Meeting
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Chordoma

PRESIDENT'S ADDRESS

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CHORDOMA is a relatively rare tumour which arises from the remnants of the notochord.

The Notochord

The notochord in the embryo extends from the spheno-occipital junction to the coccyx. It has been shown that in the human, at its upper end, it approaches the inner surface of the sphenoid and may lie in close proximity to the dura. More caudally it comes closer to the pharyngeal surface of the occipital bone. In both these situations remnants of chordal tissue have been observed on the surface of the bone. The notochord later becomes divided into segments with the development of the vertebral column, and disappears as an entity. Within the vertebral bodies occasional vestigial remnants have been observed and in the intervertebral discs the notochord is represented by the nucleus pulposus. I am indebted to Professor E. W. Walls for these illustrations of the notochord in a 21 mm. (seven weeks) human embryo cut in the sagittal plane (Figs. 1A, B).

Luschka in 1857 was the first to discover small jelly-like protrusions into the skull from the region of the clivus of Blumenbach and these were described by Virchow in 1857. He considered that they were cartilaginous in origin and named them "ecchondrosis physaliphora", believing that the vacuolated mucus-containing cells were degenerated cartilage cells.

Müller in 1858, after a study of the notochord in the fœtus and of notochordal rests in man and animals, was the first to put forward the view that these excrescences were derived from the notochord. His view was not upheld and the opinion of Virchow, as to their cartilaginous origin, was generally accepted until Ribbert in 1895 finally established their origin from the notochord, and that they were in fact "ecchord-

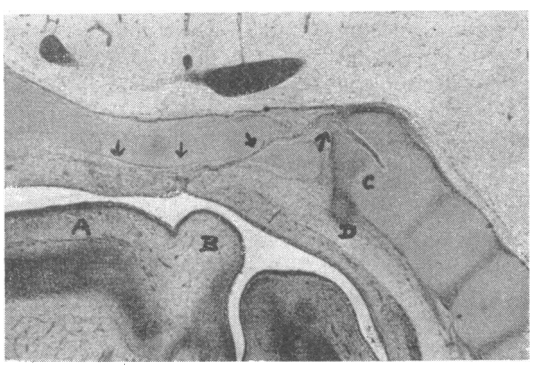

FIG. 1A.

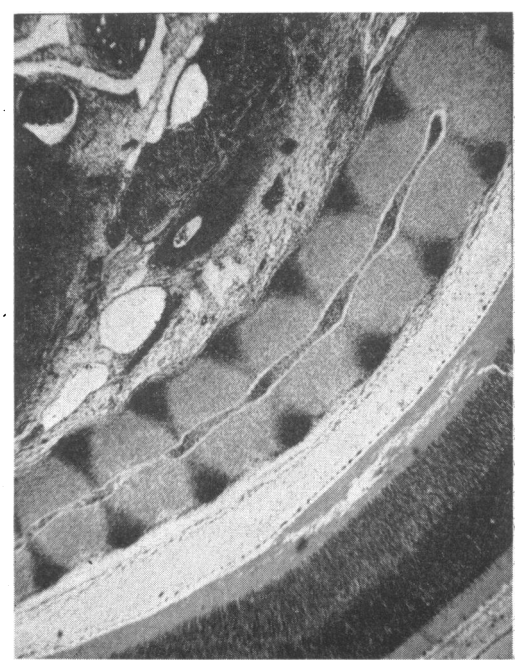

FIG. 1B.

FIG. 1.—Sections of 21 mm. (seven weeks) human embryo in sagittal plane.

FIG. 1A.—Cephalic end showing notochord (arrowed) extending through odontoid peg and curving towards the pharyngeal surface of the occipital bone and then up towards the dural surface of the sphenoid. Its termination is caudal to the developing pituitary. A, Tongue. B, Epiglottis. C, Axis. D, Anterior arch of atlas.

FIG. 1B.—Caudal end of fœtus showing the remains of the notochord in the developing vertebral column. Segmentation is commencing with the development of bulges to form the nucleus pulposus of the intervertebral discs.

oses". He was the first to apply the term "chordoma". He conducted experiments on rabbits, piercing the intervertebral discs and allowing protrusion of the nucleus pulposus. Eventually these herniations enlarged with an appearance in all ways resembling that of a chordoma.

Ecchordoses, or heterotopic notochordal remnants in the spheno-occipital region, are uncommon, but are occasionally found if carefully sought at autopsy. Ribbert found them in 2% of examinations. Stewart and Morin (1926) found 4 in 350 autopsies, and Willis (1953) found 5 in over 1,000 examinations in which he had looked for them. They are quiescent, symptomless protrusions consisting of irregular vacuolated cells in a mucoid matrix. Similar heterotopic notochordal remnants have been described in the coccygeal region. It seems probable that they may in some cases be the starting point of a chordoma.

Several cases of chordoma, a progressive tumour causing symptoms, were described in the region of the clivus in the first decade of this century, and sacrococcygeal chordoma was first recognized in 1910 (Alezais and Peyron, 1914; Berard *et al.*, 1922; Feldmann, 1910). In 1922, Stewart described the first case in Britain, a sacrococcygeal growth, when there were only 26 cases in the literature. Since then there have been increasing numbers of cases reported (Alexander and Struthers, 1926). Mabrey (1935) reviewed the literature up to that date and collected reports of 150 cases. Harvey and Dawson (1941) reviewed 240 cases, and in 1944 Faust *et al.* brought the total up to 252. Dahlin and MacCarty (1952) reported 59 cases from the Mayo Clinic and in later years there have been smaller series and individual cases. Crawford (1958) described the staining reactions of these tumours and added a further 6 cases. The total number described up to the present must be more than 350.

This present study is based on observations on 29 cases which have not previously been reported. 4 of them were under the care of Sir Stanford Cade at the Westminster Hospital, and I am indebted to him for placing his cases at my disposal. The remainder have been in the Middlesex and Mount Vernon Hospitals, where I have had the opportunity of studying them and of treating the majority of them.

The cases of chordoma which have been described arise most frequently at the upper and lower extremities of the vertebral column, the portions which are the last to develop. 90% are

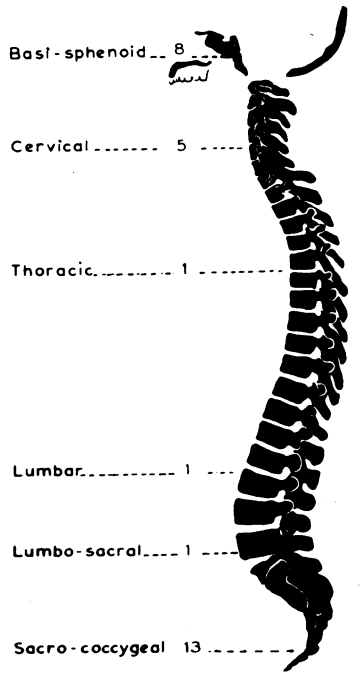

FIG. 2.—Sites of origin of chordomata in author's series.

in the spheno-occipital and sacrococcygeal regions, but they also occur in the cervical, thoracic and lumbar regions (Fig. 2, Tables I and II).

TABLE I.—SITES OF ORIGIN IN AUTHOR'S SERIES

Spheno-occipital	8
Cervical spine	5
Thoracic spine	1
Lumbar spine	1
Lumbosacral spine	1
Sacrococcygeal	13

The distribution is similar to that in other series.

TABLE II.—SITES OF ORIGIN IN OTHER SERIES

	Present series	Dahlin and MacCarty (1952)	Harvey and Dawson (1941)	Mabrey (1935)
Sacrococcygeal	13	32	122	87
Spheno-occipital	8	15	88	47
Vertebral	8	12	30	14
Others	—	—	—	2
	29	59	240	150

Age and Sex

Chordoma has been reported in any age group from infancy to the very old. They are more frequent in later life and those arising in the sacrococcygeal region are usually in an older age

group than those in the region of the clivus or elsewhere in the spine.

In the present series the youngest was 8 and the oldest 66. All but 3 were over 30 and more than half were over 40. The average age of patients with tumours in the sacrococcygeal region was 49 years, as against 36 years for the remainder. In the reviews of Harvey and Dawson, of Mabrey and of Dahlin and MacCarty, there was a preponderance of males over females in the proportion of about 2 to 1. In this series, however, the sex incidence is nearly equal. 15 were male and 14 female.

It has been pointed out that in the sacrococcygeal tumours there is a frequent history of trauma preceding the development of the tumour. In 4 of the cases which I have studied there was such a history of a severe fall followed by persistent pain, leading on to recognition of a tumour. 3 of them were sacrococcygeal and the other one was in the lumbar region. The observation of a history of trauma is an interesting one, but a correlation of cause and effect must be treated with some reserve. There is always a tendency in any case of tumour formation for the patient to remember some particular blow or fall preceding the discovery of the growth, and in sacrococcygeal chordomas there is the possibility that the presence of the neoplasm may have produced some weakness of the legs and been instrumental in the fall. As Cappell (1928) has pointed out, the experiments of Ribbert are, however, suggestive that trauma may play a part in the development of these tumours.

Pathology

The typical chordoma is a slowly growing, locally invasive tumour which causes irregular destruction of bone and extends into the soft tissues, tending to compress and push them aside and erode them rather than to infiltrate them. The bone at the site of origin may show extensive absorption and the periosteum and cancellous bone at the edges of the defect, where the tumour has broken through into soft tissue, are expanded and pushed out over the soft tissue mass.

Macroscopic appearance (Fig. 3).—On cut section the tumour has usually a variegated appearance. Much of it is solid, of a gelatinous consistency, and there may be large cystic areas filled with a semi-fluid mucinous material.

The tumour may be divided into lobules by bands of fibrous tissue and it may appear to have a fibrous capsule around it. In some parts there may be areas of necrosis and others may be deeply pigmented from old hæmorrhages, and in some cases there are bone fragments apparently carried out with the advancing tumour from the vertebra of origin. In one of the cases in this series there was considerable calcification in the tumour.

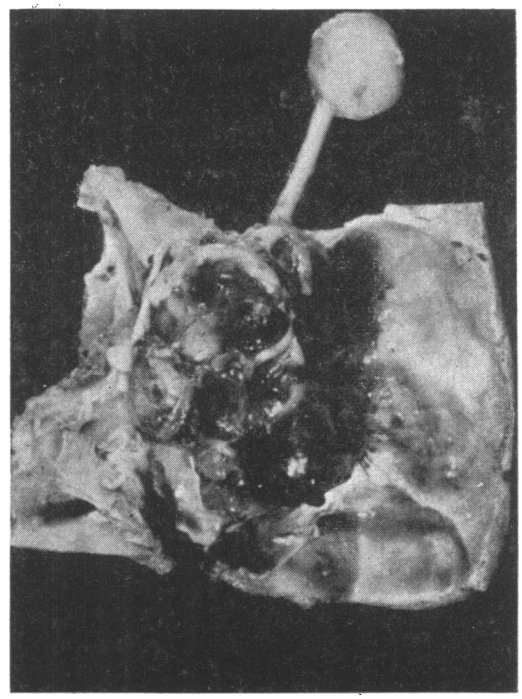

FIG. 3 (*Case I*).—Macroscopic appearance of a chordoma.

Histology (Fig. 4).—The histological appearances of chordoma have been described in detail in several publications. They show a picture in which there is great variation. The cells may be packed together in solid irregular groups separated by connective tissue or by a mucinous matrix. They may be in cords or trabeculæ and in the case of Alezais and Peyron (1914) there was the presence of regular cavities lined with cuboidal cells.

There is great variation in the size of the cells. In the youngest part of the tumour there are small polyhedral cells, and in the older parts the cells are larger with vacuolated cytoplasm and intercellular vacuolation. These were described as physaliphorous cells by Virchow, and are the characteristic feature of chordoma. The blown-up physaliphorous cells may gradually lose their cell outline and in parts there may be the appearance of a vacuolated syncytium, with few cells swimming in a sea of mucus. The vacuolated

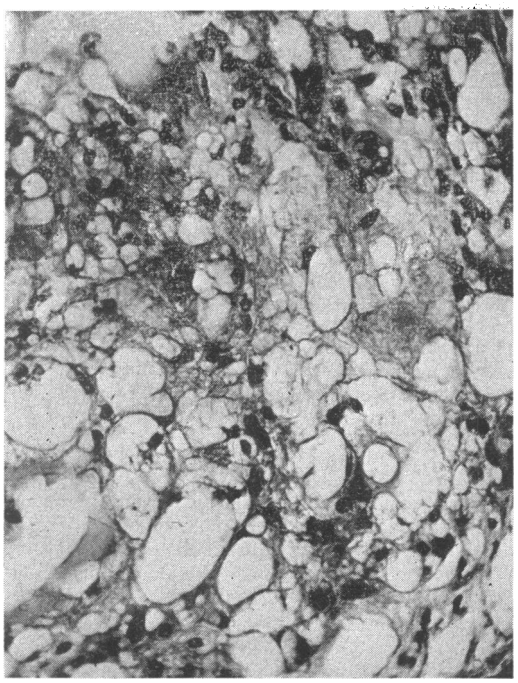

FIG. 4.—Histological appearance.

cells may have a signet ring appearance and vacuoles in the nucleus have been described. In one of Willis's cases there were many giant cells, some of huge size, and both Cappell (1928) and Willis (1953) have described a distinct hyaline sheath delimiting the chordoma from the surrounding tissues. Cappell has also pointed out that the great variation in histological appearance of chordoma is consistent with the histological appearances of the notochord in its various stages of development. In some parts of a chordoma a definitely spindle cell appearance has been described, closely resembling fibrosarcoma. In some, the mucinous matrix and the vacuolated cells may be difficult to distinguish from cartilage and there may be difficulty in differentiating chordoma from chondrosarcoma, a differentiation which Crawford (1958) considers can be made with special staining methods. Lichtenstein (1952) draws attention to the possibility of a false diagnosis of colloid carcinoma of the rectum being made in sacrococcygeal chordomas.

Stewart (1922) considered that chordoma could only be diagnosed with certainty if the characteristic vacuolation and mucin production are present in at least a portion of the growth.

Metastases in Chordomas

The usual course of a chordoma is to increase gradually but slowly, and there may be a history of discomfort or pain extending over some years before a diagnosis is made. In some cases, however, the progress may be much more rapid and there may be a history of only a few months before a tumour of considerable size is found, and in many of the cases described the rate of growth has increased considerably in the later stages of the patient's illness. Chordoma, particularly in the sacrococcygeal region, may develop into an enormous tumour weighing many pounds. The eventual outcome in the majority of cases has been death, due to local extension of the tumour mass causing interference with function by reason of its bulk, or by involvement of nerves.

Distant metastasis, though rare, has been reported and described in detail. Metastasis may be to lymph nodes, liver, lungs and to subcutaneous tissues, and apparently occurs both by lymphatic channels and by the blood stream. Stewart's case in 1922 had metastases in the buttock and in the scapular region. Willis found metastases in 2 of his 5 cases, one of them having blood-borne metastases in many organs and the other hepatic metastases.

When Graf described his case with multiple metastases in 1944, there was a total of 10 cases with metastases in the literature. One of the 6 cases described by Crawford in 1958 had pulmonary metastases. There were no cases which have so far developed metastases in the present series, and Dahlin and MacCarty (1952) found none in their 59 cases.

Clinical Features

The clinical features of chordoma will obviously vary according to the site of the tumour.

The basisphenoid region is the second most common site for these tumours to arise. The clinical features in this situation are variable. They may be those of a slowly growing tumour, gradually infiltrating upwards from the base of the skull and causing nerve palsies and increased intracranial pressure, and there may be no evidence of any spread downwards into the nasopharynx. Until a histological diagnosis is made they may be indistinguishable from other tumours arising in this situation, such as craniopharyngioma or even an intracranial extension of a carcinoma of the nasopharynx hidden in the fossa of Rosenmuller.

I am indebted to Dr. S. P. Meadows and Sir Stanford Cade for the details of a patient who was under their care and who exemplified this type of tumour.

Case I.—Male, aged 54, admitted to the Westminster Hospital in April 1949. In September 1947 he developed diplopia and pain in the right eye with occasional twitching of the right side of the face. In March 1948 slight proptosis of the right eye was observed with complete right VI nerve palsy. On admission he was found to have right VI, IX, X and XI cranial nerve palsies. Radiographs showed destruction of the posterior clinoids on both sides and of the dorsum sellæ and the tip of the right petrous bone.

He was treated by X-rays on several occasions in 1949 and 1950, with improvement, and by the 10-gram teleradium unit for recurrence in 1954. His condition deteriorated and he died in March 1955, seven and a half years after his symptoms began.

At post-mortem examination of the head and contents after removal of the cerebral hemispheres, a large nodular, well-circumscribed tumour was seen occupying the medial part of the right middle fossa and basisphenoid region. Laterally the tumour was entirely extradural, the dura being stretched over it but not eroded. Posteriorly it was pushing the brain-stem backwards, deeply indenting the anterior aspect of the pons. The sella turcica could barely be defined on the surface of the mass which was growing upwards and pushing the optic chiasma to the left of the mid-line. Both optic tracts were attenuated and soft and tightly stretched over the upper surface of the growth.

On removing the tentorium, cerebellum and dura mater, the growth was seen firmly attached to the base of the skull and was 6 to 7 cm. in diameter. The bulk of the tumour lay on the right of the mid-line, and in colour it varied from green to white, on the whole appearing vascular.

It was difficult to identify the cranial nerves on the right side of the brain-stem, but those that were identified were seen to be thinned by stretching. The anterior surface of the left petrous bone had a slightly worm-eaten appearance, but the only bone to be extensively destroyed was the basisphenoid bone.

Fig. 3, illustrating the macroscopic appearance of a chordoma, is taken from this case.

Histologically there was some modification, presumably due to irradiation, in the form of fibrosis and large cells with bizarre, hyperchromatic nuclei, but the growing margin, including that in bone at the base of the growth, showed the typical branching strands of physaliphorous cells and the production of mucin.

3 of the 8 patients with basisphenoid growths presented with swelling in the nasopharynx (Cases II, III and IV), and one with a tumour both inside the cranium and in the nasopharynx (Case V).

Case II.—Woman, aged 63, who had nasal obstruction for many years and was found to have a fleshy tumour growing from the vault of the nasopharynx. This was removed by diathermy after transpalatal approach by Mr. Douglas Ranger in June 1958, and was found to be a chordoma. At present there is no sign of recurrence.

Case III.—Man, aged 33, who had a three months' history of nasal obstruction and deafness in the left ear. After biopsy he was treated by supervoltage irradiation with the Theratron cobalt unit for a mass filling the nasopharynx. There was marked regression of the tumour when his treatment finished three months ago.

In neither of these cases was there any radiological evidence of bone involvement despite attempts to demonstrate it by special views and tomographs.

Case IV.—Woman, aged 29, admitted to the Middlesex Hospital in June 1933. She had a smooth, slightly lobulated tumour in the posterior wall of the nasopharynx. A little gelatinous material was obtained on aspiration. X-ray examination of her skull showed no evidence of bone erosion, though the posterior part of the sphenoidal sinus was opaque. On frozen section the tumour was stated to be a nonmalignant mucous gland tumour, though Professor R. W. Scarff at the time considered that it was a chordoma.

She was treated by radium needle implantation with regression of the tumour, but a year later was readmitted and died. Autopsy showed an intracranial chordoma which indented the inferior margin of the pons.

Case V.—Man, aged 37, referred to Mount Vernon Hospital from the Westminster Hospital in April 1955. He had a year's history of increasing pain in the right side of the forehead and face, and had been under observation for six months. He had a left VI nerve palsy and gradually developed a V and III nerve palsy. There was a tumour in the right side of the nasopharynx.

X-ray examination showed a partly calcified tumour in the mid-line, obliterating the pituitary fossa (Fig. 5). A left carotid arteriogram showed displacement of the carotid siphon but no blood supply to the tumour.

The mass in the nasopharynx was found to be cystic and was punctured and material removed for histology. The histological report stated that it was not particularly suggestive of a chordoma and that it was probably mucinous degeneration in a connective tissue tumour. He was treated by the Theratron telecobalt unit, receiving 7,200 r over fifty-seven days. Three days after completion of treatment he had an intracranial accident, and died two weeks later.

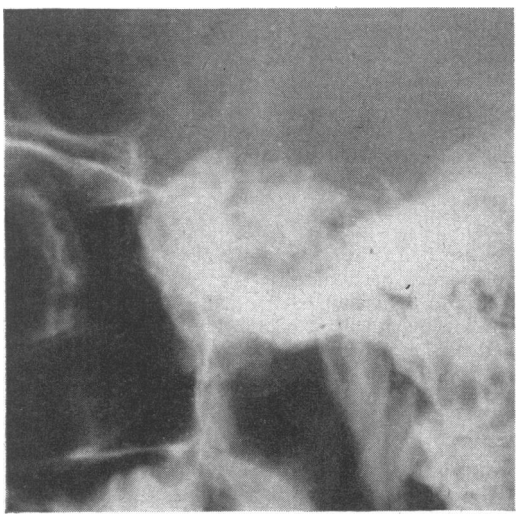

FIG. 5 (*Case IV*).—Partly calcified chordoma of basisphenoid region.

Considerable interest was taken in the diagnosis of this condition because, although we had made a provisional diagnosis of chordoma, the biopsy from the nasopharynx failed to confirm this and it was considered that the dense calcification seen on radiographic examination made this diagnosis unlikely.

At post-mortem examination of the head and contents there was a massive tumour 6 cm. in its longest diameter occupying the sphenoidal region in the mid-line, invading the pituitary fossa and surrounding the optic nerves. The dura was elevated over the rounded contours of the tumour. It was bulging into the nasopharynx as a smooth, flat, domeshaped swelling. An extension spread into the right middle cranial fossa, infolding itself deeply into the temporal lobe of the brain; a large recent hæmorrhage surrounded this extension within the temporal lobe, pressing on the internal capsule and basal ganglia on this side. The cut surface of the tumour was gelatinous with a bluish tint, flecked with hæmorrhage. It was thought that the hæmorrhage might have been caused by regression of the tumour as the result of irradiation. Histologically several sections were examined and finally the typical structure of chordoma with physaliphorous cells was found.

X-ray Appearances

The X-ray appearances of chordoma are usually of a lesion which causes irregular destruction of the affected bone, bursts through the cortex and forms a soft-tissue mass extending out from the bone. The edges of the bone where the tumour has burst through may be expanded outwards and flakes of bone may be carried out in the soft tissue tumour. Arteriography shows displacement of vessels by the tumour but no new blood supply to the tumour mass. Calcification in the tumour has been described and in fact is stated as one of the four cardinal signs of sacrococcygeal chordoma by Hsieh and Hsieh (1936). Dahlin and MacCarty (1952) found it in 2 of the 15 clivus tumours in their series and Wood and Himadi (1950) observed it in 6 of 7 clivus tumours. Mabrey (1935), however, stated "the lesion is not a bone tumour and bony proliferation is not to be expected".

Only one patient in the present series showed extensive calcification. The others had little or none, and I think a more typical example of spheno-occipital chordoma with intracranial spread is the patient illustrated in Fig. 6, at present under the care of Mr. Valentine Logue.

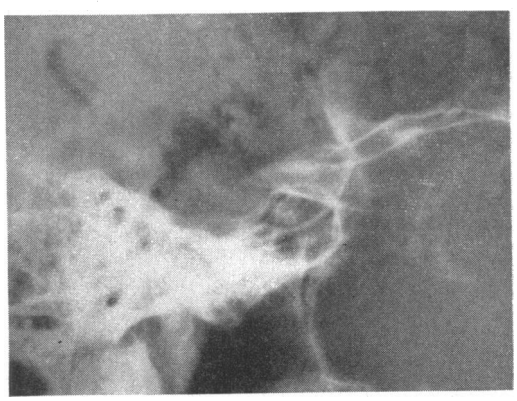

FIG. 6.—Chordoma of basisphenoid region. Bone destruction. Soft-tissue tumour outlined by encephalography.

There is bone destruction and no new bone formation.

The clinical features of vertebral chordoma depend on the site of origin of the tumour, not only on the level of the vertebra involved but also on the direction of spread. Pain is usually the earliest symptom, and it may be local at the site of origin of the tumour or be referred along a nerve distribution and be accompanied by tingling and paræsthesia. There may be the typical signs of pressure on the spinal cord as seen in other types of tumour, with nerve palsies and ultimately paraplegia. On X-ray examination there may be collapse and distortion of the vertebræ. A soft-tissue mass may protrude, most frequently anteriorly but sometimes at the side of the vertebræ or even posteriorly, and will cause symptoms which will vary according to the level of the spine which is involved.

These features, as far as the cervical region is concerned, are exemplified in the following 3 cases:

Case VI (Fig. 7).—Woman, aged 31, admitted to the Westminster Hospital in 1950. For two and a half years she had persistent pain in the neck. In September 1949 she developed a lump in the right side of the neck. After biopsy she was treated by 200 kilovolt X-ray therapy and the 10-gram teleradium unit, receiving over three months a tumour dose of approximately 5,000 r. There was slow but gradual improvement in pain, and the tumour decreased to about one-third of its former size. She died, however, in May 1952.

Case VII.—Girl, aged 8, admitted to Mount Vernon Hospital in October 1956. She was in normal health until one month before, when she developed pain in the right hypochondrium. She had marked wasting of scapular muscles and both sternomastoids, and of her arms and forearms. There was weakness of all groups of muscles in both legs. X-ray examination showed erosion of the spinous process and right lateral mass of the fifth cervical vertebra. Biopsy showed chordoma. She was treated by the four million volt linear accelerator to a tumour dose of 5,000 r over thirty-five days, and after an initial favourable response relapsed and died seven months later.

Case VIII (Fig. 8).—A man aged 41 in April 1954 complained of swelling in his neck, that his collars were too tight and that he had slight discomfort on swallowing and slight pain in the right arm with pins and needles in the fingers. He was found to have a hard mass behind his larynx, pushing his larynx forwards.

After biopsy he was given a course of treatment with 200 kV X-rays to a tumour dose of 4,750 r, following which there was a decrease in the mass and he felt much better. In March 1955 he was referred to Mount Vernon because of a return of pain and tingling in his hand, and he was treated by the

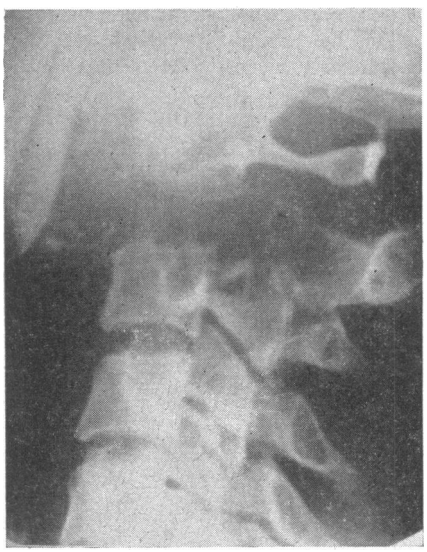

FIG. 7 (*Case VI*).—Chordoma involving 2nd cervical vertebra with soft-tissue tumour extending into pharynx.

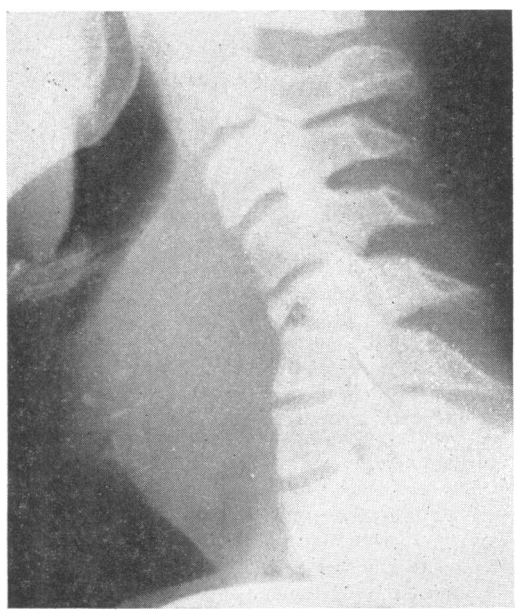

FIG. 8 (*Case VIII*).—Chordoma of fifth cervical vertebra with soft tissue mass pushing forward the posterior pharyngeal wall and displacing the larynx.

Theratron cobalt unit. It was not possible to see his larynx on indirect laryngoscopy because of distortion by the tumour mass. There was regression of the tumour and his cords could be seen with the laryngeal mirror. After initial improvement in his pain and tingling he began to deteriorate, developed paraplegia and died.

Case IX was the only patient with a chordoma in the thoracic region. He was admitted to the West Middlesex Hospital under the care of Mr. Illtyd James in September 1947, with symptoms and signs of a spinal cord tumour. The physical signs suggested a lesion of the third thoracic segment. After laminectomy he was found to have extensive thickening of the dura mater from the first to the fifth thoracic vertebræ. The extradural mass was found to be spreading a considerable distance up and down the extrathecal space. Sufficient of the tumour was removed from the posterolateral and anterior aspects of the cord to relieve pressure. This was found to be a chordoma with mucoid degeneration. Following the operation there was some return of power in his legs. He was given post-operative treatment with X-rays at 200 kV with an estimated dose at the lesion of 3,650 r. He had been in a home for paraplegics and at present, nearly twelve years later, he is reported as unchanged and able to do a full day's work in a factory.

Case X, the patient with a lumbosacral tumour, was a woman aged 29 when admitted to the Middlesex Hospital in 1953 with a most extensive lesion. At the age of 12 she fell down some steps and shortly afterwards developed severe pain. X-ray examination showed no abnormality, but the pain persisted. She had her spine manipulated as she became unable to straighten her legs. At the age of 17, foot-drop developed and she lost sensation in her feet. She later lost control of her bladder and her bowel. At the age of 24 she was manipulated by an osteopath who, after five years, recommended her to seek advice at the Middlesex Hospital.

In June 1953, after extensive investigations, no treatment was advised as we considered that conventional X-ray therapy offered no hope of improvement. She was able to control her bladder and bowel function and was able to get about with crutches. X-ray examination (Fig. 9) showed osteolytic areas in the third, fourth and fifth lumbar vertebræ extending anteriorly from the posterior aspects of the bodies. There was a similar appearance of the upper two-thirds of the sacrum. There is well-marked kyphosis at the fourth lumbar vertebra, which shows considerable destruction.

In 1954, when we had treated several cases of chordoma with the Theratron cobalt unit, I decided to treat this patient, with the object of at least preventing further extension of the tumour. A biopsy was carried out from the lumbar area, as it was presumed to be the area of most recent extension.

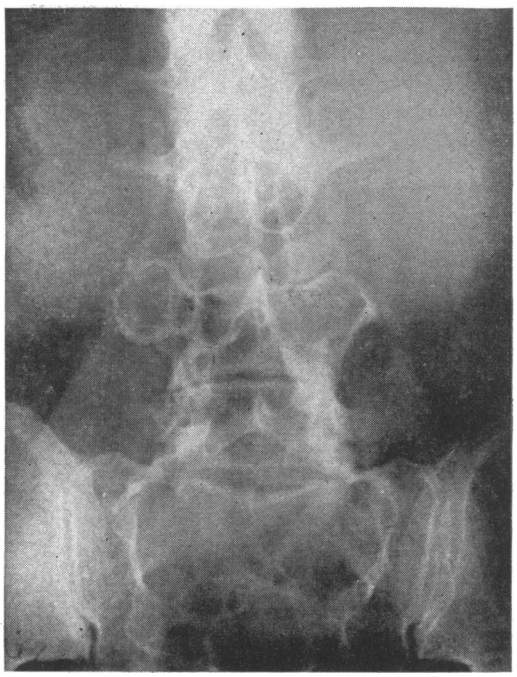

FIG. 9 (*Case X*).—Extensive lumbosacral chordoma.

Although typical physaliphorous cells were not seen the appearance was highly suggestive of chordoma.

She was treated by the Theratron at Mount Vernon, receiving a tumour dose to the lumbar spine of 5,600 r over forty-five days with a cycling rotation technique, and two months later further treatment was given to the sacral area to a tumour dose of 5,800 r over fifty-five days with a similar technique. It is now four years since the treatment was given and there is no appreciable change in her symptoms. She is still at work as a telephonist, over 21 years since her first symptoms began.

The cases of sacrococcygeal chordoma have formed the largest group in this series, as has been the case in all the larger reported collections. A clear clinical picture of the condition can be built up from a study of the 13 cases in this group. In all of them without exception, pain was the first symptom of which they complained. It was described as a gradually increasing aching pain, and its distribution varied from the lower back, the buttocks, the hips, over the sacrum to extension down one or both legs. It was usually followed by the recognition of a lump, either externally or within the pelvis, by reason of its size causing pressure and interference with micturition or bowel action. Later there develops loss of control over bladder and bowels due to interference with their innervation and, later still, there

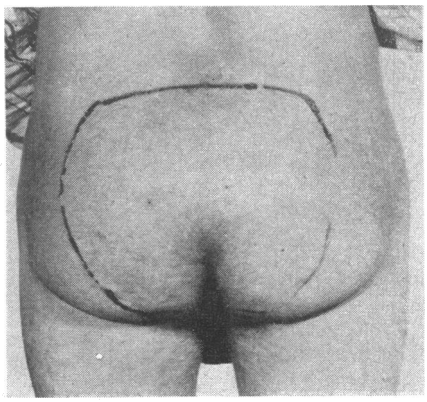

FIG. 10.—Sacrococcygeal chordoma. Tumour involving sacral region and both buttocks.

may develop nerve palsies with perhaps foot drop and weakness of legs.

The tumour itself, having eroded bone, in most cases extensively, appears to push the soft tissues aside and to compress them rather than to infiltrate into them. When an external mass is present it may appear in the mid-line over the sacrum, particularly in its lower part, or it may appear in either buttock, or there may be a generalized swelling of the whole region of sacrum and medial parts of both buttocks. It is generally firm, smooth and elastic to feel, rather than stony hard. It is not painful and there is no tenderness and no signs of increased vascularity (Fig. 10).

The tumour within the pelvis can be felt, sometimes of vast extent, almost filling the pelvis and pushing the rectum forward. It fills up the concavity of the sacrum and may arise from the midline or from either side. In all the cases which I have observed it is smoothly irregular and lobulated and has a definitely firm, elastic feel. It is firmly fixed posteriorly and invariably too large and too high for the examining finger to reach more than the lower part of it. The rectal wall, although pushed forwards, is freely mobile and not attached to the tumour. Radiologically there is gross destruction of the lower part of the sacrum and coccyx with irregular edges of the destroyed bone and areas of rarefaction extending upwards into the sacrum where the tumour is infiltrating. There may be a large soft-tissue shadow which causes difficulty in recognizing details of the edges of the destroyed bone. In the lateral view the sacrum is expanded with irregular areas of rarefaction. Arteriography shows displacement of vessels and no new vessel formation (Figs. 11 and 12).

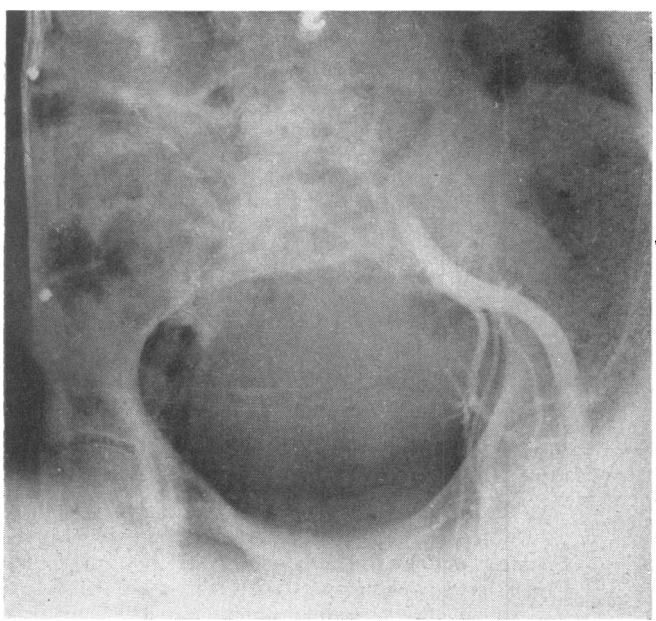

FIG. 11.—Sacrococcygeal chordoma. Extensive destruction of sacrum. Massive soft tissue tumour. Displacement of blood vessels and no new blood vessel formation.

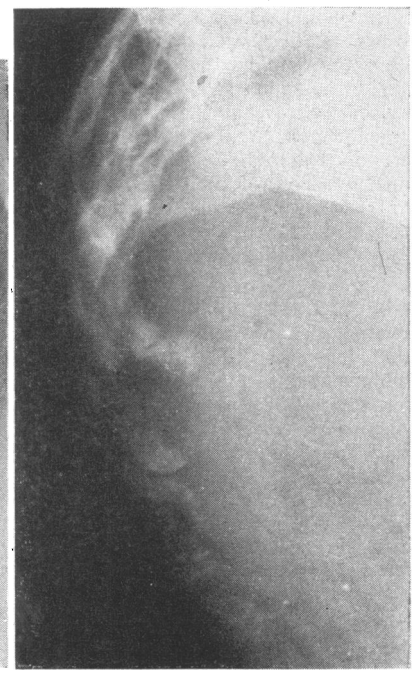

FIG. 12.—Sacrococcygeal chor
Partial destruction of sacrum.

Hsieh and Hsieh (1936) described four cardinal features: (1) Expansion of the sacrum in the lateral or anteroposterior diameter; (2) areas of rarefaction or destruction that tend to be loculated; (3) trabeculation of the undestroyed bone; (4) areas of calcification in the tumour.

Some illustrative examples are the following:

Case XI.—A man aged 53 who came to the Middlesex Hospital in 1939 with a recurrent chordoma. Following a six years' history of pain he had partial removal of a sacral tumour in 1935, followed by a radium needle implant and later X-ray therapy. He had a mass the size of a football in the right buttock extending across the midline to the left buttock. It was hard, but cystic areas were palpable. At the Middlesex Hospital he had X-ray therapy with palliative intent, gradually deteriorated and died twelve years after the development of his first symptoms.

Case XII.—Man, aged 64, referred to the Middlesex Hospital in 1943. He had a six months' history of lumbago. While his doctor was removing a chronic olecranon bursa under local anæsthesia the patient complained of the discomfort of the operating table. He was turned over and a softish tumour the size of an apple was found over the sacrum. This was thought to be a lipoma and removal was attempted forthwith. It was found to be a cystic tumour which shelled out easily, but large quantities of friable material came away at the same time with small pieces of bone. The patient developed urinary retention. In 1943 he was given 200 kV X-ray therapy, 5,250 r in forty-one days, and remained well until 1948. He was then given a further 5,000 r in sixty-six days for a recurrence of the tumour. He gradually deteriorated and died with loss of bladder function, uræmia and infection in 1952, nine years after his first symptoms began.

Case XIII.—Man, aged 58, referred to Mount Vernon by Sir Stanford Cade in February 1955. A year previously he had pain in the region of the sacrum, and in December 1954 he first noticed a swelling over the sacrum to the right of the mid-line and an area of numbness between the swelling and the anus. On rectal examination he had a lobulated semicystic tumour the size of a tangerine, pushing the rectum forwards, and there was a soft diffuse mass overlying the right half of the sacrum. X-ray showed typical destruction of the lower half of the sacrum towards the right side. Biopsy showed chordoma. He had treatment by the Theratron cobalt unit, 6,500 r being given over fifty days, with complete rotation technique. He is well at the present time with no symptoms and no tumour palpable, four years since his treatment.

Case XIV.—A woman aged 55 came to the Middlesex Hospital in 1951 complaining of low back pain on bending and stretching. X-ray examination at that time showed no bony lesions. The pain became worse and extended into the right buttock by 1955. In 1956 she became obstinately constipated and developed pruritus. An X-ray taken in August 1956 showed some destruction of the sacrum. By January 1957 she had gone through the stages of stress incontinence to complete incontinence of urine.

In April 1957, when I first saw her, she had pain down her right leg and swelling of both buttocks. After biopsy, which showed chordoma, she was treated at Mount Vernon by the Theratron to a dose of 6,300 r in thirty-nine days with cycling rotation. She had already had a colostomy and then had deviation of her urine into an ileal loop carried out by Sir Eric Riches.

Although the tumour regressed she still had foot-drop and severe pain, which was treated by chordotomy by Mr. Valentine Logue. At present she is bedridden and her nerve involvement and consequent muscle weakness are too severe for any progress to be made in rehabilitating her. I suspect that she may be developing a recurrence.

There have been two interesting cases in this group who had second primary tumours of a different nature (Cases XV and XVI).

Case XV.—Man, aged 49, treated by nephrectomy by Sir Eric Riches in 1952 for a carcinoma of the right kidney with post-operative X-ray therapy. He had complained of low back pain for some years and had had a manipulation for this. In 1956 his pain was worse and a swelling was felt over the right side of the sacrum. It was thought to be a metastasis from the kidney and a biopsy was taken. This proved to be a chordoma. He was treated by the Theratron but was unable to complete the course owing to infection of the biopsy wound, and he died six months later.

Case XVI.—Woman, aged 42, referred to Mount Vernon Hospital by Sir Stanford Cade in March 1957. She had an extensive sacrococcygeal chordoma with low back ache and incontinence of urine and fæces. She had already had a colostomy and a biopsy which showed chordoma. She was treated by the Theratron to a dose of 7,500 r over sixty-two days by full rotation. At that time she was advised to have deviation of her urine into an ileal loop, but she refused. The tumour regressed and she gradually regained control of her bladder so that it functioned normally. In September 1958 she had a return of pain in her ischial tuberosities and was found to have a stricture of her rectum. This was proved to be an adenocarcinoma of the rectum and was removed by Sir Ralph Marnham. Her convalescence was uneventful, except that she was slow in healing, but she is well at present and retains her bladder function.

Summary of Clinical Features

Tumours in the spheno-occipital region may present with intracranial tumour, with naso-pharyngeal tumour or both, and there is no particular symptomatology of chordoma to distinguish it from other slowly growing tumours in these regions.

Patients with these tumours are of a somewhat younger age group than those with sacrococcygeal chordomas and they do not live as long, owing to nerve involvement.

In the vertebral region, the least common site, there is pain, involvement of the cord by pressure or vertebral collapse and consequent nerve palsies, and there may be a soft-tissue tumour, protruding in any direction, but most frequently anteriorly. In the sacrococcygeal region there is pain and a tumour either protruding posteriorly or into the pelvis, or both, and sometimes of great volume. It is lobulated and semifluctuant. There is extensive bone destruction of the lower end of the sacrum and coccyx. There may be nerve involvement causing paralysis of bladder and rectum and of the nerves supplying the lower limb. It is the most frequent site of chordoma; the patients are mainly over 40 and they tend to live for many years, even with active growth present.

Treatment

In all publications on the subject it is recognized that to devise any curative treatment for chordoma is a matter of great difficulty. Complete surgical ablation is hardly ever possible owing to the situation in which these tumours arise. It is not possible to remove the base of the skull and it is not possible to remove the sacrum in its entirety. Partial excision is an excellent palliative, although there is usually recurrence within a few years, but many patients have been kept going for a number of years by repeated partial excisions. The case reported by Stewart in 1922 and operated on by Moynihan lasted nineteen years from the beginning of symptoms.

At the Mayo Clinic there is now a programme of attempting a more radical surgical removal of the sacrococcygeal tumours, with wider ablation of the sacrum (MacCarty *et al.*, 1952). The sacral nerves are dissected out from the tumour mass and preserved. Owing to the rarity of the condition and its slow rate of growth, it will take some years before this procedure can be evaluated.

In this series there are a number of cases who have been treated by surgery. One patient had partial removal of a tumour in the clivus region in 1952, and excellent palliation was obtained for four years, when the tumour began to grow again, and he has been referred for radiotherapy. There is in addition Ranger's patient with nasopharyngeal presentation and no evidence of intracranial spread or bone involvement, but this is only nine months ago.

Case XVII, a girl aged 16, was the one patient with a lumbar spine chordoma. She was quite well until aged 11 when she had a fall on her lower back followed by pain in the legs and sphincter disturbances. Two to three years later she began to develop deformity of the spine, which continued to increase in spite of treatment. Her legs became weaker and she was unable to walk unaided. In 1949, Miss Diana Beck performed a laminectomy and removed a tumour of the cauda equina; the histological examination showed it to be a chordoma. Six years later she was apparently unchanged; she was able to get about with crutches and had marked sensory loss in both legs.

It is, however, the possibilities of radiotherapy which have been my chief interest in treatment in the cases which I have seen. The majority of publications on the treatment of chordoma, when the subject is mentioned at all, begin with the statement that these tumours are radio-resistant and that radiotherapy is of no value, and I believe that the majority of radiotherapists are of the same opinion.

The reports from Boston (Mabrey, 1935) state, however, that radiotherapy has been of value as a palliative in the relief of pain, and Dahlin and MacCarty (1952), from the Mayo Clinic, cite 2 cases who had complete remission of symptoms for five and a half years, and for six and a half years, having been considered inoperable and having been treated by radiotherapy alone. I became impressed with the regression that was obtained in one or two cases when conventional 200 kV X-rays were given to high dosage, and determined when supervoltage became available to attempt to treat other cases if I could find them. The first and most spectacular of these results is one which I have included in this series of 29 chordomas, although there is no histological evidence of the diagnosis. It is the only case in the series which has not a histological diagnosis.

Case XVIII.—In April 1941 a man aged 31, recently qualified in medicine, sought advice at the Middlesex Hospital. He had had calcified tuberculous glands in his neck for many years. For seven years he had been conscious of a mass projecting forwards into the pharynx from the left posterior wall. He had developed a sudden pain in his neck which limited movement of his head. There was a retropharyngeal mass pushing his palate forward, which was thought to be fluctuant, and a provisional diagnosis of tuberculous abscess was made. The mass was aspirated and some mucinous material obtained. It was thought to be incompatible with the contents of a cold abscess, and there was a hard tumour remaining. A clinical diagnosis of chordoma was made.

X-ray examination showed a retropharyngeal mass with some irregular bony destruction of the body of the second cervical vertebra (Fig. 13A). He was treated with 200 kV X-rays to a tumour dose of 9,200 r, including 5,200 r by direct intrabuccal

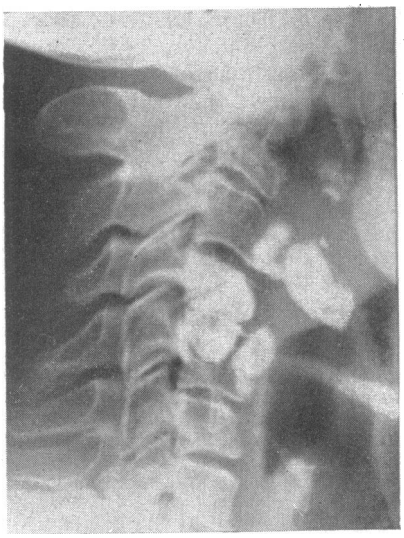

FIG. 13A (*Case XVIII*).—1941: Irregular decalcification of second cervical vertebra with soft tissue mass pushing forward the posterior pharyngeal wall.

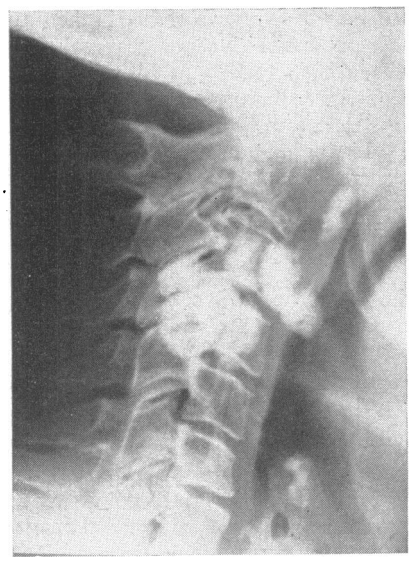

FIG. 13B (*Case XVIII*).—1955: Disappearance of soft tissue tumour. No recalcification of second cervical vertebra.

application over forty days. Three months later, in October 1941, a further tumour dose of 2,800 r was given over eighteen days and in April 1942, a further tumour dose of 1,500 r in ten days.

The patient is now well and continues his work in a specialized branch of medicine. X-ray examination (Fig. 13B) shows that the destroyed vertebra has not calcified, and he continues to wear a plastic support.

After his heavy radiation dose he has marked skin changes on his neck.

If the diagnosis were mistaken and this condition had been a tuberculous abscess, the other most possible diagnosis, I would have expected that the heavy dose of radiation would undoubtedly have aggravated the condition. It is now eighteen years since he was treated and twenty-five years since his symptoms began.

Case XIX, a woman aged 36, was admitted to the Middlesex Hospital in November 1954. For six months she had had pain down the posterior surface of the right thigh, gradually getting worse and spreading down the leg to the foot. She was found to have a smooth, firm, rounded tumour projecting into the rectum from the right. It did not extend to the mid-line and was firmly attached to the sacrum. The upper surface could just be reached by the finger. There was also a firm tumour in the right buttock. After biopsy an attempt was made to carry out a hind-quarter amputation but it was found that the tumour extended widely across the front of the sacrum upwards, almost to the brim of the pelvis, so that the sacro-iliac notch was completely obliterated on its inner aspect and the operation was abandoned.

She was treated by the Theratron cobalt unit at Mount Vernon receiving a dose of 6,100 r in fifty-two days with complete rotation technique. By the end of the treatment the tumour in the buttock had disappeared and the rectal tumour was no longer palpable.

At the beginning of treatment she had pain in the right foot and ankle which prevented her putting her foot to the ground. X-ray examination showed a patchy osteoporosis consistent with Sudeck's atrophy. Vigorous physiotherapy was continued after the completion of her radiotherapy. The pain in her right foot disappeared and she began to walk normally. We have wondered at the cause of the Sudeck's atrophy and consider that it might have been caused by damage to sympathetic nerve supply during the course of the attempted hind-quarter amputation.

She is now well, with no sign of recurrence, over four years since her radiotherapy, but is again walking badly because of severe arthritis of her right hip also due, I believe, to the damage to her nerve supply.

Histologically this is not completely definite, but I believe she had a chordoma and have counted her in this series.

When dealing with a tumour which is slowly growing and which rarely metastasizes, in which the natural history of the disease extends over many years, it takes many cases and a long period of time to arrive at definite conclusions as to the curative value of any method of treatment. Some of the cases which I have described and others who had tumours in the sacrococcygeal region, who are now alive and well without sign of

disease, may develop recurrence in the future, but of the 8 patients with sacrococcygeal chordomas who have been treated at Mount Vernon with supervoltage radiation since 1954, 6 are at present without symptoms or signs of the disease, 1 of these 6 having had a second course of treatment for recurrence a year ago, after freedom for four years. Of the remaining 2, 1 has probable recurrence after two years' freedom, and 1 is too early to evaluate.

I hope I have shown that radiotherapy has some value in the treatment of chordoma. I consider that, at least as a palliative, it should be given in preference to partial operation in sacrococcygeal tumours.

Supervoltage radiotherapy, and in this series it has been mainly by the Theratron cobalt unit, has given the opportunity of delivering the high dose necessary without excessive damage to the overlying and surrounding tissues. Rotational techniques are also particularly appropriate, as we believe that in this way we can more accurately limit the dose to the volume of tissue required. This is of considerable importance as there must be some danger of causing transverse myelitis, which has been described as the result of irradiation, if excessive dosage is given to the spinal cord.

With supervoltage irradiation, given over a prolonged period of time, and by limitation of the volume of tissue irradiated, it may be possible to operate with safety in some cases, even after such a heavy dose as 7,500 r delivered over sixty-two days, as had to be done in the case of the woman who developed carcinoma of the rectum (Case XVI).

I would like to acknowledge with gratitude the help that I have had from many clinical colleagues, especially Sir Stanford Cade, and from Professor R. W. Scarff, Dr. A. C. Thackray, Dr. A. D. Thomson, and Dr. R. E. Cotton of the Bland-Sutton Institute of Pathology, from Dr. G. W. S. Andrews of Mount Vernon Hospital Pathology Department, from Dr. Campbell Golding and Dr. H. S. Murray, and from Mr. M. S. Turney of the Photographic Department of the Middlesex Hospital, who has produced my slides.

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