Chordoma in Siblings

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IN HUMAN embryological development[6] the evolution of the notochord can be demonstrated. In all higher vertebrates, the notochord arises in essentially the same manner from Hensen's node, which is a thickened mass of rapidly proliferating cells at the anterior end of the primitive streak. In the most primitive vertebrate groups it develops as a fibrocellular cord, directly ventral to the central nervous system and becomes the chief axial support of the body. In the elasmobranchs (shark family), ring-like cartilaginous vertebra are formed around the notochord and compress it. In the process of evolution, the notochord is further compressed when bony vertebrae replace the cartilaginous vertebrae of the lower forms of life, but, even in the higher mammals, the centra of the vertebrae remain to mark its location and the central portion of the nucleus pulposus of the intervertebral disc is, in its microscopic structure, clearly a notochordal remnant. Its remnants, found in both the centra of the vertebra and in the intervertebral discs, may develop as an invasive malignant tumor (chordoma) and thus become important pathologically.

Neoplasia of the chordal rest tissue is a relatively rare occurrence.[3] The commonest sites are sacrococcygeal and, less frequently, basisphenoid. Very rarely they may occur elsewhere in the spine. Intraspinal and extradural chordomas have been reported[7] and here they produce symptoms of spinal cord tumors. Ordinarily these tumors are extraspinal and have their origin in vertebral rather than disc chordal vestiges.[5] They grow slowly by expansion, ordinarily destroying the bone by an osteolytic process. Occasionally they may incorporate bone. They extend into the retroperitoneal space from their sacrococcygeal origin and into the retropharyngeal area from the basisphenoid nidus. They are more frequently in males than females, occur at any age, but usually in the fifth decade.[1] Their slow growth often occasions symptoms for as long as five years before diagnosis. As the tumor pushes its way into the retroperitoneal space from its sacrococcygeal origin, it often impinges on the bowel or causes bladder symptoms referable to the bladder. It is usually felt as a firm mass behind the rectum. Chordomas are nonencapsulated and what may appear as encapsulation does not truly limit the tumor. They spread by extension and infiltration of surrounding tissue. In a few instances it may be possible to excise them radically but usually complete removal proves impossible[1] and recurrence is the rule rather than the exception.

In gross appearance, a freshly removed chordoma is often much like a chondroma. It is usually pinkish-gray, firm and elastic, although some have a mucinous appearance and are soft in consistency. There may be areas of hemorrhage and necrosis.

Microscopically the tumor grows in cords and sheets of cells. The pattern closely resembles that of notochordal tissue as found in the nucleus pulposus of the intervertebral disc, and the various appearances of chordomas are parallel to the development of the primitive notochord.[2] Individual tumors and tumor areas vary from small cell structure with small nuclei to very large cells with prominent nuclei and nucleoli. These latter large cells predominate in the usual tumor and have a vacuolated or bubbly cytoplasm which is the origin of the term *physaliferous* (or bubble-bearing) cell. Cell boundaries are indistinct and blend with the pale-staining, homogenous mucoid matrix. These features may suggest a colloid carcinoma, and chordomas must be distinguished from them.

The characteristics of chordomas are as follows[4]: (a) formation of intra and extracellular mucus, (b) physaliferous cells, (c) lobular arrangement of cells. (d) vacuolization of nuclei, (e) resemblance to nucleus pulposus.

As previously noted, the tumors usually grow slowly; however, where the microscopic pattern presents the criteria of malignancy (such as bizarre nuclear patterns, hyperchromatism and mitoses). there is rapid growth, invasiveness and metastasis (as in Case 1 herein reported).

REPORTS OF CASES

CASE 1. Beginning about January, 1952, a 52-year-old white housewife noted low backache after falling off a stool onto the floor. This became progressively worse. It was not associated with any change in bowel habits, but the patient did have an

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8-pound decrease in body weight. She was initially treated by her family physician for arthritis.

In 1947, a hysterectomy and appendectomy had been performed for "fibroids." In 1948, she had had a hemorrhoidectomy.

The patient's mother was 70 and her father 77 years of age and both were living and well. A brother, who was then 46 years of age, was apparently well. A 35-year-old daughter was also healthy. There was no history of cancer, tuberculosis, heart disease or diabetes in the family.

Upon pelvic examination it was noted that a large mass displaced the vagina and rectum anteriorly; the uterus was absent. Rectal examination revealed a mass, approximately 8x5x4 cm., between the rectum and sacrococcyx. This was firm and bulged into the posterior rectal wall and seemed to be fixed to surrounding tissues. Barium enema and sigmoidoscopic examination also showed a large pelvic mass displacing the rectum anteriorly and to the right. In February of 1952, the radiologist reported no definite bony involvement of the lumbar or sacral spine except for increased lordosis.

At laparotomy in March, 1952, a reddish purple, vascular encapsulated pelvic mass about 10 cm. in diameter was observed tightly adherent to the sacrum and coccyx. The entire tumor was not completely removed because of extreme vascularity, bleeding and adherence to surrounding structures. The portion removed weighed 310 gm.

The pathologist reported "a rapidly growing and highly malignant chordoma."

In December of 1952, a partial resection of the sacrum and coccyx was done because of recurrent tumor, microscopically confirmed to be chordoma. On May 25, 1954, two years after the first operation, the patient was admitted for terminal care with evidence of a large abdominal and pelvic mass the size of a full term pregnancy. The left gluteal region was infiltrated with tumor. The diaphragm was displaced upward because of the large intra-abdominal mass. In addition, a mass was noted to be protruding into the rectal and vaginal walls. The patient died of uremia on May 26, 1954.

Pathologist's Report

The abdomen was protuberant and there were nodular protrusions along the lower abdominal scar. The arms, breasts and other tissue, which were not edematous, were wasted and cachectic.

There was no gross pathological change observed in the brain or chest except for pulmonary edema.

Upon section of the abdominal wall it was noted that the nodules in the lower abdominal region were within the wall and were distributed along the line of a previous surgical incision, apparently implants in the tissue. A large tumor, multinodular and massive, protruded from the pelvis, pushing the bladder forward and partially obstructing the bladder neck against the pelvis. The bladder extended in flattened broad fashion up to a point half way to the umbilicus as nearly as the adhesions present would permit. The intestines were thrust forward by neo-

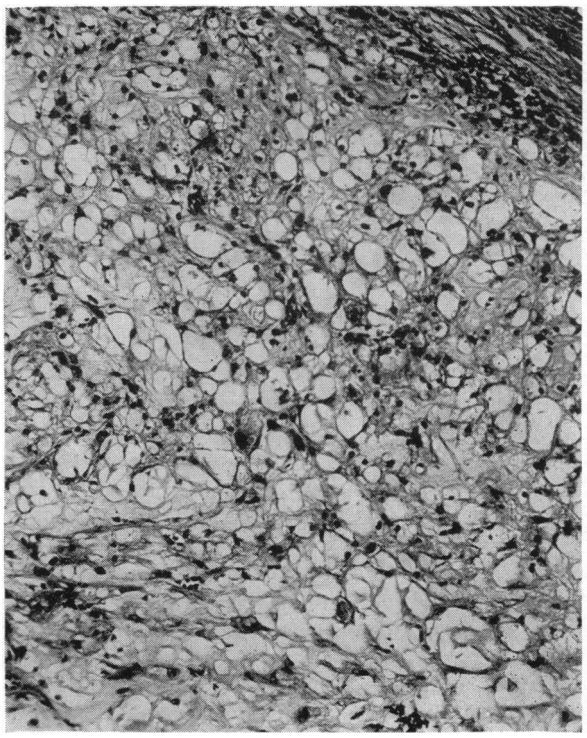

Figure 1.—Low power (×120) view of a field from tumor in Case 1. Note the vacuolation of cytoplasm and nuclei, and criteria of malignancy; that is, bizarre nuclear patterns, hyperchromatism and mitotic cells.

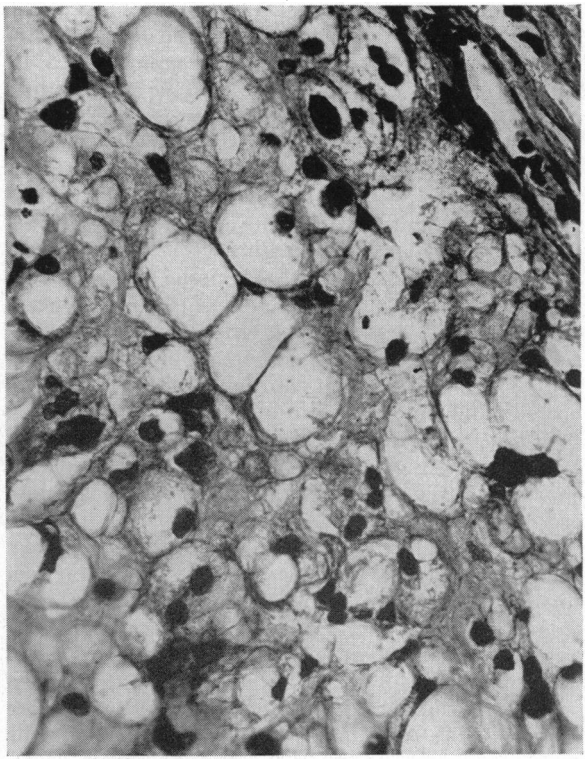

Figure 2.—High power (×500) view of a field from tumor in Case 1. The cellular and nuclear criteria of malignancy are more clearly seen.

plastic growth which extended retroperitoneally along the spinal column up to and beneath the liver. A large mass of neoplastic tissue protruded from the retroperitoneal space, pushing the right kidney upward. This mass was approximately 12 cm. in diameter. On section, the centers of most of these masses of neoplastic tissue were soft and hemorrhagic. Elsewhere, the tumor was not softened and hemorrhagic in appearance, and the cut surface was white or pearly white, occasionally mucoid or cartilaginous in appearance. The liver was filled with metastases of a similar character. Both ureters were obstructed by the outward and upward pressure of the retroperitoneal tumor masses. The ureters on both sides were greatly dilated, being fully 2 cm. in diameter, close to the pelvis of the kidney. Upon removal of the kidneys, pronounced hydronephrosis with renal atrophy was observed on both sides. The adrenal glands and the pancreas were grossly normal, and the gastrointestinal tract, exception for the compression previously described and the partial obstruction of the intestine both large and small, showed no gross abnormality. A tumor nodule about 2 cm. in diameter was observed at the hilus of the spleen but no tumor tissue was found on multiple sections of the splenic tissue.

Summary of gross pathological findings: Chordoma with sacral and pelvic invasion and body destruction, liver metastasis, retroperitoneal extension of chordoma causing partial intestinal obstruction, partial bladder neck obstruction, partial ureteral obstruction with chronic hydronephrosis, and edema—pulmonary, fascial and dependent.

CASE 2. A 52-year-old white man, a tiling contractor, brother of the patient in Case 1, said that he had "low back bone pain," which he first noted in December, 1954, as a dull aching pain which he also felt made his bladder empty poorly. These symptoms disappeared completely until July, 1955, when throbbing pain recurred, again localized about an inch or two above the end of the sacrum, starting on the left; and again it was associated with difficulty in urination. The pain kept him from sleeping and it became progressively worse in the course of about a week. Earlier in the year 1955 the patient had noted a significant change in bowel habits—pronounced constipation necessitating daily enemas. There were no other urinary symptoms except for nocturia one time. Libido had been decreased since 1953 but there had been no significant change in sexual activity. It was noted that the pain, which originally began on the left, became more pronounced on the right side and became bilateral.

Upon examination the patient was observed to be well developed and well nourished, and to have no definite physical abnormalities of significance except a firm, smooth, regular, symmetrical 5 to 6 cm. rounded mass firmly attached to the sacrum at its lower end, which was palpated at the end of the examining finger on rectal examination. The mass was tender and nonmovable. The coccyx itself did not seem to be involved and was movable.

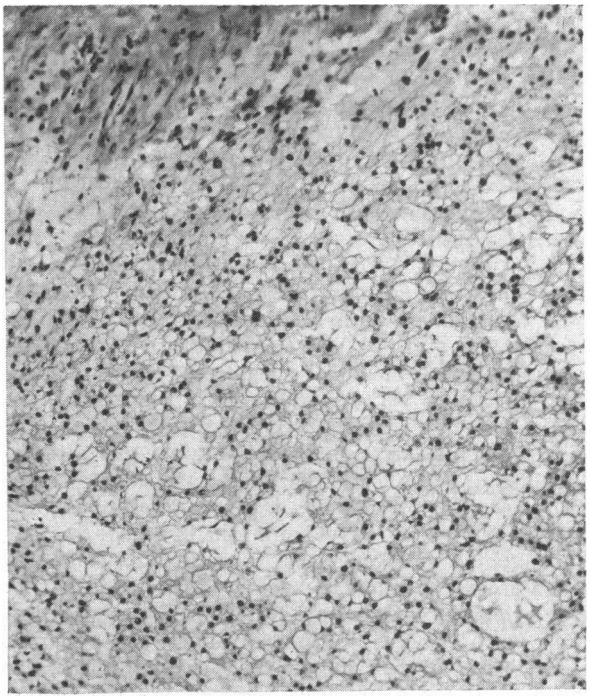

Figure 3.—Low power (×120) view of a field from tumor in Case 2. Note the smaller cells, greater uniformity of size and shape of nuclei than seen in Figures 1 and 2, and the vacuolation of cytoplasm and ground substance.

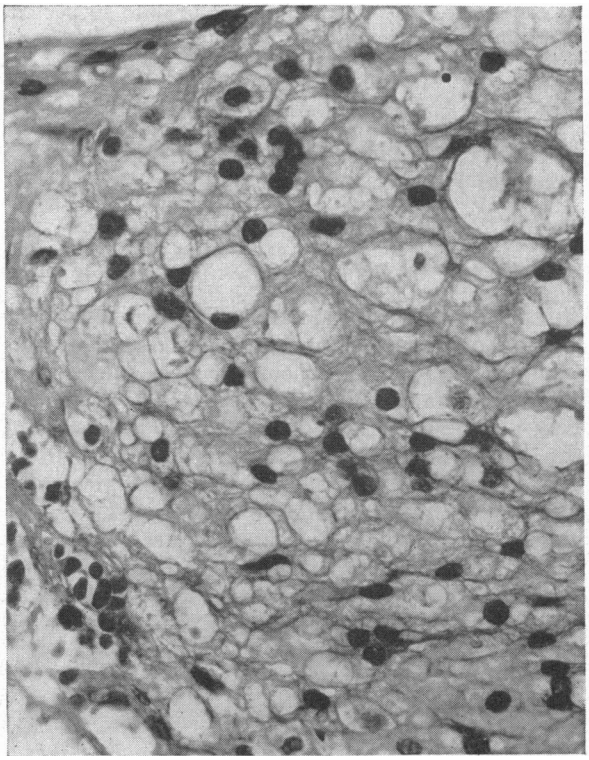

Figure 4.—High power (×500) view of a field from Figure 3. The nuclear characteristics (small size and uniformity in size and shape) as well as the absence of mitotic cells are more clearly seen.

Upon neurological consultation and careful testing, including the saddle areas, no motor changes or reflex changes were observed. Especially detailed sensory evaluation with light touch, pinpoint and vibratory stimuli did not demonstrate deficits.

It was felt that the mass arose from the sacrum and encroached on the posterior rectal wall without actually involving it. Surgical intervention was advised.

On July 29, 1955, with the patient in a prone position, coccygectomy was performed. A tumor was immediately seen at the lowest end of the remaining sacrum. It was bluish gray, had a thin capsule and seemed to be relatively soft and gelatinous. It lay between the ventral surface of the sacrum, which it had partially eroded, and behind the presacral fascia and probably the periosteum. A portion of the lower end of the sacrum was carefully removed. The tumor was not noted to involve any of the sacral nerves nor did it invade into or through the anterior sacral foramina. It seemed to have eroded the ventral surface of the sacrum, the bone being somewhat excavated in this region and softened around it, and had a rather wide adherent base at the level of the fourth sacral foramina. All portions of the tumor were removed and the ventral surface of the sacrum carefully curetted of all possible gross remnants. The tumor was 5x5x3 cm. The patient recovered readily. When last observed, some two years after the operation, he had stopped working because of intractable pain "at the lower end of my spine." Recurrence of tumor was suspected.

SUMMARY

Two cases of chordoma, in siblings, a man and a woman of middle age, are reported. This is the first reported occurrence in siblings. In the woman the tumor recurred locally and spread to the liver. In the man the tumor was in the sacrococcygeal area. It was removed and the patient was well for two years, but at the time of this report there were symptoms of recurrence. In a review of the literature no reports or suggestions were found with regard to a hereditary or genetic basis for the occurrence of chordoma.

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Meckel's Diverticulum Incarcerated in a Femoral Hernia

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THE DIAGNOSIS of strangulation of a herniated diverticulum of the intestine that lies within a greater hernia is usually very difficult to make. The classical signs of strangulation—inflammation, fever and pain—are less severe and occur late in the course of the disease process. Often there is no intestinal obstruction, the fecal stream passing readily since only a diverticular process, not the entire bowel, is incarcerated. The only positive findings are a mass and perhaps tenderness if there is compromised bowel in the sac. Fecal fistula at the site of the hernia was noted by Weinstein[7] in 25 per cent of cases he reviewed in a report published in 1938.

HISTORY

In 1700 Littre[4] published a description of a new form of hernia observed in three cases. The hernial sac described contained a Meckel's diverticulum. This combination now is called Littre's hernia. The process is to be contrasted to a Richter's hernia in which a portion of the bowel wall is strangulated in a hernial sac but no preformed diverticula is involved.[3] The older literature does not differentiate clearly between the two entities, mixed terminology making it difficult now to determine how many cases there were of each.[2] In 1946, Watson collected reports of 211 cases of Littre's hernia from the literature; 30 of them were of the femoral type.

In 1808 Meckel[5] described the ileal diverticulum which bears his name and postulated it was of congenital origin. Meckel's diverticulum is reported to occur in from 0.2 to 2.0 per cent of persons.[1,6] The incidence noted at postmortem is slightly higher than that observed at laparotomy, since the search usually is less intensive in live subjects. Meckel's diverticuli may vary considerably as to size, length and location, but usually the sac projects from the antimesenteric border of the ileum at a point anywhere from 12 to 36 inches proximal to the ileocecal valve. It may be a fibrous tract from the ileum to the umbilicus or it may be patent, and it may or may not have its own mesentery. Meckel's diverticulum may contain ectopic gastric, duodenal or pancreatic tissue

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