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Case Report

Clear cell sarcoma of soft tissue arising in the supraclavicular region: A cytological diagnosis

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ABSTRACT

Introduction: Clear cell sarcoma of soft tissue is a rare malignant neoplasm which constitutes about 1% of soft tissue tumors. It most commonly arises in the distal extremities of young adults. Peak incidence is in the third decade. More commonly seen in women than men.

Case Presentation: A 30 year old female presented with swelling in right supraclavicular region associated with pain since 3 months. CT scan findings revealed irregular and non-enhancing, hypodense lesion measuring 2.5x2x1cm in right supraclavicular region. Cytological features revealed abundant cellularity comprising of atypical cells mainly dispersed singly and arranged in loose cohesive groups. The cytoplasm was abundant, vacuolated at places, pleomorphic round to oval nucleus, placed centrally to eccentrically with prominent nucleoli. Diagnosis of malignant mesenchymal neoplasm was made on the basis of the cytomorphological details. Clear cell sarcoma was included in the differential diagnosis. Cytomorphological diagnosis was confirmed by histological and immunohistochemical diagnosis.

Conclusion: When cytology of a soft tissue tumor shows high cellularity alongwith dispersion and loosely cohesiveness of cells, nuclear pleomorphism and conspicuous nucleoli, diagnosis of clear cell sarcoma may be considered.

1. Introduction

Clear cell sarcoma of soft tissue (CCSST) is a rare tumor that originates from neural crest cells. It is histologically characterized by clear cells with intracellular glycogen accumulation.[1–3] They commonly arise from tendinous sheaths and aponeuroses, with the majority located in the lower limbs, particularly around the ankles.[4] While less cases are seen describing tumor in the upper extremities and very few cases of primary CCSST have been described in the literature as arising in the chest wall and scapular soft tissues.[5 6] Due to the histochemical similarities, Clear cell sarcoma(CCS) was initially described as "malignant melanoma of soft parts". Immunohistochemical markers similarity and gene expression profiling are suggestive of CCSST as a genomic subtype of malignant melanoma.[7] Most clear cell sarcomas are associated with a t (12;22) (q13-14;q12) translocation.[8,9]

CCS may occur at any age but only a limited number of reports have been published revealing the incidence risk of CCS over the age of 40 years. This tumor shows slightly female preponderance.[10]

Clinically, CCS follows a slow yet progressive course which is characterized by frequent local recurrences alongwith lymph node metastasis. Visceral metastasis is also seen.[11]

The primary suspicion of CCS can be achieved by imaging studies. Exact diagnosis is made finally using

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histological assessment.[10]

The cytodiagnosis of CCS requires a high degree of suspicion, due to its rarity of presentation and overlap of morphology with malignant melanoma, alveolar soft part sarcoma, metastatic adenocarcinoma and, extra renal rhabdoid tumor, synovial sarcoma and epitheloid sarcoma.[9]

Here, we report an unusual case of clear cell sarcoma arising from supraclavicular region in a 30year female.

2. Case Presentation

2.1. Clinical history

A 30year old female presented with complaint of swelling in right supraclavicular region associated with pain since 4months. On examination, there was a tender, soft to firm, irregular mass measuring approximately 2x2cm in right supraclavicular region. CT scan findings showed an irregular, non-enhancing 2.5x2x1cm hypodense lesion in right supraclavicular region.

2.2. Cytological findings

Fine needle aspiration (FNA) procedure of the swelling was carried out using a 23gauge needle and 10ml syringe. Air dried and 95% ethanol fixed smears were prepared and stained by Leishman stain and Papanicolaou respectively.

Smears examined were highly cellular comprising of atypical cells mainly dispersed singly and arranged in loose cohesive groups alongwith lymphoid cells and plasma cells (Figure 1) against a background of amorphous granular debris and few RBCs. The atypical cells had high N:C ratio, central to eccentrically placed, round to oval pleomorphic hyperchromatic nuclei, prominent nucleoli and moderate to abundant amount cytoplasm which was vacuolated at places (Figure 2).

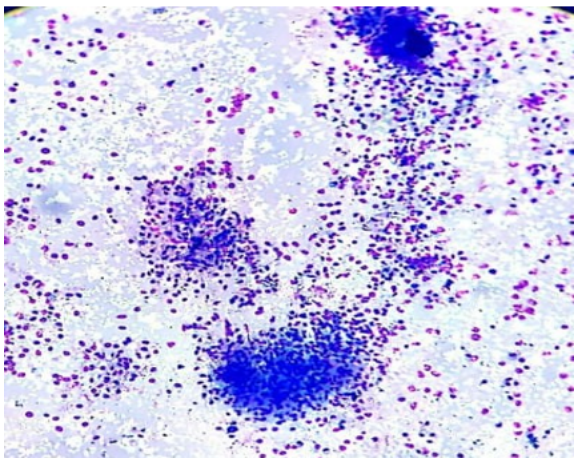

Fig. 1: Atypical cells mainly dispersed singly and in loose cohesive groups admixed with lymphoid cells and plasma cells. Leishman stain X10

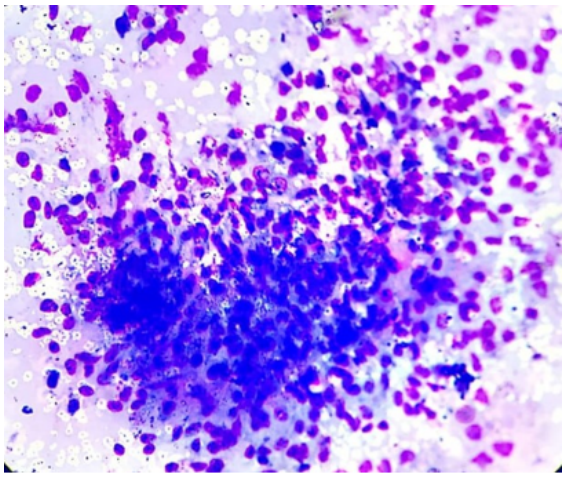

Fig. 2: Atypical cells have high N:C ratio, round to oval pleomorphic nuclei with moderate to abundant cytoplasm. Leishman stain X40.

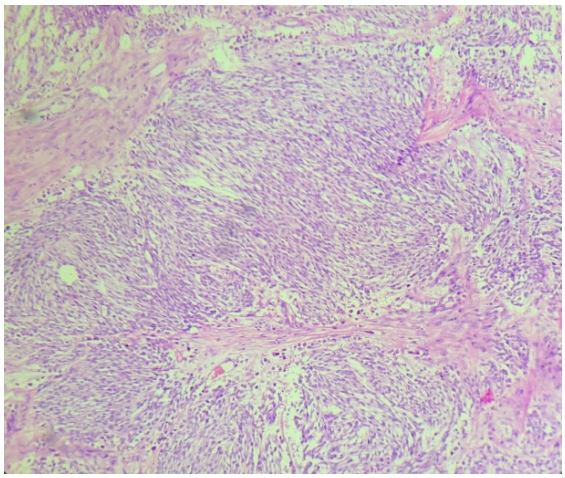

Fig. 3: Tumor cells arranged in nest and fascicles Separated by fibrous septa. H&E stain X10.

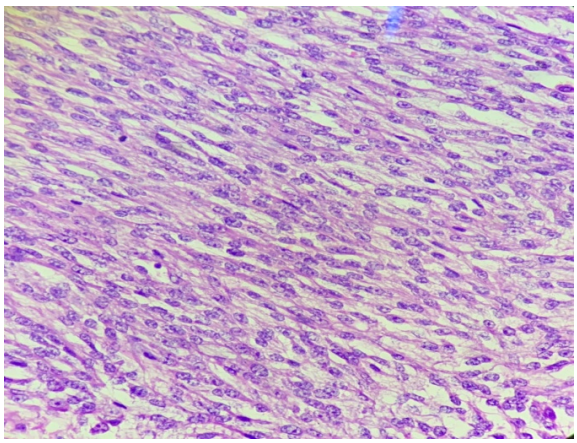

Fig. 4: Round to spindle shaped cells with large Pleomorphic nuclei.

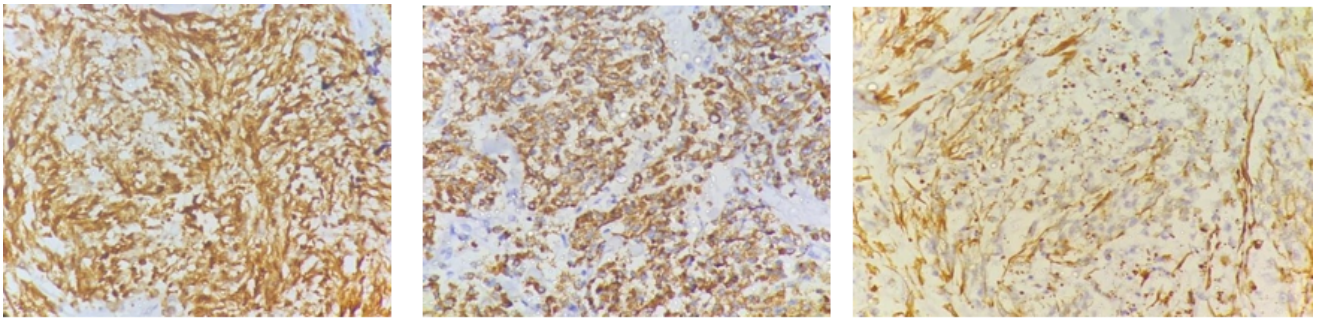

Fig. 5: Immunohistochemistry

Considering cytomorphological details, clinical history, age of the patient, location of the tumor, a diagnosis of malignant mesenchymal neoplasm was made including the differential diagnosis of CCS.

2.3. Histopathological findings

On gross examination, the specimen was partially skin covered globular soft tissue mass measuring approximately 3x3x2cm. The cut surface showed a grey-white, well circumscribed, firm and fleshy tumor.

Representative microsections examined show tumor cells arranged in nests and fascicles which were separated by fibrous septa (Figure 3). Tumor cells were round to spindle with large pleomorphic nuclei, fine granular chromatin, conspicuous nucleoli and eosinophilic to clear cytoplasm (Figure 4). The tumor cells were infiltrating in the surrounding fibroadipose and fibrocollagenous tissue. Mitosis of 18/10 hpf was seen.

The nesting pattern was illustrated on the special stain, reticulin. Fontana Masson stain performed for melanin pigment was negative.

The histopathological findings were suggestive of CCS. The diagnosis was confirmed by imunohistochemistry. The tumor cells were positive for S100, Vimentin and HMB-45.

3. Discussion

Clear cell sarcoma is a slowly progressing malignant tumor mainly arising in the soft tissue of lower extremities. It is more common in women and mainly affect young adults between third and fourth decade.[9]

The common sites of the neoplasm are mainly in the extremities, especially the foot and ankle region followed by the knee, thigh and hand. Few cases mention unusual sites involving lung, chest wall, scapula, cervical spinal cord, rectus abdominis and retroperitoneum.[10]

There are few cytological publications of CCS. The features reveal abundant cellularity comprising of polygonal cells mainly dispersed singly and in clusters with centrally to eccentrically placed hyperchromatic nuclei and abundant clear to finely granular cytoplasm.[10]

On the basis of cytomorphological details, age of the patient, site and presentation of tumor, differentials of epithelioid sarcoma, metastatic adenocarcinoma and alveolar soft part sarcoma can be considered.[11]

On IHC, tumor cells express S-100 and HMB-45.[10] In the present case, tumor cells were positive for S-100, HMB-45 and Vimentin.

These tumors have poor prognosis with increased incidence of recurrence and metastasis. Necrosis and tumor size (>5 cms) factors are associated with a high rate of distant failure.[12] Patients with tumors arising in the extremities have favourable prognosis.[10].

The treatment plan for CCS include surgery followed by chemotherapy.[10]

4. Conclusion

Clear cell sarcoma arising in the supraclavicular region is rare. Recognition of CCS on cytology is difficult although cytological features revealing high cellularity with discrete polygonal pleomorphic cells with granular to vacuolated cytoplasm can guide towards the diagnosis. However, for confirmation, histopatholgical and immunohistochemical assessment is mandatory.

5. Conflict of Interest

None.

6. Source of Funding

None.

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