Parachordoma: Unusual entity at an unusual site

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ABSTRACT

Parachordoma is a rare tumor of soft tissue with unspecified lineage. According to the World Health Organization (WHO) classification of 2002, the tumour has been included in the same category as mixed tumors and myoepitheliomas. Metastasis and recurrence are rarely reported. We have reported a case report of Parachordoma in Left Palm.

Cytological features revealed moderately cellular aspirates comprised of dual population of epithelioid and spindle shaped cells in a chondroid to myxoid background. Histopathologically, the tumor was composed of lobules of polygonal to spindle shaped cells with pale eosinophilic to clear cytoplasm with mild pleomorphism and infrequent mitotic activity in a chondro myxoid stroma with intervening fibrous tissue. Tumor cells were positive for EMA, S-100 and vimentin on immunohistochemistry(IHC). Occurrence of parachordoma at a rare site prompted us to present this case report.

Key words: Chordoma, Parachordoma, Palm, World Health Organization (WHO) Classification

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INTRODUCTION

Parachordoma represents a soft tissue tumor comprising of cells having histological and ultrastructural characteristics similar to those of chordoma cells. It has been included in the same class as mixed tumors and myoepitheliomas, according to the World Health Organization (WHO) classification (2002).

On the other hand, with immuno-histochemistry, they have features similar to that of chondroid tumor cells [1,2].This tumor most often arises from upper or lower limbs.

Parachordomas are treated by complete removal of lesion [3]. Sometimes, these tumors are locally destructive and tend to recur if incompletely excised [4-6].

Parachordoma was first reported by Dabska in 1977 [1]. They typically develop in sites adjacent to synovium, tendon, or osseous structures in extremities and usually present as slowly growing but locally destructive, lobulated neoplasms. They are prone to recurrence if incompletely excised [3]. These tumors are commonly localized in soft tissue, particularly at upper or lower limbs. Ectopic notochord, could be the origin similar to chordomas or other neuron-related cells [7].A role for trauma is implied in the etiopathogenesis of parachordomas[3] .Generally, these tumors are considered to be a benign mass; however, they may also be locally aggressive or lead to distant metastasis [1,4].

On macroscopic evaluation, parachordomas are frequently seen as a well-defined nodular mass, ranging from 3 to 7 cm in diameter .They have a yellow-white cross-sectional surface with myxoid or gelatinous condition [4].

Histopathologically, it is characterized by well circumscribed lobules comprising of cellular aggregates, as well as single, large, vacuolated cells embedded within a hyaline and chondroid matrix divided by fibrous bands. There is mild pleomorphism and minimal mitotic activity[3] .

They show two distinct patterns: cellular areas with cohesive nests or ribbons of cells with abundant, clear cytoplasm and a high nuclear cytoplasmic ratio set in a large myxoid background and chondroid areas, with clear cells set in a cartilagenous matrix [7].Approximately 25% to 50% of the tumor cells express cytokeratin 8/18, focal chromogranin A, epithelial membrane antigen (EMA) ,S-100 protein and vimentin. However, CD

34, actin, glial fibrillary acidic protein (GFAP) and calponin are typically negative [3,5,6,8].

CASE REPORT:

A 30 year male patient presented with a diffuse swelling over left Palmer aspect measuring 5x4x1cm.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 chondomyxoid stromal background (Fig 2g).

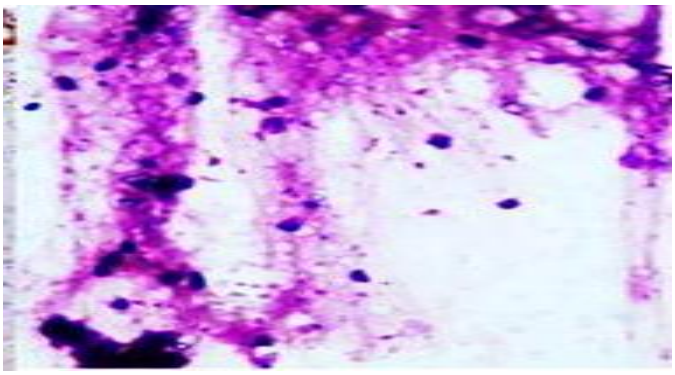

Figure 2(g): FNAC shows oval to spindle shaped cells in a chondromyxoid background (Giemsa,100X)

Thus, the features were suggestive of a benign mesenchymal lesion.Surgical excision of the mass was performed. Specimen was received as a globular, enacapsulated soft tissue mass measuring 4.5X3X1.5 cm. Cut surface showed grey white, with solid and gelatinous areas. (Figure1a-b).

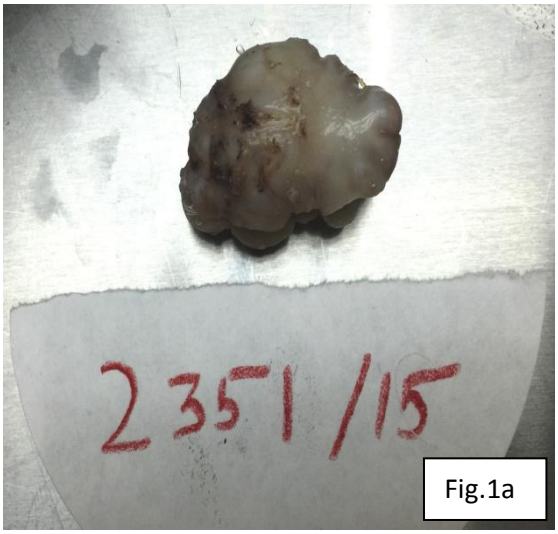

Fig.1a

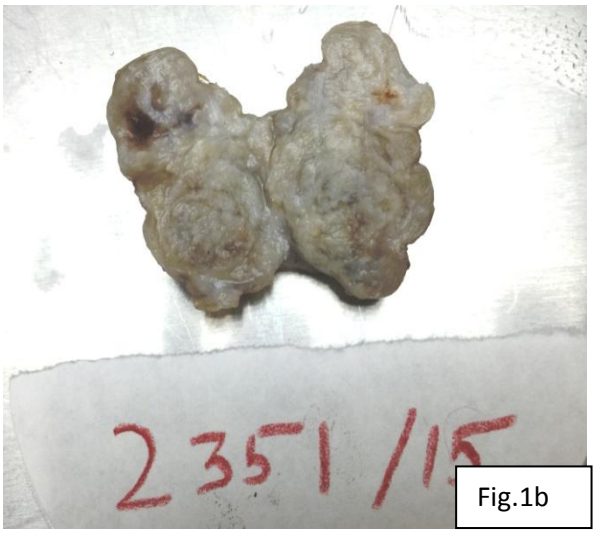

Fig.1b

Figure 1 (a)-Gross specimen received as globular, enacapsulated soft tissue mass.

Figure 1 (b) Cut surface showed grey white, with solid and gelatinous areas

Sections stained in Haematoxylin and Eosin, showed tumor tissue arranged in lobular arrangement separated by a loose fibromyxoid stroma having a pseudochondroid appearance with presence of polygonal to spindle cells which were uni and binucleated with cytoplasmic vacuolization with cells showing mild anisonucleosis and infrequent mitosis fig. 2(a-c).

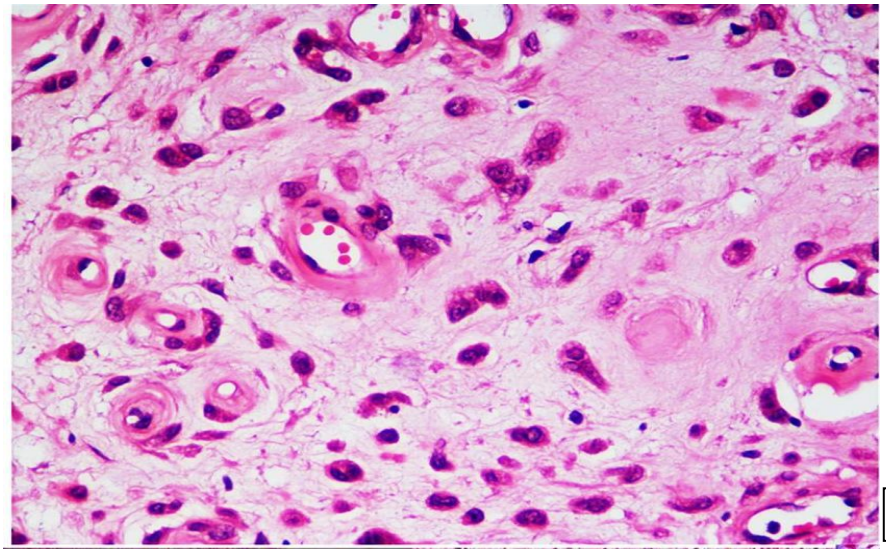

Fig.2a

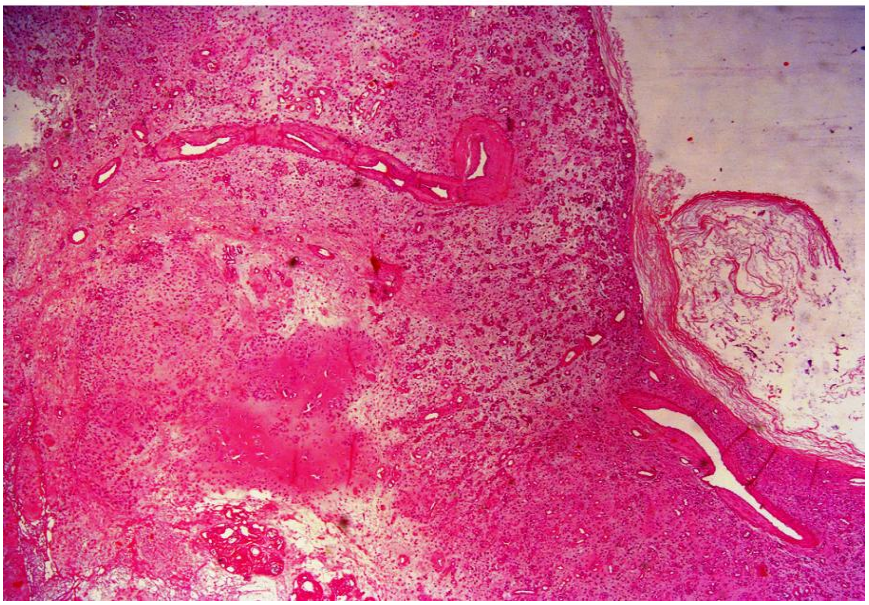

Fig.2b

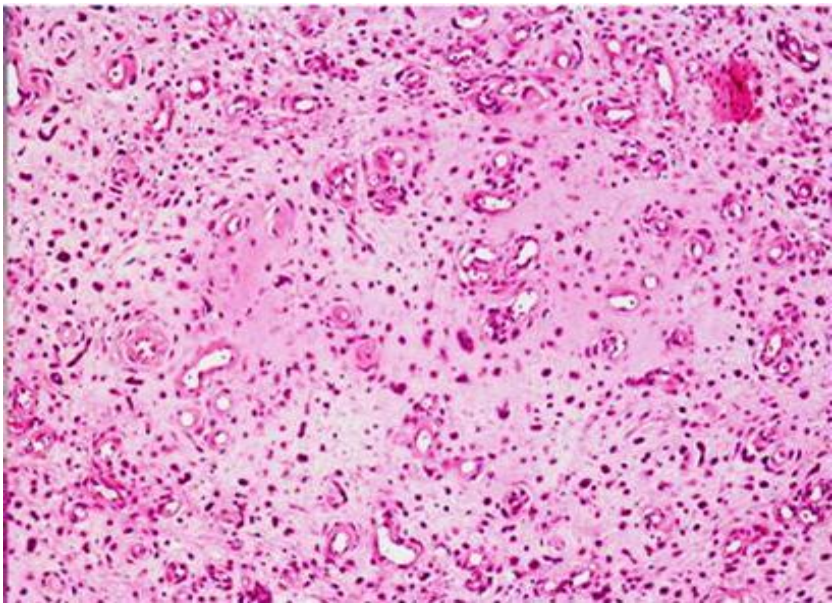

Fig.2c

Figure 2(a)- Polygonal to spindle cells which were uni and binucleated with cytoplasmic vacuolization(H&E 400X)

Figure 2(b-c)- Tumor tissue arranged in lobular arrangement separated by a loose fibromyxoid stroma having a pseudochondroid appearance(H&E 40X,100X)

PAS demonstrates intracytoplasmic positivity of the cells (Figure 2d)

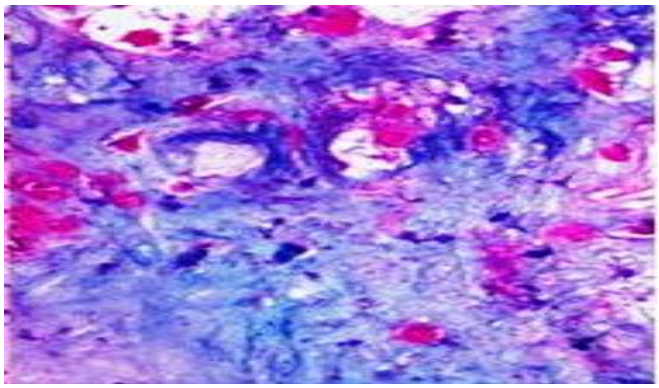

Fig.2d

Figure 2(d): Tumor cells showing intracytoplasmic PAS positivity(100X)

IHC STUDY (clone retrieval): Tumor cells were diffusely immunopositive for Vimentin, EMA (Figure 2e) and S-100 (Focal) but negative for CD34 (Figure 2f),calponin, SMA and Desmin.

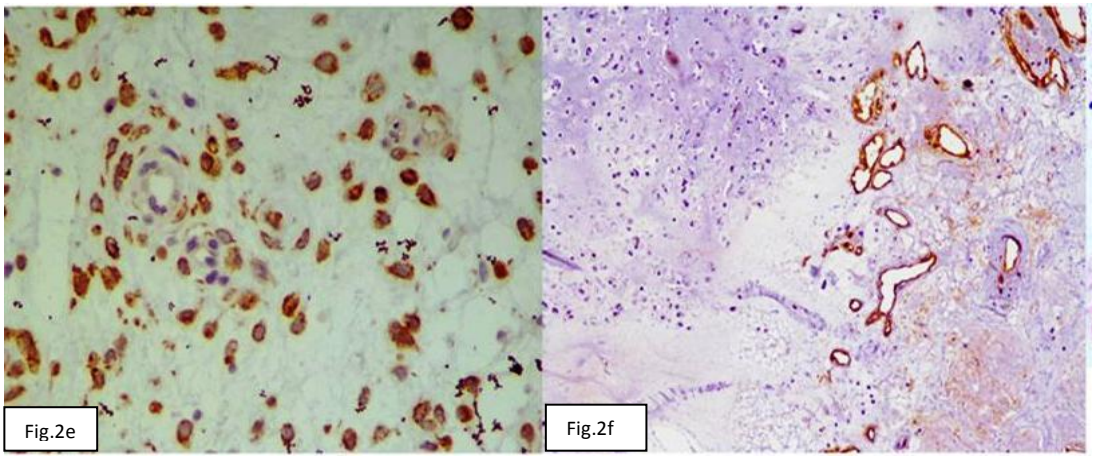

Figure 2(e)- Tumor cells positive for EMA (100X)

Figure 2(f)-Tumor cells negative for CD34(100X)

DISCUSSION

Parachordoma is an uncommon benign soft tissue tumor, in adults with infrequent recurrence [3]. It is first reported by Laskowski in 1959 [6] .There is a slight male peponderance [6] .The mean age is 34.4 years [range 4-86 years], however 20% of the cases are in pediatric population [9,10]. The common sites of the parachordoma are the extremities, head, neck, thorax, bones and rarely in the pelvis. Parachordomas are usually painless and slow-growing masses [12] .

There are various theories about the Histogenetic origin of parachordomas is debatable with no widely accepted view.The commonly accepted theory suggests development from progenitor cells with a chondroid differentiation capability or the bone surface associated with tenosynovial or aponeurotic tissue or the periosteum, or subcutaneous tissues [12]. It's relation with ectopic notochord remnants, synovial tumors including synovial sarcomas and lipoblasts and chondroblasts has also been postulated. Most widely accepted belief is it's myoepithelial cell origin [13,14] .

The tumor is usually well-circumscibed, sometimes surrounded by a thin fibrous pseudocapsule. It's size varies from 3 to 7 mm and sometimes grow up to 12 cm [11,12] .Low magnification on histopathology characteristically shows that the tumor consisting of small nests of epithelioid cells, cords and focal pseudoglandular structures, with a myxohyaline stroma. Pseudoglandular

structures was not noted in this case.These proliferating cells resemble physalipharous cells of classical chordoma.

The myxoid material is oulined by intracytoplasmic vacuoles having glycogen as demonstrated by PAS staining. The nuclei are variable from vesicular chromatin with prominent nucleoli to a small pyknotic type. Multinucleation is usually noted. Mitotic activity is however minimal. No lymphatic or vascular invasion is seen. Occasionally they have spindle cells with narrow cytoplasm , hyperchromatic nuclei, arranged in cords which are surrounded by a fibrous stroma. The stromal matrix varies from pure myxoid to chondroid or hyalinelike composition [12] .Morphological heterogeneity is a usual feature.

Depending on the dominant cell type and stromal component the differential diagnoses are considered and includes extraskeletal myxoid chondrosarcoma, chordoma, ossified fibromyxoid tumor, metastatic carcinoma, clear cell sarcoma, metastatic malignant melanoma and epithelioid sarcoma.

Extraskeletal myxoid chondrosarcoma shows presence of eosinophilic spindle cell cords ,creating multinodular patterns and ovoid cells within a myxoid matrix. It generally extends to the deep skeletal muscle. They have smaller cells which are more eosinophilic having a less vacuolated cytoplasm than parachordoma cells [12] .Although few extraskeletal myxoid chondrosarcoma cases stained with S-100 but CK expression is not seen in extraskeletal myxoid chondrosarcomas ,unlike parachordoma as seen in this case [2]. Chordomas are seen in older patients and have a higher risk of recurrence and have sacral and vertebral unlike the parachordomas[10] . The microscopically chordomas and parachordomas may resemble but a chondroid matrix is relatively rare in parachordomas[2]. Epithelioid sarcomas express CK and EMA but are negative for S-100, GFAP and myogenic markers. Melanomas characteristically express melanocytic antigens i.e. Melan-A, HMB-45 and S-100[2].

Few differentiating points between the entities based on IHC are given in the table below -

IHC	CK7	S-100	EMA	CK-19	CEA
Parachordoma	-	+	+	-	-
Chordoma	+/-	+	+	+	+
Extraskeletal Myxoid chondrosarcoma	-	-	-	-	-

(Positive +, Negative -)

Surgical treatment can be curative tumors with incomplete excision may recur [12] .There is no clear relationship between the positive surgical margins and subsequent recurrence but there is a correlation with histologically malignant appearance [2] .

CONCLUSION

Parachordoma is a rare entity that can affect any site of the body. Imaging and Fine needle aspiration cytology can direct us to the tumor but exact diagnosis can only be made on basis of histo-pathological features including the immuno-histochemistry. A clear diagnosis has definite therapeutic implications. Palmar aspect is extremely rare site for occurrence of Parachordoma.

REFERENCES

1. Gimferrer JM, Baldo X, Montero CA, Ramirez J. Case report: Chest wall parachordoma. Eur J Cardiothorac Surg 1999;16(5):573-5.

2. Weiss SW, Goldblum JR: Enzinger & Weiss's Soft Tissue Tumors. 5th ed., Philadelphia, Mosby Elsevier, 2008, 1109-1116.

3. Sangueza OP, White CR. Parachordoma. Am J Dermatopathol 1994;16(2):185-8.

4. Dabska M. Parachordoma: a new clinicopathologic entity. Cancer 1977;40(4):1586-92.

5. Guedes A, Barreto BG, Barreto LG, Araujo IBO, Queiroz ,AAAthanazio DA etal . Metastatic parachordoma. J Cutan Pathol 2009;36(2):270-3.

6. Karakaya YA, Ozekıncı S, Büyükbayram H, Mizrak B. Parachordoma:a recurrent case and review of the literature. Turk Patoloji Derg 2011;27(2):173-6.

7. Tihy F, Scott P, Russo P, Champagne M, Tabet JC, Lemieux N. Cytogenetic analysis of a parachordoma. Cancer Genet Cytogenet 1998;105(1):14-9.

8. Behzatoğlu K, Canberk S, Bahadir B, Oznur M. Parachordoma or myoepithelioma?: a case report of a rare soft tissue tumor. Pathol Int 2007;57(3):167-70.

9. Tirabosco R, Mangham DC, Rosenberg AE. Brachyury expression in extra-axial skeletal and soft tissue chordomas: a marker that distinguishes chordoma from mixed tumor/myoepithelioma/parachordo-ma in soft tissue. Am J Surg Pathol 2008;32:572-580.

10. Lococo F, Cesario A, Meacci E. Pulmonary metastases from parachordoma. Ann Thorac Surg 2009;88:9-10.

11. Hornick JL, Fletcher CD.Myoepithelial tumors of soft tissue: a clinicopathologic and immunohistochemical study of 101 cases with evaluation of prognostic parameters. Am J Surg Pathol 1997,27:1183-96

12. Silverberg SG, DeLellis RA, Frable WJ, LiVolsi VA, Wick MR:Silverberg's Principles and Practice of Surgical Pathology and Cytopathology. 4th ed., Philadelphia, Churchill Livingstone, 2006, 395-6.

13. Kilpatrick SE, Hitchcock MG, Kraus MD, Calonje E, Fletcher CD: Mixed tumors and myoepitheliomas of soft tissue: a clinicopathologic study of 19 cases with a unifying concept. Am J Surg Pathol 1997, 21:13-22

14. O'Connell JX, Berean KW: Parochordomas. Am J Surg Pathol 1997, 21: 395-6.