CHORDOMA OF THE THORACIC SPINE
Report of a Case

F. Husain, London, England

Chordoma is thought to arise from remnants of the notochord. This is the embryonic axial skeleton of vertebrates which becomes surrounded and submerged by the mesenchymal elements forming the spinal column and base of the skull. Vestigial remnants may be present anywhere along the cranio-spinal axis—in the basi-sphenoid, odontoid process, vertebral bodies and nuclei of the intervertebral discs. Chordoma is a rare tumour. Of low-grade malignancy (Meaney, Greenwald and Phelan 1956), it grows slowly and invades locally but seldom metastasises. Luschka (1856) and Virchow (1857) described the condition under the name of ecchondrosis physaliphora. Müller (1858) was the first to recognise its origin from notochordal cells, and Ribbert (1894) first introduced the term chordoma. Coenen (1925) classified the tumours as 1) cranial—clival, hypophysial, nasopharyngeal and dental; 2) vertebral—cervical, thoracic and lumbar; 3) caudal—antesacral, retrosacral and central. Faust, Gilmore and Mudgett (1944) and other workers studied the incidence of the tumour according to age, sex and site of affection. Chordoma may occur in patients aged from seven months to 82 years (Allen 1955) but is commonest in the sixth decade (Dahlin and McCarty 1952). The average age of affection is fifty-three years for caudal and thirty-five years for vertebral chordomata. Males are more frequently affected than females in the case of caudal and vertebral chordomata (4 : 1 and
2:1), whereas both sexes seem to be equally affected in the case of the cranial tumours (Utne and Pugh 1955). Caudal chordomata account for from 50 to 60 per cent of all cases, and cranial growths for 37 per cent: truly vertebral chordomata are rare (13 per cent). In the last group, cervical involvement is the most common and lumbar affection is the next in frequency. Occurrence in the thoracic region is extremely uncommon.

**CASE REPORT**

A woman aged fifty-eight complained of pain in the mid-thoracic region of the spine, with swelling in that region. She related the onset of symptoms to a strain of the back sustained when a bus in which she was standing moved suddenly. Pain was increased when she coughed or sneezed, and had slowly increased since its first onset. There were no other related symptoms or any relevant features in the past or family history. She was admitted for investigation in September 1957, one year after the onset of symptoms.

At that time examination showed tenderness in the mid-thoracic region posteriorly, rather to the left of the mid-line. At that site there was a fluctuant swelling four inches in

![Image of a tumour after removal](image)

**Fig. 3**
The tumour after removal.

length and breadth, not adherent to the skin. There was no sign of affection of the spinal cord or nerve roots, and examination of other systems showed no abnormality.

*Investigations*—Radiographs of the spine showed a soft-tissue mass in the mid-thoracic region with scoliosis to the right at this level. The outline of the left pedicle of the seventh thoracic vertebra was indistinct (Fig. 1). There was no evidence of affection of the vertebral bodies, and radiographs of the rest of the spine and of the chest showed no abnormality. Examination of the blood showed: haemoglobin 88 per cent; white cells 8,000 per cubic millimetre. Erythrocyte sedimentation rate was 13 millimetres in the first hour. Examination of the urine showed no abnormality. Aspiration of the swelling produced only a small amount of bloodstained fluid which was sterile on culture.

One week after admission biopsy was performed. Sections of the specimen showed the appearances of chordoma.

*Treatment and progress*—Three weeks after admission operative removal of the tumour was attempted. A large mass of tumour was found extending across and on both sides of the mid-line from the fourth to the eighth spinous processes. It invaded the paravertebral muscles and the trapezius and rhomboids. It seemed to arise from the seventh thoracic vertebra, whose laminae and left pedicle were considerably eroded. There was no involvement of the dura mater. The tumour was soft, gelatinous and greyish. Tumour, muscle, spinous processes and
Figure 4—Fibrous stroma forming capsule and septa. (×35.)

Figure 5—Small separate collapsed tumour cells. Much mucoid tissue. (×112.)

Figure 6—"Signet-ring" cells. (×465.)

Figure 7—Physaliphorous cells. (×475.)
laminae were excised, but the removal of the tumour was necessarily incomplete because of its anterior extension on both sides of the vertebral column. The patient's progress after this operation was uneventful, and she left hospital after five weeks. Radiograph at that time (Fig. 2) showed more clearly the destruction of the pedicle of the seventh thoracic vertebra.

**Examination of tumour**—The gelatinous mass removed was $14 \times 10 \times 7$ centimetres in size (Fig. 3). On section it appeared lobulated, with a thick fibrous stroma forming a "capsule" and septa (Fig. 4). There was much mucoid tissue. The tumour cells were mostly small and separate with a collapsed appearance (Fig. 5), but in some areas "signet ring" cells (Fig. 6) or physaliphorous cells (Fig. 7) predominated. In other areas the cells tended to form epithelial cords (Fig. 8). The stroma contained scattered collections of lymphocytes and other leucocytes and there were occasional granulomatous areas with giant cells (Fig. 9).

**Subsequent progress**—The patient was well until July 1958 when pain recurred. There was a swelling over the right seventh rib. Radiographs showed no evidence of further destruction of bone. Shortly after the recurrence of pain the patient developed paraplegia which rapidly became almost complete. She was admitted to University College Hospital where attempts at relief were made by operation and by radiotherapy. At the second laminectomy some recurrent tumour was found, without evidence of compression of the cord. However, the apparent softening of the cord itself seemed to indicate that thrombosis of the vessels of the cord had occurred. The paraplegia was unaffected by treatment. The patient developed urinary infection and bedsores and died of toxaemia in March 1959, eight months after the recurrence of pain.

**DISCUSSION**

There are several specially interesting features about this case. The tumour itself is a very rare one, and its occurrence in the thoracic spine is most unusual. Its presentation here was simply with pain in the back and with local swelling, without any considerable radiographic changes. The absence of destruction of the vertebral body is an unusual feature, though the
presence of a swelling is the rule in these cases. The possible diagnoses of infection such as tuberculosis, of a primary condition such as myeloma or haemangioma, and of secondary malignant affection had to be considered (Sennett 1953). The diagnosis was in fact made on the histology of the mass as shown by biopsy. The histological appearances were characteristic of chordoma, though the behaviour of the tumour was perhaps rather more malignant than was suggested by the large amount of mucoid tissue present. Recurrence of symptoms occurred after incomplete removal, and the subsequent behaviour of the tumour confirmed the view that chordoma is not radiosensitive. Horwitz (1941) showed by examination of human embryos that notochordal remnants were most often present in those sites favoured by chordomata. The question of the role of trauma in precipitating neoplastic metaplasia in these notochordal rests has been raised, and certainly in this case symptoms appeared after injury.

SUMMARY

1. A case of chordoma of the thoracic spine is described.
2. Certain features of this case—namely the absence of vertebral destruction—are contrasted with those of cases previously described.

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REFERENCES