Research Paper Medical Science

A Rare Case of Sacral Chordoma

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ABSTRACT

Chordoma is a rare, slow growing but locally aggressive malignant tumor derived from primitivenotochordal elements, and it is usually found in the sacrococcygeal area.

Chordomas are difficult to excise completely because preservation of sacral stability and sacral nerve pathways to the rectum and bladder limit the extent of surgery.

The role of adjuvant treatment is uncertain and surgery remains the mainstay of its treatment. We present such a rare case of sacral chordoma managed in our hospital.

The clinical implications of these rare tumors are discussed.

KEYWORDS

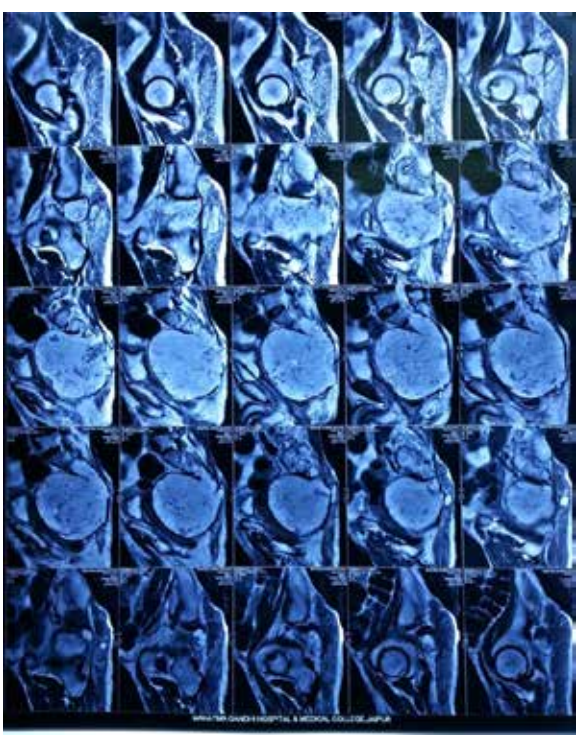
CASE REPORT

A 55 year old male was admitted with swelling and pain in lower back since 6 months.

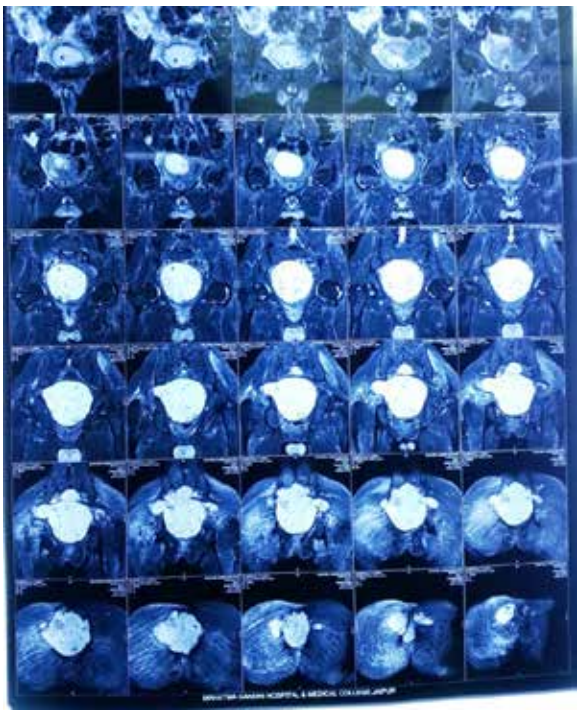
Weakness and numbness of right lower limb since 3 months and difficulty in defeacation and micturation since 8 days.

Examination revealed a 7cm x 5cm swelling in sacral region . Extending from first to fourth sacral bone.

On **rectal examination** ,a firm growth was palpable in the presacral plane and rectal mucosa was free.

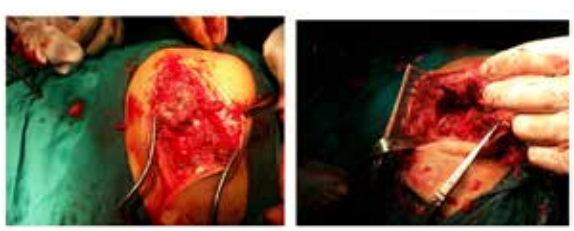


Magnetic resonance imaging (MRI) revealed a large mixed signal mass involving sacrum causing extensive bony erosion and extending into the presacral space displacing rectum anteriorly .It measures 10.5x10.2x11.0 cm. Posterior was into bilateral gluteal muscles with edema in the muscles. The mass lesion shows heterogenous contrast enhancement.Foci of hemorrage seen in the mass. Extension into bilateral SI joint was seen but there was no extension into Hip joint .



Histopathological examination shows a tumor with nodular pattern. The cells shows cord like pattern with large amount of mucinous material. The cells have vacuolated cytoplasm with loose cytoplasmic margins. At places chondroid type of differentian is also seen. **Overall picture is suggestive of chordoma.**

Near total Resection of tumor via posterior approach was done and patient had an uneventful recovery.Now patient is on regular follow up.



Tumour before excision Tumour after excision

DISCUSSION

Chordoma is a primary sacral neoplasm of ectodermal origin and makes up (1-4)% of all primary bone tumors. [1] It is usually present on the midline cerebrospinal axis and the most common locations are the spheno-clival region and the sacrum [1].

Chordomas occur in all ages and in both sexes, but the sacrococcygeal tumours are most common in fifth and sixth decades oflife [2]. It is locally aggressive slow growing malignant tumour derived from primitivenotochordal elements.[3]

About 50% arise in the sacrococcygeal area, 35% in the spheno-occipital area and the remainder along the cervico-thoraco-lumbar spine [2]. The presenting symptomin most patients is local pain. About one third of the patients also have radiculopathydue to irritation of the sciatic nerve orileo-lumbar trunk [4].

Histologically, the tumour shows classical arrangement of tumour cells in cords and lobules separated by a variable but usually extensive amount of mucoid intercellular tissue and fibrous septa [2]. Physaliferous cells are very characteristic and are large

cells with prominent vesicular nucleus and vacuolated cytoplasm. Other tumour cells are small with inconspicuous nuclei and no visible nucleoli. Mitotic figures may be scanty

or absent. Areas of bone and cartilage may also be seen. Chordoma with prominent cartilaginous foci are described as chondroid chordoma, most often seen in spheno-occipital region. The microscopic differential diagnosis includes chondrosarcoma, signet cell adenocracinoma of the rectum, myxopapillary ependymoma[2.] Immunohistochemically, the tumour cells show positivity for S-100 protein, keratin, epithelial membrane antigen, HBME-1, cathepsin k, E- cadherin, rarely for CEA [2]. Although, metastasis is infrequent at presentation, the prognosis for patients with sacral chordoma is reported to be poor. Metastases occur only in 10% ofcases to lungs, liver, lymph nodes, skin andmuscles [4]. Treatment is in the form of surgical excision, radiation therapy, or a combination of both modalities [2]. However definitive treatment is by wide excision with normal tissue margins and avoidance of spillage. Local recurrence rates are significantly increased with violation of tumour margins at initial surgery [4]

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