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Case Report: Cyto-Histological Diagnosis of a Sacral Mass, Suspected as Sarcoma Clinico-Radiologically

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Abstract*—Clinically a Sacrococcygeal mass may attain massive size causing suspicion of a sarcomatous lesion. Such masses may have diverse aetiology ranging from inflammation to neoplasia. We present here cytological findings of a sacral mass of 70 yrs. male, clinically & radiologically suspected to be sarcoma. The initial radiologic diagnosis was Sarcoma/Metastatic/ Chordoma. FNAC finding of this case was consistent with Chordoma which was later confirmed on HPE.*

***Keywords*—** *Chordoma, sacrococcygeal, cytology, histopathology.*

I. INTRODUCTION

A sacrococcygeal mass may attain massive size arising suspicion of a sarcomatous lesion. Chordoma is one such lesion which may appear sarcomatous Clinico-radiologically by its size and aggressive behaviour. It frequently occurs in fifth and sixth decades and affect much more often men than women. Site of involvement include sacrococcygeal (50%), spheno-occipital region (35%) and in the true vertebrae (15%). They are commonly located at the cranial and caudal ends of the primitive notochord. Local recurrences are common, and distant metastases may occur [3]. They are composed of large and small tumour cells arranged in lobules in a background of myxoid stroma. The cells have abundant clear, eosinophilic, or multivacuolated (physaliphorous) cytoplasm.

Case Report:

A 70-year-old male was admitted in this hospital with a complaint of constipation, urinary incontinence and pain with numbness on left hip. Pain is radiated to the left foot which increased with physical activity.

CMRI of Lumbosacral spine showed a large, expansile, destructive soft tissue tumour of sacrum (fromS2 segment to coccyx) and bilateral sacral –ala. A differential diagnosis of Sarcoma / Metastasis/Chordoma was suggested radiologically.

FNAC of the mass showed discrete and loose clusters of large round to oval cells with vacuolated cytoplasm and vesicular nuclei in a background of abundant myxoid stroma. There are some large cells with bubbly cytoplasm (physaliphorous cells) [Figure 1]. These cytomorphological features are suggestive of Chordoma.

Operative findings revealed tumor mass at S2 level and going up to lateral rectal margin and eroding pre-sacral and sacral spinal canal. It was pink, friable and soft to firm and vascular mass intra-operatively. The mass was clinically diagnosed as sacral sarcoma.

We received multiple grayish-white to brown pieces which had soft gelatinous consistency.

Histopathological examination revealed tumour cells separated into lobules by thin fibrous septa. The neoplastic cells were arranged in nests and had abundant eosinophilic bubbly cytoplasm and round regular nuclei with inconspicuous nucleoli in a background of myxoid stroma. The large cells with bubbly cytoplasm are the physaliphorous cells, which is characteristic of chordoma [Figure 2].

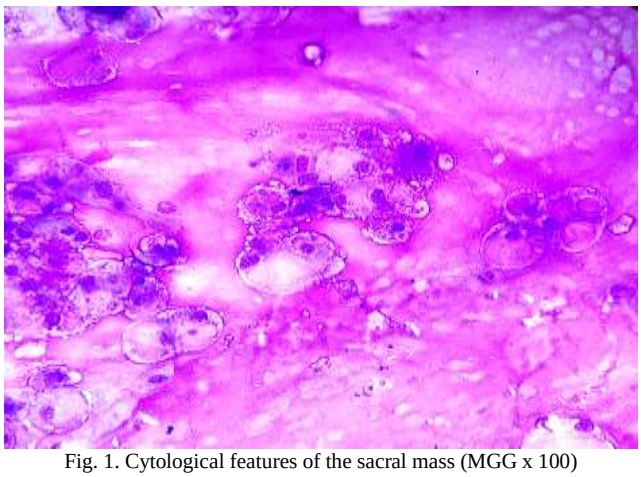

Fig. 1. Cytological features of the sacral mass (MGG x 100)

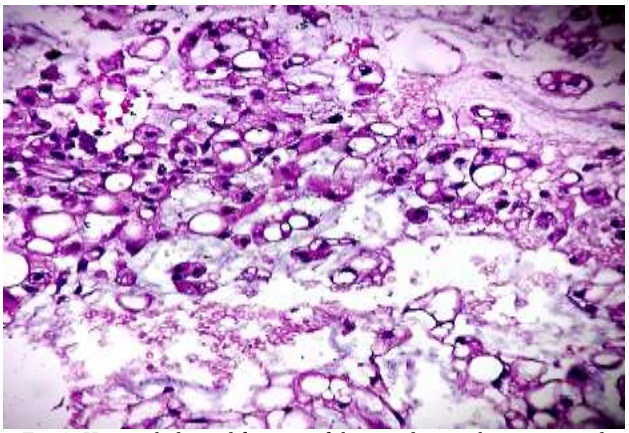

Fig. 2. Histopathological features of the sacral mass showing typical physaliphorous cells (H&E X 400)

II. DISCUSSION

An aggressive sacrococcygeal mass may rarely present with features of Chordoma and forms 1% to 4% of all malignant bone tumours [1,5]. In this tertiary care institute

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only five (05) cases had been diagnosed over a period of five years, from 2013 to 2017, indicating its rarity. Out of these, three occurred in the sacrococcygeal region and the rest two were in Clival area. All the cases were males. The tumour is common in the fifth to seventh decade of life though any age group may be affected [2]. The inherited disease is sometimes associated with duplication of the Brachury gene (T gene that control notochord differentiation) [6].

In Sacrococcygeal chordoma the presenting symptoms are mostly pain in the hip and signs and symptoms related to the involvement of sacral and Pre-Sacral areas. Chordomas arising in the sacral region may present with chronic low back pain and sometimes with urinary symptoms [2]. In our case, the patient is and old man of 70, who presented with radiating pain towards hip associated with constipation and urinary incontinence.

Morphologically, chordoma is composed of epithelioid cells showing prominent vacuoles—giving it the characteristic physaliphorous appearance—and arranged in cords within a myxoid to chondromyxoid matrix. Presence of cells having large uni or multivacuolated cytoplasm is diagnostic of chordoma. Crapanzano et al. (3) studied 12 cases and found that nine of the twelve cases showed the classic physaliphorous cells of chordoma. Sometimes the cytoplasmic vacuolation result in nuclear indentation or signet ring like cells [3, 4]. Myxoid background is also an important finding for diagnosis of chordoma. It is essential to rule out other tumours having a myxoid background. Our case showed the typical cytologic features of chordoma. Some cases might show pleomorphism, with physaliphorous cells having hyperchromatic nuclei and prominent nucleoli [3, 7].

Our case showed features of conventional chordoma with tumour cells arranged in lobules along with typical physaliphorous cells.

Differential diagnosis of chordoma in cytology is with those tumours which have myxoid stroma and vacuolated cytoplasm. (Table 1)

TABLE 1. Showing cytological differential diagnosis of tumours which have myxoid stroma and cells with vacuolated cytoplasm [1, 2, 5]:-

Features	Chordoma	Chondrosarcoma	Metastatic Carcinoma	Myxoid Liposarcoma
Site	Sacrococcygeal spheno-occipital region and true vertebrae	Long bones	Lung, Liver, Bone, Lymph node, Brain, solid abdominal organs, Peritoneum and even in the Breast	Soft tissue
Cellular characteristics	Presence of physaliphorous cells	No physaliphorous cells	Glandular or papillary clusters and columnar cells may be present. No physaliphorous cells	Lipoblasts. No physaliphorous cells
IHC	Express keratins, S100 , EMA and Brachury	S100+ve, EMA & CK -ve	Metastatic panel	Mucicarmine & CK negative

Classic radiologic finding of chordoma is a destructive soft tissue mass of the sacrococcygeal region with calcification [8]. Recurrence is common. Metastasis has been reported in many organs [2]. Our case had local extension to the spinal cord without any distant metastasis. The prognosis of chordoma depends on the size of the tumour, site, completeness of resection and postoperative irradiation received.

III. Conclusion

Sacrococcygeal neoplastic lesions are considered to be a diagnostic and therapeutic challenge. A lot of awareness and suspicion is necessary when dealing with chronic sacrococcygeal pain and sacrococcygeal masses. Chordoma having massive size and involving nearby structures may mimic many sarcomas. However, these have characteristic cytomorphologic feature of physaliphorous cells in a myxoid background, which help in accurate diagnosis. Radiologic findings may aid in both the diagnosis and preoperative planning. Histopathological correlation is important for complete diagnosis and further management of the case.

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