SACRAL CHORDOMA

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Introduction

Chordomas are rare tumours, accounting for 1-2% of all malignancies and are thought to originate from the primitive notochord. Approximately 50% originate in the sacral region, 35% in the clivus (base of skull), and 15% elsewhere along the axial skeleton. Skull tumours are relatively small but extremely difficult to remove due to their proximity to the brain. Conversely, the sacrococcygeal tumours present as large masses and are thus difficult to resect fully. The tumours are slow growing, with a high propensity for local recurrence regardless of treatment; distant metastases are rare [1]. Wide excision with postoperative radiation for positive margins is the treatment of choice.

We present a case of sacral chordoma which was treated successfully by wide surgical excision.

Case Report

A 65-years-old lady presented with a painful swelling over the lower back of 3 months duration followed by constipation and acute retention of urine of 15 days duration. Clinical examination revealed a sacral mass bulging into but free from the rectum. X-ray of the sacral spine revealed a soft tissue mass anterior to the sacrum with osteolytic lesions in S2-S5 segments. CT-Scan of the pelvis revealed a presacral mass with calcifications within, compressing the rectum and the bladder anteriorly (Fig-1). The mass was found to be infiltrating the right paraspinal muscles. FNAC was suggestive of chordoma of the sacrum.

The patient was taken up for surgery. A combined abdominal sacral approach was employed. Through a lower midline laparotomy first, the rectum was mobilized off the tumour. Thereafter through a midline sacral incision the tumour along with the sacrum and coccyx below the level of S2 vertebra was excised along with the involved part of the right paraspinal muscles in toto. The muscular gap was bridged by a prolene mesh, the wounds primarily closed. Post-operative recovery was rapid, the patient regaining bladder and bowel control by the fifth day. Histopathology revealed physaliphorous cells characteristic of a chordoma (Fig 2).

The patient received radiotherapy, 6000 cGy over 5 weeks, and is on regular follow up with no recurrence at 10 months post op.

Discussion

Sacral chordoma is a low grade malignant tumour but is locally aggressive [2]. A median interval of one

Fig. 1 : CECT pelvis showing a large presacral mass with calcification

Fig. 2 : Photomicrograph of tissue section showing typical physaliphorous cells (H & E 200X)

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year is usual between the onset of symptoms and diagnosis. Pain is the chief symptom, is non specific and comes gradually as the surrounding soft tissues are displaced and the bone eroded. The tumour usually expands ventrally, displacing the presacral fascia anteriorly without producing an obvious mass externally. The rectum is rarely invaded. As the tumour encroaches on the sacral foramina, nerve root neuropathies develop, leading to urinary retention, rectal sphincter incontinence and radicular pain.

The consistent radiographic finding in sacral chordomas is the destruction of several segments of the sacrum with an associated mass placed anteriorly. With the advent of CT, better localization of the tumour has become possible. Currently, MRI is replacing CT as the imaging modality of choice because it not only offers a higher resolution image but also allows evaluation of the sacral canal which acts as an unobstructed conduit for the superior spread of tumour [3]. A biopsy is always performed for confirmation of diagnosis via an open or tru-cut needle technique from a posterior approach. A transrectal biopsy is contraindicated as it risks seeding the bowel. Microscopically the tumours are characterized by the distinct physaliphorous (soap bubble) cells. Despite its low grade classification, sacral chordomas are malignant and metastasize, but often quite late. The reported rate in the literature varies from 5-10% [4].

The present consensus in management is that sacrococcygeal chordomas are best treated with wide resection and in most patients with adjuvant radiation [4]. The standard surgical technique is a combined anterior-posterior approach [5]. The anterior approach allows the surgeon to develop the presacral space, making subsequent sacral transection easier and safer. The importance of complete removal of the tumour with a clear margin cannot be overstated. In his review of 50 cases, Chandawarker [6] found that aggressive resec-

tion through a combined abdomino-sacral approach offers the best results. Post operative complications included urinary incontinence in 14% and rectal incontinence in 6% of patients. A posterior approach has also been described in literature [7], but it is complex, time consuming and involves more blood loss. The authors recommend use of a mersilene mesh to prevent herniation through the sacral defect. Although chordomas are not very responsive to radiation, it is routinely prescribed postoperatively. It has been found useful for palliative treatment of pain and neurological deficit. The median survival time reported in the literature is 5 years for sacral lesions, falling to 20 to 40 percent at 10 years. Prabhakaran et al of Kidwai Memorial Institute of Oncology, India [8] reported an overall survival of 56% at 5 years and a progression free survival of 36%. However with early diagnosis and improved surgical resection a better disease-free survival can be expected for sacral chordomas.

REFERENCES

1. Mindell ER. Current concepts review. Chordoma. *J Bone and Joint Surg* 1981;63-A:501-5.
2. Rich TA, Schiller A, Suit HD, et al. Clinical and pathological review of 48 cases of chordoma. *Cancer* 1985;56:182-7.
3. Rosenthal DL, Scott JA, Mankin HJ, et al. Sacrococcygeal chordoma: magnetic resonance imaging and computed tomography. *A J Roentgenol* 1985;145:143-7.
4. Huvo AG. Bone Tumours Diagnosis. Treatment and Prognosis, 2nd ed. Philadelphia: WB Saunders 1991:599-624.
5. Ignace RS, Samson MD, Dempsey S, et al. Operative Treatment of Sacrococcygeal Chordomas *J Bone and Joint Surg* 1993;75-A:1476-84.
6. Chandwarkar RY. Sacro coccygeal chordoma: review of 50 consecutive patients. *World J Surg* 1996;20(6):717-9.
7. Waisman M, Kligman M, Roffman M. Posterior approach for radical excision of sacral chordoma. *Int Orthop* 1997;21(30):181-4.
8. Prabhakaran PS, Misra S, Kannan V, et al. Sacral chordomas: a 10 year study. *Australasia Radiol* 1998;42(1):42-6.