Case Report

Clear cell sarcoma (CCS) involving the dorsum of right foot and its deeper tissue – A case presentation

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Abstract

Clear Cell Sarcoma or Malignant Melanoma of soft parts is a rare aggressive tumor involving the tendon and aponeuroses with melanocytic differentiation and distinct genetic background. It shows histological immunohistochemical and ultrasturctural resemblance with malignant melanoma causing major diagnostic confusion. Here we present a case of 35 year old female with clear cell sarcoma involving the right foot with involvement of deeper tissue.

Keywords: Clear cell sarcoma, tissue.

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INTRODUCTION

Clear Cell Sarcoma is a rare soft tissue tumor showing predilection for the deeper soft tissue of the lower extremities close to the tendon, fascias or aponeurosis[1]. It occurs in young adults between the age group of 20-40 years,[2,3] and with involvement of lower extremities particularly around the foot and ankle region accounting for nearly 40% cases. The overall incidence comprises less than 1% of all soft tissue sarcoma.[4]

CASE REPORT

A 35 years old female patient presented with swelling of 6 months duration, involving the right foot over dorsum of III and IV metatarsal phalangeal joint, adherent to the tendon with history of tenderness. The patient underwent surgical wide excision and specimen was submitted for histopathological examination.

Gross

On gross inspection, the tumor was an irregular grey white soft tissue mass measuring 4 x 2.5x2 cm, partially covered with skin. C/S – the mass is grey white lobulated.

Microscopic examination

The Histopathological examination revealed a tumor characterized by fascicular to nested pattern delineated with variable thickness fibrous septa. The tumor cells were polygonal to fusiform shaped with large pale eosinophilic to clear cytoplasm and vesicular nuclei with prominent nucleoli. Some tumor cells containing cytoplasmic pigments. The tumor did not show any evidence of connection to the epidermis.

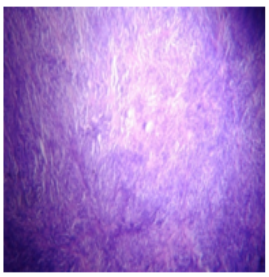
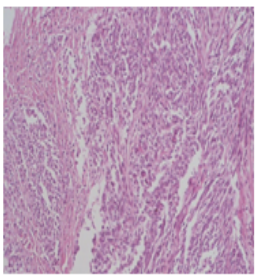


Figure 1 (a)



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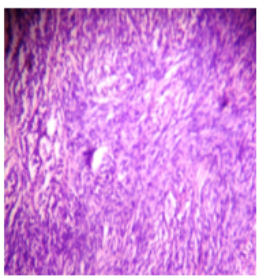

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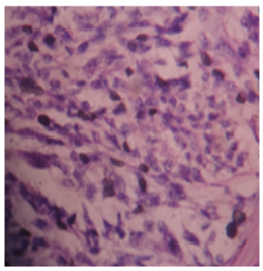

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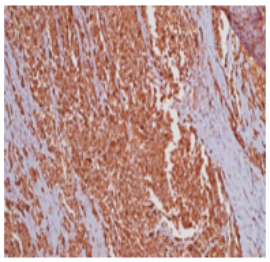

Figure 4

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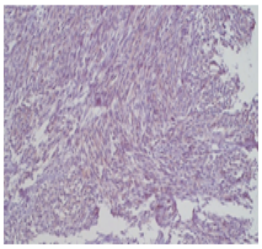
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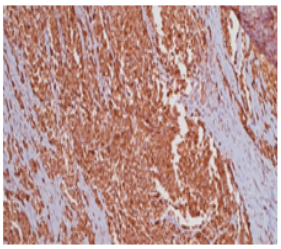
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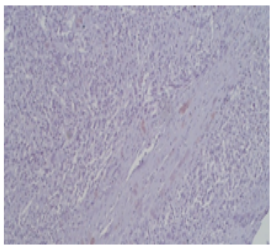
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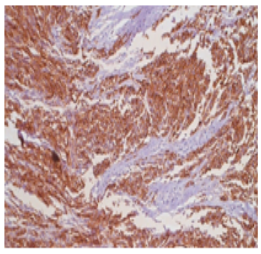
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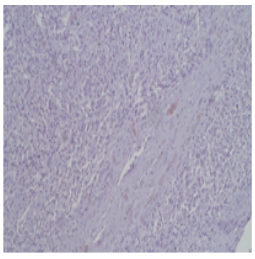
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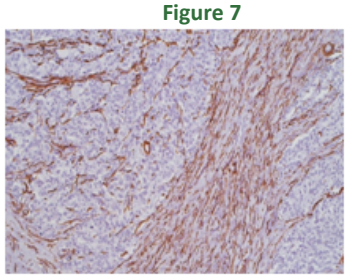
Figure 10

Figure 1 (A and B): H and E, x 100 Nests and fascicles delineated by fibrous septa
Figure 2: H and E, x 400 Spindled to epitheloid cells with pale eosinophilic cytoplasm and vesicular nuclei and prominent nucleoli
Figure 3: H and E, x 400 Spindled to epitheloid cells with cytoplasmic pigments
Figure 4: S100 Diffusely Positive
Figure 5: EMA Negative
Figure 6: BCL2 Diffusely Positive
Figure 7: CD 99 Negative
Figure 8: HMB 45 Diffusely Positive
Figure 9: Ki67 Positive 0-1%
Figure 10: SMA Negative

Immunohistochemistry

Immunohistochemistry showed tumor cells exhibiting diffuse positivity for S100, HMB45 and BCL_2 and tumor cells were negative for EMA, SMA, MIC_2.

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DISCUSSION

Dr.Franz Enizinger reported in 1965 Clear Cell Sarcoma of tendon and aponeurosis[5] and his subsequent paper with Chung in 1983 the author duo documented melanocytic phenotype of the tumor and the term Malignant Melanoma of soft parts.[6] This Clear Cell Sarcoma occurs most commonly in adolescents and young adults with no particular gender bias;[5,7] But some reports have found predominance in females;[8] It commonly occurs in the deep soft tissue juxtaposed to tendons, fascia or aponeuroses and mostly present as slow growing, frequently painful (30-60%) tumor. Though foot and ankle are frequently involved, the other rare sites involved are kidney, trunk, Penis, GIT, head and neck.[7] The tumor size is relatively small ranging from 0.4 cm up to 14.5cm diameter.[7] Histological picture of Clear Cell Sarcoma in our case showed classically compact nests and fascicles with uniform to minimally pleomorphic tumor delineated by fibrous septa and cells containing polygonal to spindle shaped with abundant clear to pale eosinophilic cytoplasm with round to ovoid vesicular nuclei, prominent nucleoli and some containing cytoplasmic pigments. Specific IHC markers were used to delineate clear cell sarcoma from other entities; More specifically, S100 and HMB45, BCL2, EMA, SMA, MIC_2 were used to differentiate from synovial sarcoma - monophasic type, MPNST, and other epithelial tumors. In our study clear cell sarcoma showed S100, HMB45 and BCL2 Positivity. The antigen for neural differentiation used is S100[11]. In MPNST S100 protein shows only focal weak staining and limited to only to a small number of cells and it is rarely to encounter an MPNST with strong diffuse positivity[11]; this helped to conclude Clear Cell Sarcoma which showed diffuse positivity for S100 in our case. Synovial sarcoma monophasic type is ruled out by EMA and MIC2 negativity[11]. Usually this tumor express strong positivity for EMA and focal positivity for S100, In synovial sarcoma MIC2 gene is expressed in the cytoplasm or cell membrane[11] but in our study in clear cell sarcoma it showed negativity. Melanocytic tumors show HMB45 positivity and a high Ki-67 favours

malignant melanoma for diagnosis[12]. Though our case showed HMB45 positivity, Ki67only 0-1% positivity, S100 positivity and deeper location, without epidermal connections, ruled out clearly malignant melanoma. Metastatic melanoma, other remote possibility is also ruled out by IHC features and also by the negative history of malignant melanoma elsewhere in the body. To distinguish Clear Cell Sarcoma accurately form Malignant Melanoma authors have outlined the histologic criteria that most often Clear Cell Sarcoma is characterized by hyaline sclerotic and reticulated stroma with fascicles of uniform population of tumor cells encased by delicate fibrous septa, which is seldom observed in Malignant Melanoma; The pagetoid pattern of atypical melanocytes in Malignant Melanoma is totally absent in Clear Cell Sarcoma.[9] So, with this histopathological picture, though we thought of Clear Cell Sarcoma, other entities like synovial sarcoma – monophasic type, MPNST, epithelial tumors, if any, had to be ruled out. The histopathological picture did not show neural differentiation; IHC with S100 protein, HMB45 and BCL2 showed strong diffuse positivity favours Clear Cell Sarcoma over other entities like synovial sarcoma – monophasic type, MPNST and other epithelial tumors. Recently molecular genetic study shows t (12;22)chromosomal translocation is more specific for Clear Cell Sarcoma,[7,9] but because of technical impossibility, cytogenetic was not studied in our case. A careful histological pattern analysis, coupled with Immunohistochemistry helped us to establish the diagnosis of Clear Cell Sarcoma involving the deeper tissue.

CONCLUSION

Clear Cell Sarcoma in an aggressive unique rare tumor with propensity for local recurrence, early metastasis and overall poor survival. It is diagnosed by histological and Immunohistochemical study. Eventhough it shares many features with melanoma, Clear Cell Sarcoma has a distinct genetic background[7,9,10] which segregated it as a unique tumor of soft tissue. IHC helps by its low positivity for Ki-67 and HPE by the absence of epidermal connection. The tumor involving the deeper soft tissue was diagnosed in our case as Clear Cell Sarcoma based on the typical histological pattern of nests, delineated by fibrous septa and immunohistochemistry showing diffuse positivity for S100, HMB45 and BCL_2. Hence whenever we come across tumor with histopathological and IHC pattern involving deep tissues we should keep in mind always Clear Cell Sarcoma because of its aggressive nature reoccurrence and metastatic potential.

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