

CASE SERIES

Cytological Features of Parachordoma: Two Case Reports of a Rare Entity*Neelam Sood¹, Kumar Sharad Sinha^{1*}**¹Department of Pathology, Deen Dayal Upadhyay Hospital, Govt. of NCT,
New Delhi- 110064(Delhi) India***Abstract:**

Parachordoma is an uncommon tumour of soft tissue of uncertain origin. Recurrence and metastasis are rarely seen. We report two cases with one presenting as recurrent case of pelvic parachordoma in a 45 year old female who presented with painless pelvic mass since 2 months. The other case report is of a 30 year male patient who presented with a diffuse swelling over left palmar aspect. Cytology findings of the pelvic swelling revealed moderately cellular smears composed of large physaliphorous cells with abundant bubbly cytoplasm in clusters and singly scattered medium sized epithelioid like cells with round nuclei, vesicular chromatin and prominent nucleoli in background of abundant myxoid ground substance. Cytology of the Palmar swelling revealed moderate cellularity comprised of oval to spindle shaped cells with coarse chromatin, prominent nucleoli and abundant vacuolated cytoplasm in a chondromyxoid stromal background. The preliminary diagnosis on cytology with further confirmation on histopathology with the aid of immunohistochemistry helped clinch the diagnosis of this rare entity of parachordoma occurring at unusual sites.

Keywords: Cytology, Recurrent, Parachordoma, Physaliphorous

Introduction:

Parachordoma (or myoepithelioma) is a rare tumour which was first described by Laskowski, but later named and described by Dabska in 1977 [1]. The tumour was initially categorized as mixed tumour / myoepithelioma in 2002 (WHO classi-

fication). However, it has now been placed in the category of tumours of uncertain differentiation and is renamed as myoepithelioma / myoepithelial carcinoma / mixed tumour by WHO in 2013 [2, 3]. To the best of our knowledge, these are the first cases of pelvic parachordoma showing recurrence based on radiological findings and parachordoma occurring in the palm.

Case Reports:**Case Report-1**

A 45 year old woman presented with slowly increasing mass in pelvic region since 2 months. Computerized Tomography (CT) revealed a 78x74x48 mm solid cum cystic mass lesion with lobulated outline in pelvic subcutaneous fat (Fig.1). Previously it was diagnosed parachordoma occurring in the same site 20 months back. Fine needle aspiration was done and smears were stained with Giemsa stain and Hematoxylin and Eosin stain. Cytology revealed moderately cellular smears composed of large physaliphorous cells with abundant bubbly cytoplasm along with clusters and single scattered medium sized epithelioid like cells with round nuclei, vesicular chromatin and prominent nucleoli in background of abundant myxoid ground substance. Anisonucleosis was noted but there were no mitotic figures [Fig.2, 3 (a, b)]. Based on CT findings and detailed history, a diagnosis of recurrent parachordoma was given.

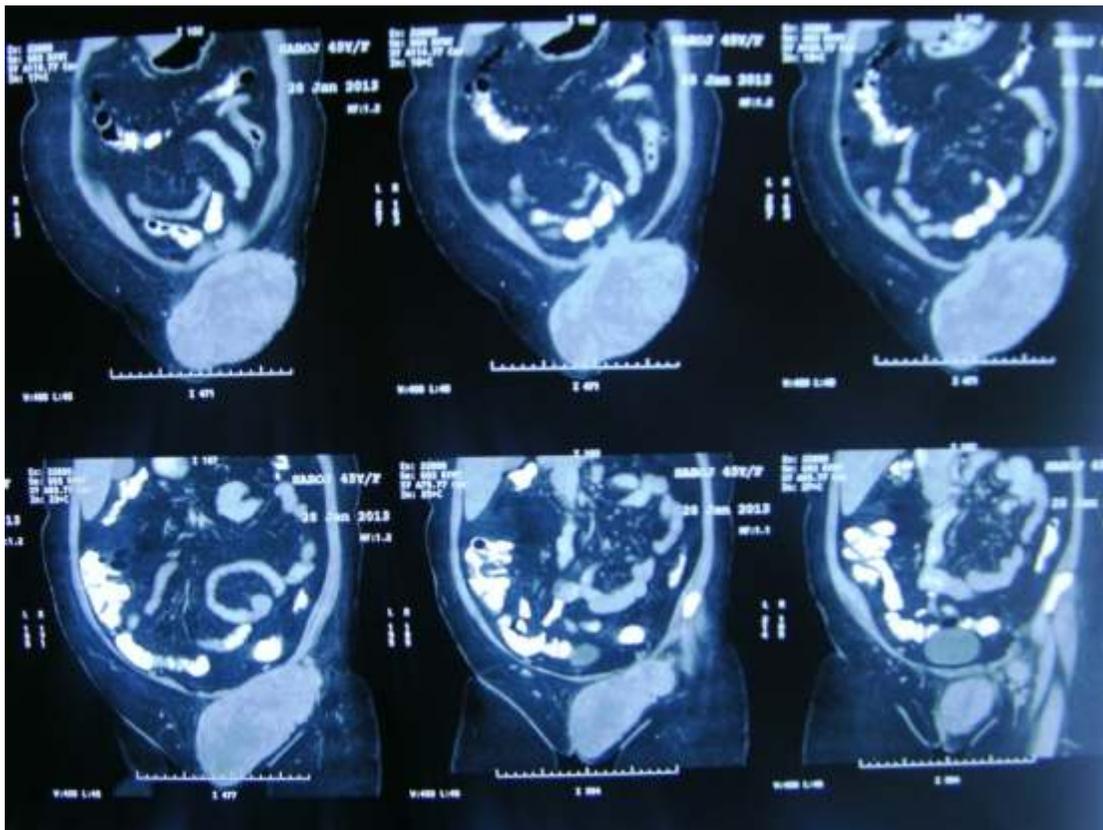


Fig. 1: CT Scan Showing Lobulated Cystic Mass

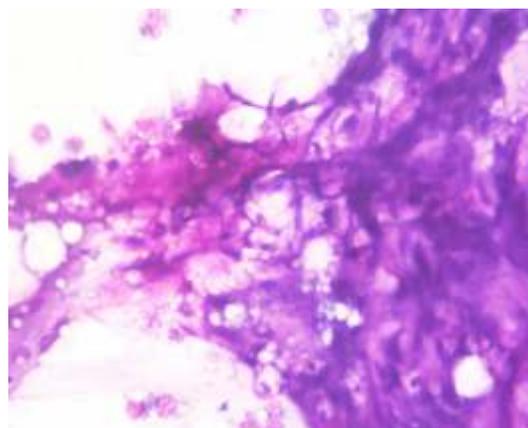


Fig. 2: Physaliphorous Cells and Epithelioid Cells in a Myxoid Background (Giemsa, 100X)

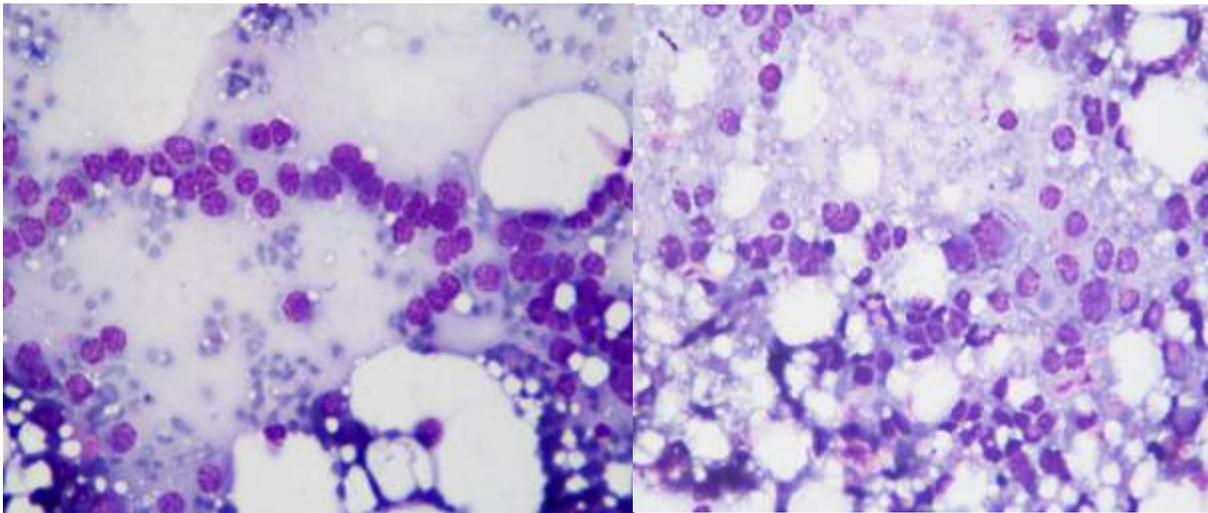


Fig. 3(a, b): Epithelioid Like Cells in a Myxoid Background (Giemsa,100X)

Case Report-2

A 30 year male patient presented with a diffuse swelling over left palmar aspect measuring 5 x 4 x 1 cm. FNAC of the swelling was performed which revealed moderate cellularity comprised of oval to

spindle shaped cells with coarse chromatin, prominent nucleoli and abundant vacuolated cytoplasm in a chondromyxoid stromal background (Fig.4).

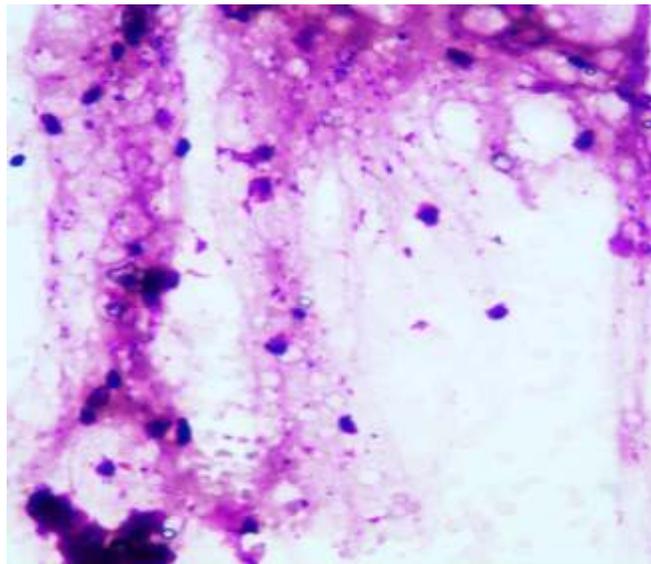


Fig 4: FNAC Shows Oval to Spindle Shaped Cells in a Chondromyxoid Background (Giemsa, 100X)

Discussion:

Currently, parachordoma is considered a part of the broad morphological and immunophenotypic spectrum of soft tissue myoepithelioma/mixed tumours. The histogenesis of parachordoma is still uncertain [4]. Common reported sites are extremities, less common sites are head and neck, trunk and bone [5]. This tumour has little male predilection,[1] however, in our case one patient is male and other female. In the largest study of 101 myoepithelial tumours carried out in Brigham and Women's hospital it was found that it occurs in a wide age range (3-83) with mean age of 38 yrs. It has been reported to be as small as 0.7 cm to as large as 20 cm with mean size of 4.7 cm [6]. Clinically patient presents with slowly enlarging painless mass, the tumour may be situated primarily in the subcutis or deep to the fascia [5]. In our two cases, patients presented with a slow growing mass since 2 months in the subcutaneous fat of pelvis and swelling of the left palmar aspect. Cytomorphology of this tumour is not widely documented and is reported only twice in the literature [7, 8].

The preliminary diagnosis on cytology with further confirmation on histopathology with the aid of immunohistochemistry helped clinch the diagnosis of this rare entity occurring at unusual sites. This group of tumours show morphological heterogeneity. Thus, the differential diagnosis depends on the dominant cell type and stromal component and includes extraskeletal myxoid chondrosarcoma, chordoma, ossified fibromyxoid tumour, metastatic carcinoma, clear cell sarcoma, metastatic malignant melanoma and epithelioid sarcoma. Extraskeletal myxoid chondrosarcoma shows eosinophilic spindle cell cords that create multinodular patterns and ovoid cells within a myxoid matrix. Extraskeletal myxoid chondrosarcoma cells are smaller and more intensely eosinophilic with a less vacuolated cytoplasm than parachordoma cells [2]. Parachordomas generally express CK and especially CK8/18, while CK expression is not seen in extraskeletal myxoid chondrosarcomas [2] which shows S-100 positivity [9]. Chordomas presents on axial skeleton, shows lot of physaliphorous cells and are positive for CK 1/10, CK 7, 20, 19 and CK 12-17. Alcian blue staining is

Studies	Age	Sex	Site	Clinical features	Cytology	Recurrence
Zardawi <i>et al</i> 2010 [7]	17 y	M	Buttock	Slow growing painless mass	Cellular smears, atypical cells including spindled and epithelioid cells in myxoid background	No
Samaka <i>et al</i> 2012 [8]	46 y	F	Pelvic	Painful mass, irritative urinary bladder symptoms	Cellular smears, spindled and epithelioid cells in myxoid background	No
Present cases	45 y	F	Pelvic	Painless mass	Cellular smears, large physaliphorous cells with epithelioid cells in myxoid background	Yes
	30 y	M	Palm	Painless mass	Moderate cellularity with oval to spindle shaped cells with coarse chromatin, prominent nucleoli and abundant vacuolated cytoplasm in a chondromyxoid stromal background	No

also positive in case of chordoma after hyaluronidase predigestion in contrast to parachordoma [9]. Epithelioid sarcomas frequently express CK and EMA but S-100, GFAP and myogenic markers are negative. Melanomas typically express melanocytic antigens such as Melan-A, HMB-45 and S-100 [2].

Recurrence and metastasis are rarely seen. There are contradictory views on the surgical margins and recurrence association in the literature. Surgical treatment can provide a cure in parachordoma but tumours that have not been fully excised can recur and local recurrences and metastases are more frequent in patients with a histologically malignant appearance [2]. Sometimes, recurrences and metastasis may occur from the tumour negative margins [10]. One of our cases had a recurrence after 20 months of initial surgical resection.

Parachordomas are benign tumours but metastases and recurrences are not unusual. Recurrence is seen as early as 3 months or as late as 12 years after the initial surgery, although early

recurrence is rare. Metastatic potential in parachordoma has been poorly described in the literature, however, there have been six reported cases of metastasis from parachordoma out of which four cases showed metastasis in lung and other two cases in lymph node, testis and hip respectively [4]. On recurrence, our patient also showed a 5 cm large lung nodule and an enlarged left inguinal lymph node on CT scan suggestive of metastasis however it could not be evaluated further. Parachordomas are benign tumours but they can turn malignant and show metastasis and recurrence and cases should therefore be followed-up closely.

Conclusion:

The clinical follow up of the primary site is important because of the significant potential for local recurrence in parachordoma. In addition, long term follow-up, including lung CT scan and bone scintigraphy are important in cases of locally invasive and/or recurrent tumours because of their metastatic potential.

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