LETTER TO EDITOR

Sacral Chordoma - A Rare Case Report with Brief Review of Literature

Nabaneet Majumder[1], Abdul Wahid Ayubi[1], Deepak G. Paricharak[1], Seema S. More[1]*
[1]Dept. of Pathology, D. Y. Patil University, Kolhapur - 416006, (Maharashtra), India

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 of life [2]. It is locally aggressive slow growing malignant tumour derived from primitive notochordal elements. We here in report a case of sacral chordoma in a twenty seven years old female with review of literature.

Here we report a case of young female of twenty seven years presented with low backache since 5 months. X-ray of the pelvis showed a mass in the sacral region and MRI confirmed the same. The mass was surgically excised and sent for histopathological evaluation. Excised gross specimen was well encapsulated, gelatinous, globular tissue mass measuring 4.5×3×2.5 cm in diameter, soft to firm in consistency. Histology revealed a tumour composed of cells arranged in lobules over myxoid background separated by fibrocollagenous tissue. Characteristic physaliferous cells are seen which are large cells with prominent vesicular nucleus and vacuolated cytoplasm. So a diagnosis of chordoma was considered. Immunohistochemically, the tumour cells showed positivity for S-100 protein, cytokeratin, epithelial membrane antigen, thus confirming the diagnosis. Chordoma represent rare primary tumour of bone (the incidence rate is approximately 0.1/100 000/year) [3]. It is locally aggressive slow growing malignant neoplasm that arises from the remnants of the primitive notochord [4].

Chordomas occur in all ages and in both sexes, but the sacrococcygeal tumours are most common in fifth and sixth decades of life, where as many of the spheno-occipital neoplasms occur in children and adolescents. 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 symptom in most patients is local pain. About one-third of the patients also have radiculopathy due to irritation of the sciatic nerve or ileo-lumbar trunk [5].

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% of cases to lungs, liver, lymph nodes, skin and muscles [5]. 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 [5].

To conclude, we have described a case of sacral chordoma in a twenty seven year female and highlighted the morphological features of the disease. Sacral chordomas are associated with poor prognosis and if not diagnosed early, may attain large size and cause tumour related metabolic and/ or hematological complications which may limit surgical intervention. However in our case, early diagnosis was done followed by surgical excision and regular follow-up upto 6 months was possible which showed no evidence of local recurrence or any complication. So, early diagnosis and treatment are very essential.

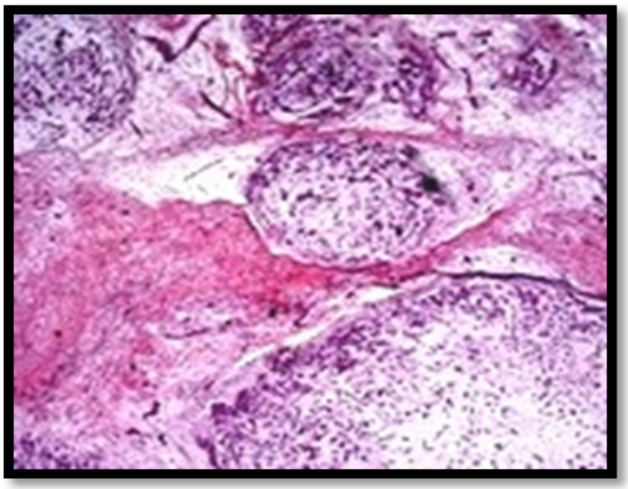

Fig. 1: Microphotograph shows tumour cells arranged in lobules over myxoid background separated by fibrocollagenous tissue (H & E, x 4)

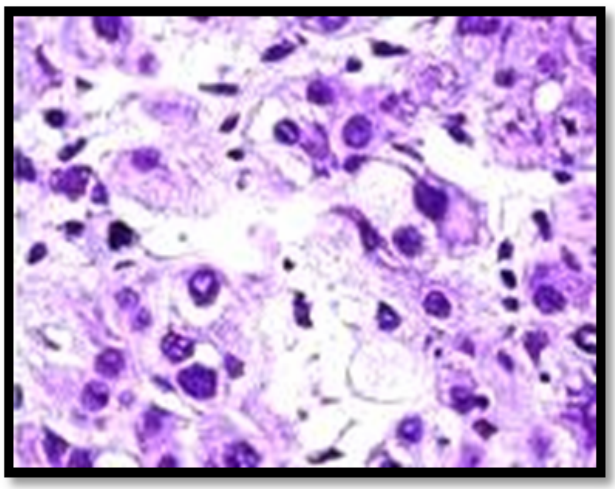

Fig. 2: Characteristic physaliferous cells (H and E, x 40)

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***Author for Correspondence:** Dr. Nabaneet Majumder, Dept. of Pathology, D. Y. Patil University, Kolhapur - 416006, Maharashtra, India Cell: 08237788472 Email: drnabaneetmajumder@gmail.com*