CHORDOMA WITH A REPORT OF TWO CASES

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WHILE still comparatively rare, cases of chordoma are being reported with greater frequency, probably owing to better recognition of the condition. As far as can be ascertained, none have as yet been reported in the Canadian literature. Two cases which have recently come to our attention, therefore, seem worthy of mention.

Luschka, in 1856, first described small benign jelly-like nodules at the clivus Blumenbachii, which Müller, in 1858, concluded were of notochordal origin. This was confirmed a few years later by the work of Ribbert, who also noted their occurrence in 2 per cent of autopsies. More recently, Stewart and Morin[7] examined 350 consecutive autopsies specially for these nodules and found them to occur in slightly less than 1.5 per cent. Stewart[6] designated these small benign tumours "Ecchondrosis physaliphora sphenooccipitalis," referring thus to their origin and to the bubble-like appearance seen on microscopic examination. When large enough to give rise to symptoms, they are termed "chordomas," as are growths of similar origin in the sacrococcygeal region and elsewhere along the course of the notochord.

Normally, the notochord disappears early in embryonal life, remaining only as a jelly-like substance, the nucleus pulposus, in the centre of the intervertebral discs. Several observers have shown that in the fetus small remnants of notochordal tissue may lie outside this usual location, and it is these aberrant cells which are believed to give rise to tumours. Thus, they may arise anywhere along the course of the primitive notochord. The commonest sites, however, are at its extremities, *i.e.*, the spheno-occipital and sacro-coccygeal regions. The spheno-occipital type gives rise to pressure symptoms characteristic of a tumour at the base of the brain — headache, vomiting, and optic neuritis. Various cranial nerves may become involved, as they are embedded in the growth. Tumours in this area need not be very large to give rise to symptoms. The largest one is reported by Jeliffe and Larkin,[4] measuring 11 by 7 by 6 cm. In the sacro-coccygeal region, if the growth projects posteriorly, there may be signs of an external tumour only. If it projects anteriorly, there is interference with defæcation and urination and signs of pressure on the sacral nerves. In this location the tumour may become very large. Stewart and Morin[7] report one measuring 80 cm. in circumference, and Bernard, Dunet and Peyron[1] one measuring 22 by 21 by 15 cm. Willis's[9] recent case, measuring 15 inches in length and weighing approximately 200 oz., is the largest yet reported.

Chordomas are usually slow growing and locally malignant, though a number of instances of metastases are recorded. Willis' case gave widespread metastases to the lungs, liver, spleen, kidneys, heart, thyroid and skin, probably due to invasion of the iliac vein by direct extension.

CASE 1

K. J., a male, aged 62, labourer.

Complaint.—Pain and presence of swelling at the end of the spine.

Family history.—Negative.

Past illnesses.—Irrelevant.

Present illness.—In October, 1928, the patient fell on the edge of a wagon-box striking the base of his spine. He had pain for several days but no swelling was noticed. During the winter of 1928-29 he had no pain in the region of his injury. In March, 1929, he noticed soreness but no swelling. During the summer of 1929, he drove horses and had only occasional stiffness in the region of the sacrum. In the spring of 1930, he drove steadily in a car for two days, became sore, and noticed a swelling of the base of his spine. He had considerable pain after sitting for any length of time. The swelling was incised in April, 1930, but only blood was obtained. The wound healed but the swelling persisted and was incised again.

Physical examination.—July, 1930: Fairly well developed and nourished. Head, neck, thorax and abdomen showed negative findings. Usual weight 145 lb.; present weight 123 lb. Blood pressure 240/130, with slight arteriosclerosis of temporal and radial arteries. Hæmoglobin 70 per cent; white cells 6.500; erythrocytes 3,750,000.

Special examination.—Flattened over the end of the sacrum, a tumour was found, measuring approximately 4 inches in diameter and irregular in outline. It was firm in consistency and was attached to the skin, which was thickened and indurated.

At operation, the tumour was found to be irregular in outline, attached to the skin but not to the sacrum. It was not encapsulated. Through it were 6 or 7 areas of coagulated blood, each the size of a walnut. The

tumour was dissected out and a series of deep x-ray treatments was given. On October 11, 1930, another swelling in the same region was also dissected out and followed by deep x-ray therapy.

On January 23, 1931, another tumour was present on the upper portion of the left thigh posteriorly, which was of irregular outline, well encapsulated, firm and rubbery in consistency. It measured 7 x 7 x 4.5 cm. On section, it was seen to be divided into numerous compartments of various sizes by thick fibrous bands. Many

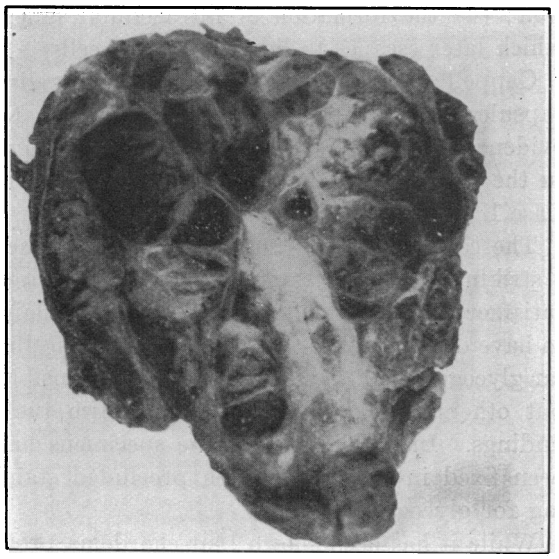

FIG. 1.—*Case 1.* Gross appearance of the tumour.

of these compartments contained a whitish mucoid material; others were brownish and evidently the seat of hæmorrhage, while others contained a mixture of the two.

Microscopically, bands of dense fibrous tissue were seen enclosing islands of tumour cells. This fibrous tissue showed some areas of lymphocytic infiltration. There were several areas of hæmorrhage. The tumour cells tended to grow in a cord-like or alveolar arrangement. They varied greatly in size. The smaller areas had one or two nuclei, were polyhedral or irregular in shape, with a solid pink-staining cytoplasm and a well-defined cell membrane. There were all gradations between these and large multinucleated cells with many large clear vacuoles and a mere rim of cytoplasm. A few whorled bodies resembling those described by Stewart and Morin[7] were noted. The nuclei also varied in size, were round or ovoid and usually contained a well marked nucleolus. No mitotic figures were seen but the nuclei were often lobulated, and frequently indented by vacuoles. Some nuclei were small and hyperchromatic. True nuclear vacuolation was not found. Running between the cells, very marked in some regions, was a bluish hazy homogeneous substance, mucin, apparently extruded from the tumour cells. In some areas, the cells

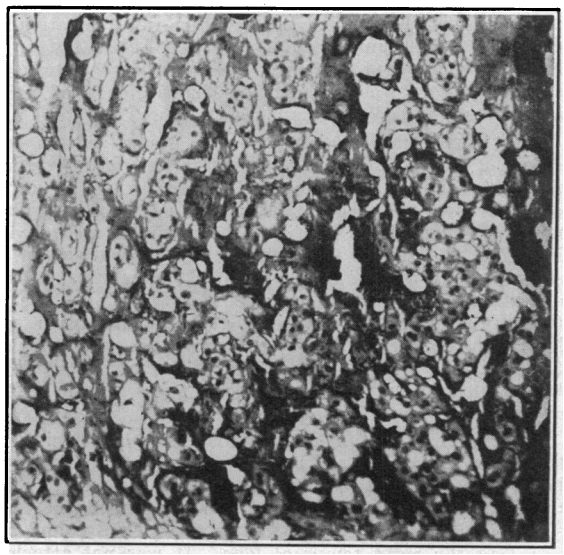

FIG. 2.—*Case 1.* Microscopic appearance of the tumour. Low magnification.

appeared to be small and compressed and separated by large amounts of mucin. Apart from the trabeculated bands of fibrous tissue, the stroma was scanty or absent. Blood vessels were found only in the above-mentioned trabeculated bands of fibrous tissue.

Sections of a small secondary growth in muscle presented the same picture. Considerable mucin was present.

CASE 2

M. W., a male, aged 35, farmer.

Family history.—Irrelevant.

Past illnesses.—He had had a fall on the head four years ago. This left him dazed, but not unconscious.

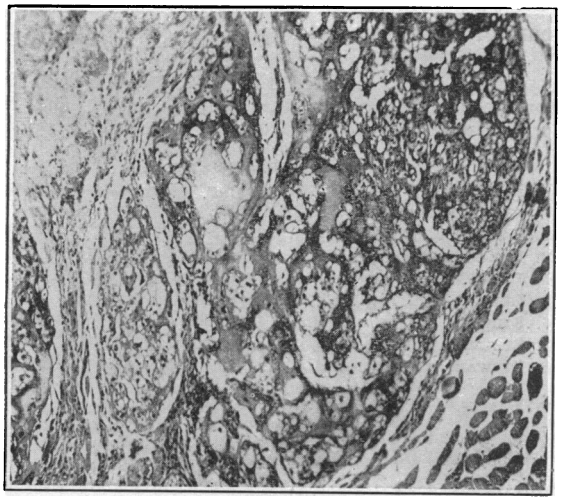

FIG. 3.—*Case 1.* Secondary nodule in muscle.

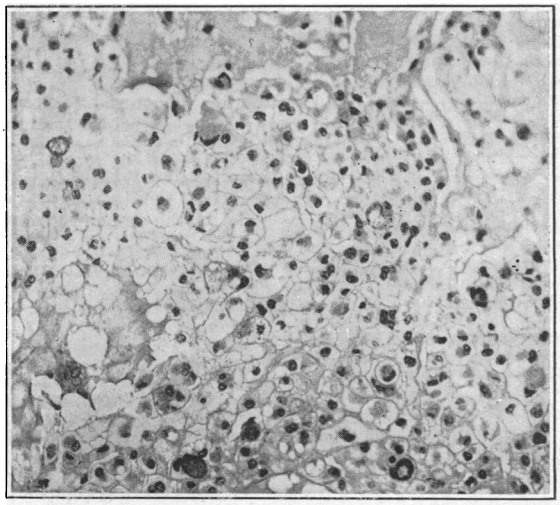

FIG. 4.—*Case 2.* Characteristic structure of the tumour.

He had no headache. Recovery was complete. His history was otherwise negative.

Present illness.—The patient was first seen on September, 1929. He complained of occasional vomiting attacks after meals. The general physical examination was negative. There was no loss of weight. He was mentally slow and memory for dates and places was poor. He complained later of pain in the upper jaw and a number of teeth were removed. In December, 1929, he showed paresis of the left side of the face and complained of headache in the right frontal region and pain in the upper jaw. The eye-grounds were negative at this time. In January, 1930, paresis of the left arm and leg was noted. Mental reactions were noticeably slowed. There was considerable incontinence of urine and fæces. The Wassermann reaction was negative.

At operation, a tumour was found within the cranium, having a deep attachment in the right temporo-frontal region. A portion of it, measuring 6 cm. in diameter, was removed. This was found to be irregular, nodular, encapsulated and in part cystic. It was greyish in colour with many purplish red areas. On section, it showed numerous engorged blood channels. After operation, the patient developed increasing weakness of the left side and paralysis of the left arm with epileptiform seizures. The pressure of the cerebro-spinal fluid was increased with a high cell count. He finally lapsed into coma and died about two months after operation.

At autopsy, a tumour was found firmly attached to the inner surface of the basilar process of the occipital bone. It measured 4.5 by 3.5 cm., and was soft, gelatinous and irregularly nodular. It extended into the sella turcica, compressing the pituitary body, and infiltrating the right temporal lobe. It was not attached to the dura. The optic nerves were stretched over the tumour and displaced to the left.

Microscopically, sections showed all gradations between comparatively small cells with solid pale pinkish cytoplasm and large typical physaliphorous cells, 100μ or more in diameter, with many vacuoles and a mere rim of cytoplasm. Many cells contained two or more nuclei. The nuclei were mostly large and pale, with a well marked nucleolus. Many showed indentation by the vacuolated cytoplasm. Several nuclei showing true nuclear vacuolation were noted. In some areas where an alveolar arrangement. Separating the cells were large amounts of stringy, hazy, bluish-staining mucin. There was very little stroma, but some fibrous bands ran through the tumour and in these lay thin-walled blood vessels. Through this fibrous connective tissue were also a number of lymphocytes and collections of large endothelial cells, containing granules of brownish pigment. Among the tumour cells, several rounded bodies vacuolation was less marked, the cells tended to assume were noted measuring up to 100μ in diameter. These were made up of large bluish granules with a few small round pinkish hyaline-like bodies scattered amongst them. They were probably degenerating cellular material.

Sections of the spheno-occipital bone from various areas at the point of attachment of the tumour failed to show any invasion of the bone by the tumour growth.

These two cases are typical both in their clinical course and pathological findings of chordoma occurring at the two most common sites,—the sacro-coccygeal and spheno-occipital regions. While the microscopic findings usually vary considerably in different areas, the composite picture should never leave the diagnosis in doubt. Stewart described the outstanding histological findings as:— (1) the alveolar character of the growth; (2) the solid epithelial aspect of the younger, more cellular, areas; (3) the cytoplasmic and intercellular vacuolation; (4) the formation of intracellular mucin which later escapes to lie between the cells.

Cappell[2] and Willis[9] described a hyaline capsule surrounding secondary growths. No evidence of such a structure could be made out in the small secondary nodule in the muscle in Case 1.

The cause of the vacuolization which is such a striking feature of these tumours has not been satisfactorily explained. Several authors claim to have demonstrated the presence of intracellular glycogen, and suggest that this is responsible, but others have been unable to confirm these findings. In both our cases the specimens had been fixed in formalin, which precluded staining for glycogen.

While it has been shown that chordoma probably arises from aberrant notochordal tissue yet what causes these cells to proliferate is unknown. In a great number of instances, and in both our cases, there is a history of antecedent trauma. It would seem reasonable that in some cases trauma might be the exciting factor.

Treatment of the condition is unsatisfactory. Practically all spheno-occipital cases are inoperable. Radiation therapy is ineffective. In the sacro-coccygeal type, the usual course is to excise the growth as completely as possible and deal with the invariable recurrences in the same manner.

I am much indebted to Dr. H. J. Hassard, of Portage la Prairie, for the clinical history and the surgical material obtained from the first case.

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