SACRAL CHORDOMA

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In the present paper, the writers offer another authentic case of chordoma to add to the rather meagre number now reported. A review of the literature has revealed accounts of only slightly more than eighty cases of all varieties. Most of these have been situated, as was this one, in the sacral area, the next most frequent site being the spheno-occipital region, although in the past few years some cases occurring along the spine at various levels have been reported. Probably, chordoma is not so rare as the number of reported cases indicates, many such tumors being either overlooked or incorrectly diagnosed.

History of the Case. This patient was a man aged forty-one, a manufacturer, who came to the clinic September 10, 1929, complaining of pain in the lower part of his back. This pain began in December, 1928. It was very mild at first, but increased slowly in intensity. It was of a constant boring character not affected by activity, and interfered with the patient's sleep. Heat, aspirin, and periods of rest had failed to give relief and after months of annoying and unrelievable pain in the lower spine, occasionally radiating down the left leg, a consultation was sought.

The findings on physical, laboratory, and x-ray examinations were negative, except for a tender area the size of a fifty-cent piece over the lower third of the sacrum exactly in the mid-line. This area could be definitely delimited, and neither pressure over the surrounding parts nor manipulation of the lumbosacral or sacro-iliac joints produced any pain. Rectal examination revealed a bulging area on the anterior surface of the sacrum in its lower third which was tender and semifluctuant. There was no fixation of the soft tissues, and the coccyx was free and movable and not painful.

Exploration, September 14, 1929, revealed a tumor mass, yellowish, soft, and very friable, which protruded from the posterior surface of the sacrum and extended through to its anterior surface, the area of bone erosion being about 2 centimetres in diameter. A portion of the tissue was removed for pathological study, and frozen sections were made at this time but were not sufficiently clear to permit of a diagnosis. The tumor was curetted out as thoroughly as possible, its bed was packed with vaseline gauze, and the wound was closed.

Our pathologist, Dr. Allen Graham, reported that macroscopically, the specimen consisted of numerous grayish-pink pieces of

tissue, soft, friable, and of a somewhat gelatinous consistency. Microscopically, the appearance of the tumor tissue was variable. There were areas in which small and large solid nests, strands, and masses of tumor cells were lying in a homogeneous pink-staining mucoid stroma. The tumor cells varied considerably in size and shape. In general, they were made up of a large amount of homogeneous, pink-staining cytoplasm, containing a relatively small,

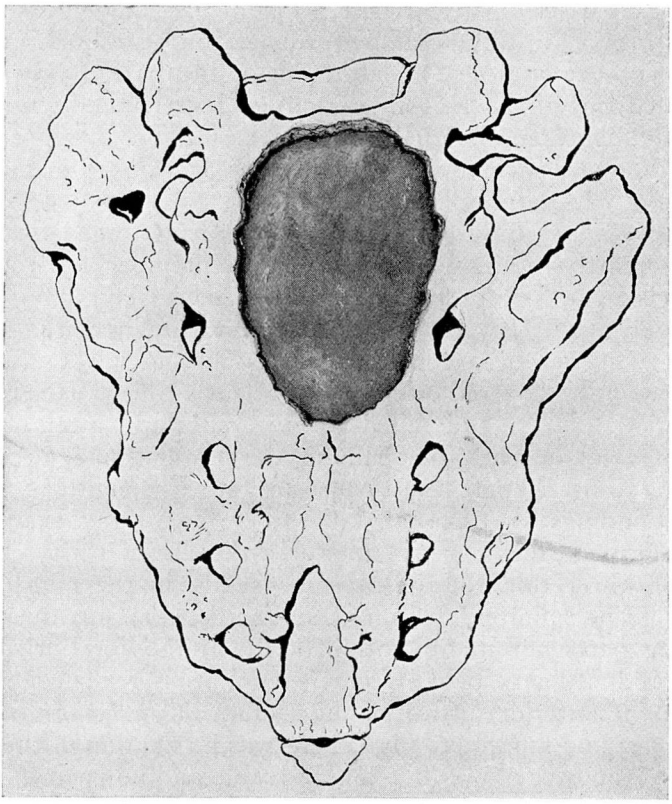

Fig. 1. Drawing of sacrum showing the relative size and position of the chordoma at the time of the second operation.

round, spindle-shaped, or irregular nucleus. The nuclei were vesicular, and had well-defined nucleoli. The cell outlines were not distinct. There were large masses of multinucleated cytoplasm which had the appearance of syncytial tissue. The cytoplasm was vacuolated in many instances. In some areas the tissue was made up of compact masses of spindle cells, forming fibrillae and whorls, little of the mucoid stroma being present, and the picture was not unlike that

seen in a fibroma or fibrosacroma. The nuclei of the cells were variable in size and shape, and a few mitotic figures and irregular nuclear divisions were observed. In other areas there was a mixture of the two histological types described above. In a few areas the arrangement of the tissue was somewhat suggestive of cartilage.

The pathological diagnosis was *sacral chordoma.*

September 19, 1929, the sacrum was radiated with 900 r-units. The patient was discharged from the hospital September 22, 1929. He was free from pain, but there was a slight seropurulent discharge from the wound. Subsequently he reported regularly for dressings and observation until March 10, 1930. Although there had not been any return of discomfort nor any external evidence of a recurrence of the tumor up to this time, on rectal examination a mass was palpable which seemed to be slowly increasing in size. Another attempt at complete excision was therefore advised.

The second operation was performed March 27, 1930. Preoperative rectal examination revealed a sessile mass in the hollow of the sacrum, about the size of a silver dollar and approximately 8 millimetres thick. The centre of this mass felt softer than the surrounding portion.

The old scar over the sacrum was excised. Considerable scar tissue was found under the skin. The spines of the sacrum were exposed with the periosteal elevator, and the ligamentous structures were reflected. There was a small opening into the sacral canal of about 1½ by 3 centimetres. Through this could be seen a soft mass, bluish-gray in color. The roof of the sacrum was cut away by rongeurs, leaving an opening 5 by 10 centimetres in size. The whole of the sacral canal was filled with a fairly firm, bluish-gray tumor mass, which was remarkably avascular except at the periphery (Fig. 1). In the lateral portion of the mass, bundles of tissue could be seen which were identified as the sacral nerves. The tumor apparently filled the entire sacral canal, and was intimately associated with the sacral nerves. It was deemed inadvisable, therefore, to attempt to remove it because of the probability that these nerves would be injured. A rectal examination was made at this juncture, and pressure was applied to the anterior aspect of the tumor in the hollow of the sacrum. This mass could be seen and felt to bulge slightly just to the left of the mid-line at about the mid-portion of the sacrum. The patient made a satisfactory operative recovery and was discharged from the hospital April 6, 1930.

Post-operative treatment consisted of deep x-ray therapy in doses of 160 r-units on April 26, May 3, May 10, May 24, and

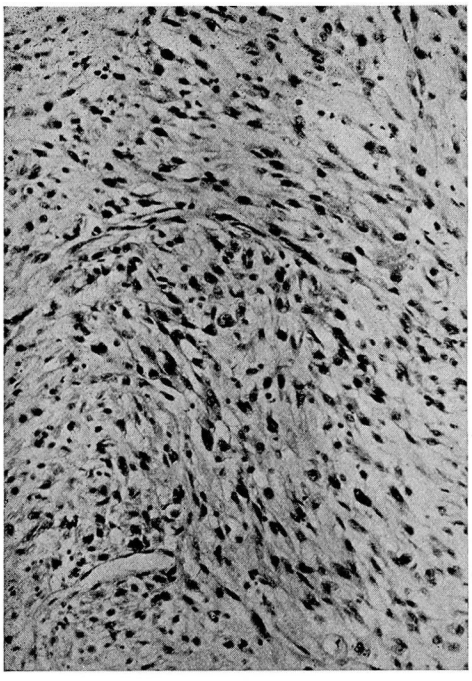

Fig. 2. View of a section, X 150, showing the spindle-cell type of tissue.

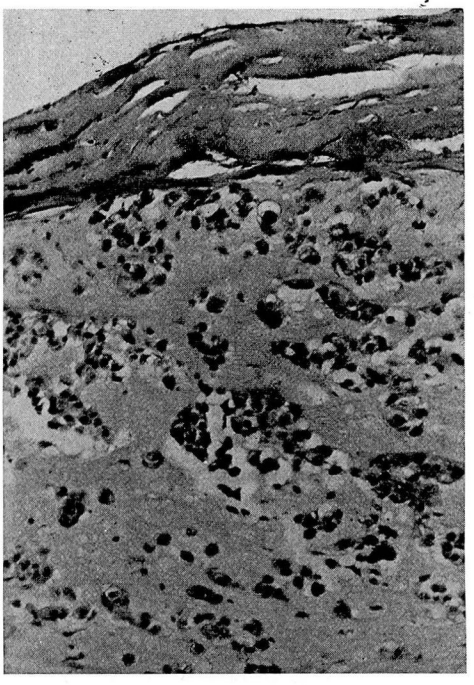

Fig. 3. View of a section, X 150, showing the fibrous capsule of the tumor and the cartilage-like tissue with mucinous stroma.

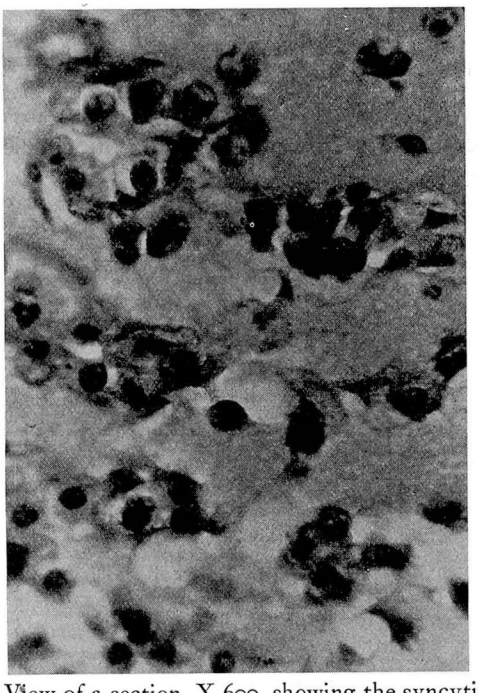

Fig. 4. View of a section, X 600, showing the syncytial-like masses of cells with vacuolated cytoplasm.

June 10, 1930. During this time the tumor progressively decreased in size and gradually became harder and more calcified.

When last examined, September 9, 1930, the patient had continued free from pain, and rectal examination revealed that the tumor was definitely smaller, while all tenderness had disappeared. The x-ray treatments apparently had been successful in checking the development of the neoplasm. Metastasis is rare in this type of tumor, and none has been found in this case.

Chordoma is a tumor arising from cellular remains of the notochord, occurring, therefore, along the spine, most frequently at its extremities. It is composed of epithelial tissue, and is of endothelial origin.

As far back as 1856 Luschka described a case of chordoma, but did not recognize its origin or importance. Müller in 1858 suggested that the notochord was perhaps the origin of these tumors. The name "chordoma" was suggested by Ribbert in 1894. The development of our present knowledge has occurred almost entirely during the past thirty years, more particularly since 1922, when Professor Matthew J. Stewart, of Leeds, presented the first case recognized in Britain. In 1926 Professor Stewart collected fifty-seven reported cases, and in 1929 reports of only eighty cases had appeared in all the medical literature, and even some of these are questionable.

The average age at the onset of these tumors is from thirty-five to forty years, although cases have occurred as early as one and a half and as late as seventy-nine years. Spheno-occipital chordomas appear, on the average, ten years later than sacrococcygeal chordomas.

Males are twice as prone as females to develop these tumors, which suggests the part that trauma may play in their etiology. As a matter of fact they have been produced experimentally in rabbits by puncturing the body of a vertebra.

The first symptom noticed usually is mild pain in the sacrum or lower portion of the spine, located exactly in the mid-line. Pain may radiate down the legs or into any region upon the nerve supply of which the growth encroaches. Relief cannot be obtained by the ordinary therapeutic measures, and the pain gradually increases until sleep becomes almost impossible. While the tumor may be discovered before the occurrence of pain, more often it is found as a result of the pain. The mass may protrude principally within the bony pelvis, and thus escape detection unless the sacrum is palpated by rectum. Chordomas grow very slowly, but their persistence has been regarded as certain.

Usually there are no symptoms except those caused by mechanical pressure. The diagnosis is suggested by the history of pain in the lower spine or skull and by the finding of a palpable tumor mass, semifluctuant in character. A positive diagnosis, however, can be made only by the microscopic appearance.

In 1926, Stewart and Morin described the gross appearance of these tumors in detail. The growth is well encapsulated, rounded, and lobulated. Gross section appears lobulated, and the lobules show mucoid degeneration, often of an advanced character. Frequently, cells of syncytial type are embedded in a sea of mucin. Some areas resemble colloid carcinoma, others cellular carcinoma. The salient microscopic features described by Stewart are as follows: aveolar character of growth; solid epithelial aspect of the younger cellular areas; cytoplasmic and intercellular vacuolation; formation intracellularly of mucinous fluid, which escapes from the cells to form, first, intercellular columns and, later, mucin in which only scattered cellular islets remain; rarity of mitotic figures except in very malignant cases. Chordomas are malignant only in a low degree, but occasionally they metastasize.

Although in the great majority of cases reported so far radiation has not helped, from the results in this case it is our opinion that the x-ray, in sufficient dosage, has greater possibilities than surgery. The end-result of our case, of course, is uncertain, but the improvement thus far has been satisfactory. The size of the tumor has decreased, the tissues have hardened, and all symptoms have disappeared.

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