SECONDARY CHORDOMA OF THE MANDIBLE

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Summary. The first case of primary sacrococcygeal chordoma metastasising to the mandible and presenting with obstructed labour is described. The pathological features of this tumour, its clinical presentation and management are discussed.

Introduction

Chordoma is a rare, malignant tumour which develops from the embryonic remnants of the notochord, but clinically becomes manifested during adulthood. To the best of our knowledge its coincidental association with pregnancy has never been recorded. The tumour usually spreads by local invasion; distant metastasis, particularly to bone, is rare. We report the first case of sacrococcygeal chordoma presenting with obstructed labour and metastasis to the mandible.

Fig. 1

Figure 1—Photomicrograph of chordoma showing sheets of vacuolated and clear signet ring cells surrounded by haemorrhagic stroma. The marked field (arrow) is magnified in Figure 2 (H & E ×100).

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Case report

A 32-year-old Pakistani fourth gravida was admitted in July 1983 to hospital in Bahrain with obstructed labour due to a pelvic mass involving the posterior rectal wall and the posterior vaginal wall. She delivered a healthy, male child by Caesarian section. After closing the uterus, the abdomen was explored and a retroperitoneal, firm, gelatinous mass measuring 10×10 cm was seen in the sacral hollow, just below the promontory. Two weeks later, she underwent exploratory laparotomy but the mass could not be mobilised due to its firm attachment to the pelvis and the posterior rectal wall. There were no enlarged lymph nodes. A biopsy taken from the mass showed the appearances of a primary chordoma (Figs. 1 & 2).

In 1984, she returned to Pakistan for radiotherapy and excision of the remaining parts of the tumour. No enlarged lymph nodes were noticed at the time of the surgery. In mid 1985, she returned to Bahrain complaining of pain radiating to the lower limbs and radiographic examination showed destruction of the right side of the sacrum. Sigmoidoscopy revealed a soft and tender bulge 5 cm from the anal verge. She was recommended to have further surgery in Pakistan and this was performed 4 months later.

In January 1986, she was readmitted in Bahrain with abdominal pain, fever, vomiting, hepatomegaly and jaundice. Blood analysis was positive for hepatitis B antigen and ultrasonography showed multiple gall stones. She was diagnosed as having infective hepatitis from previous blood transfusion, complicated by the gall stones. She also developed a urinary tract infection due to *Escherichia coli*.

In July 1986, she complained of severe pain in the rectum and buttocks but examination showed no signs of inflammation, ascites, organomegaly or any neurological deficit. Five weeks later, she was referred to the Department of Dentistry with pain and swelling at the right angle of the mandible. Radiographic examination showed an area of decreased density in the posterior body of the mandible.

![Figure 2](image-url)

Figure 2—Photomicrograph showing physaliphorous cell in a vascular stroma characteristic of chordoma. (H & E×450).
adjacent to the roots of the last right molar tooth. The diagnostic possibilities were dental cyst, ameloblastoma, giant cell tumour of bone, myeloma, histiocytosis X or metastatic tumour. The mandible was explored using an intra-oral approach under endotracheal anaesthesia. The buccal and lingual bone were found to be expanded and eroded, together with the lower border. The interior of the mandible contained purple, friable tissue and biopsy samples taken from the bone and soft tissue showed metastatic deposits similar to that of the pelvic tumour. It was decided at this point to re-investigate the primary site of the tumour. Sigmoidoscopy showed a swelling in the sacral hollow, 15 to 17 cms from the anal verge, and CT scan showed extensive, mixed density soft tissue involving the right half of the first five sacral vertebrae. The bony component was expanded and extended posteriorly while the soft part extended from around the right lateral side of the rectum to the posterior bladder wall and showed patchy but well marked enhancement suggestive of increased vascularity and recurrence of the previously excised tumour. The patient was informed about the local recurrence of the tumour and its distant metastasis in the mandible and was advised to seek further reassessment in Pakistan.

Discussion

Chordoma is a rare, slow growing, malignant tumour which develops from ectopic, embryonic remnants of the notochord along the spinal axis. It occurs with a male:female ratio of 2:1 most frequently in the sacrococcygeal region followed by the sphen-o-occipital area, and usually arises between the fourth and seventh decade of life, although no age is exempt (Ariel & Verdu, 1975; Paevolainen & Teppo, 1976).

The presenting clinical features of chordoma depends on its location (Paevolainen & Teppo, 1976; O’Neil et al., 1985). Thus sacrococcygeal tumours give rise to low back pain and bowel and urinary disturbances, whereas sphen-occipital tumours invariably present with features of raised intracranial pressure associated with cranial nerve palsies particularly those of the oculomotor and abducent nerves giving rise to diplopia. The average length of life from the onset of symptoms until death has been estimated at 5.7 years (Gray et al., 1975). As the tumour spreads to the nearby organs the function of these structures becomes affected. Thus in the present report there was obstructed labour due to the extensive pelvic spread and involvement of the posterior vaginal wall. The association of pregnancy in this case is a coincidental finding.

Chordoma occur in the form of lobulated solid or cystic, grayish brown masses with areas of gelatinous, mucinous or translucent appearance. The lobules are usually contained in a fibrous capsule but at the time of diagnosis there is frequent local invasion to adjacent parts, particularly bone. Distant metastases are rare but can occur via the bloodstream as well as the lymphatics thus combining features of carcinoma and sarcoma. The tumour has been described to spread to the regional lymph nodes, skin, adrenal glands, bone, liver and soft tissue of the thorax (Mabrey, 1935; Yarom & Horn, 1970; Paevolainen & Teppo, 1976; Chambers & Schwinn, 1979). In this report the metastatic spread of chordoma reached the mandible.

There are no pathognomonic histological features of chordoma but the tumour is characteristically pleomorphic with a wide range of structural variations in any one tumour. It is arranged in the form of lobules or alveoli with the tendency to form cords and syncytial masses of large, clear, vacuolated, mucinous, signet ring cells
giving rise to 'physaliphorous' or bubble cells. Using immunocytochemical techniques the tumour can be differentiated from atypical chondroma, chondrosarcoma, liposarcoma, ependymoma and primary or secondary metastatic clear cell carcinoma (Abenoza & Sibley, 1986; Finlay et al., 1986) and the diagnosis can be further confirmed using electron microscopy (Finlay et al., 1986).

Excision is the only treatment for chordoma and repeated surgical intervention may be necessary since local recurrence of the tumour is very common (Paevolainen & Teppo, 1976). Radiotherapy has been also recommended although the results are considered palliative rather than curative (Cummings et al., 1982). For example, a dose of radiation above 5000 cGy has been found to cause partial regression of the tumour and to increase the 5-year survival rate (Pearlman & Friedman 1970; Reddy et al., 1981). Chemotherapy has no place either in the treatment of the primary tumour or its metastatic deposits (Cummings et al., 1982).

The mandibular deposit of chordoma in the present report may be managed by excision of the involved bone and soft tissue followed by a reconstructive graft from the iliac crest. Where overlying skin and soft tissue are involved then reconstruction with a myo-osseous flap may be used, for example with a portion of the radius. Local radiotherapy to the jaw can also be used in such an accessible site or alternatively as a post-operative measure (Chambers & Schwinn 1979). At the same time, the primary sacrococcygeal tumour would require further management in view of the local recurrence of large tumour invading the pelvic organs as shown on the CT scan. The management of the mandibular deposit in this case is appropriate despite the poor overall prognosis to preserve a reasonable quality of life during the survival of the patient.

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References