Case Report

Cytology and histopathology of an unusual neck mass: Extraskeletal myxoid chondrosarcoma

Pooja Chauhan, Neelam Sharma, Lalita Negi, Sudershan Kumar Sharma

Department of Pathology, Indira Gandhi Medical College, Shimla, Himachal Pradesh, India

ABSTRACT

Extraskeletal myxoid chondrosarcomas (EMCs) are rare soft tissue tumors with estimated incidence of 2.3% among all soft tissue sarcomas. Most of these tumors occur in the extremities with only a few cases reported in the head and neck region. Histologically, these tumors exhibit a broad spectrum of morphologic features, which make distinction from other neoplasms difficult, particularly when the tumor occurs at an unusual anatomic location. We present a case report of a 55-year-old male with colloid goiter who presented with a recent increase in his long-standing anterior neck swelling. A computed tomography scan demonstrated a cartilaginous lesion in the soft tissue of the neck. Subsequent cytological and histological examination revealed EMC.

Key words: Chondromyxoid stroma, extraskeletal myxoid chondrosarcomas, neck mass

INTRODUCTION

Extraskeletal myxoid chondrosarcoma (EMCs), initially recognized by Stout and Verner in 1953, is a rare soft tissue malignancy.[1] It was defined as a distinct clinicopathologic entity by Enzinger and Shiraki in 1972 and was called chordoid sarcoma because it resembles chordoma histologically.[2] However, because of the close resemblance of the tumor cells to developing chondroblasts, electron microscopic data suggested that EMCs originates in the primitive cartilage-forming mesenchyme. The tumor has since been called EMC.[3]

EMCs is most commonly located in the deep soft tissues of the lower extremity and the buttock.[4] The present example of EMCs arose in the soft tissue of the neck, a very unusual primary location for this tumor. EMCs behave in a less aggressive manner than do other forms of chondrosarcomas, but their clinical course can vary considerably. Surgery is main-stay treatment; adjuvant radiotherapy or chemotherapy should be considered for cases displaying more cellular pattern and a less conspicuous myxoid matrix.[5]

Address for correspondence: Dr. Pooja Chauhan,
H. No 51, Housing Board Colony, Sanyard, P.O. Talyahar,
Mandi - 175 001, Himachal Pradesh, India.
E-mail: poojaigmc@gmail.com

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CASE REPORT

A 55-year-old male migratory laborer from ambient iodine deficiency zone of Nepal[6] visited the outpatient department with the chief complaint of sudden increase in anterior neck swelling for 1 year associated with progressive dysphagia, dyspnea, stridor, and hoarseness of voice. History of presenting illness revealed that the neck swelling was present for 14 years and diagnosed previously as colloid goiter by fine-needle aspiration cytology (FNAC).

General physical examination was remarkable for increased jugular venous pressure and enlarged cervical lymph nodes.

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A firm, irregular, nontender swelling was observed in the anterior neck more to the right side. Overlying skin was normal with prominent jugular veins [Figure 1]. Thyroid function test, complete hemogram, and biochemical tests were within normal limits. Contrast-enhanced computerized tomography of neck and chest showed 12 cm × 10 cm × 8 cm heterogeneous mass in the soft tissue of neck extending from the angle of the mandible to superior mediastinum internally. It invaded the thyroid cartilage with encasement of brachiocephalic trunk. The patient was referred to our department for FNAC and subsequently underwent incisional biopsy from the neck mass.

FNA showed cellular smears comprised of abundant chondromyxoid stroma with cells present singly and in cords. Tumor cells appearing to be in lacunae had round to oval grooved hyperchromatic nuclei and eosinophilic cytoplasm [Figures 2 and 3]. The possibility of EMCs was suggested. Subsequent incisional biopsy revealed multiple lobules separated by fibrous septa with areas of chondroid differentiation. The individual tumor lobule consisted of the round to slightly elongated tumor cells having mild pleomorphism, hyperchromatic nuclei, and deeply eosinophilic cytoplasm set in a myxoid stroma [Figures 4 and 5]. The diagnosis of EMCs on histopathology supported the cytological diagnosis.

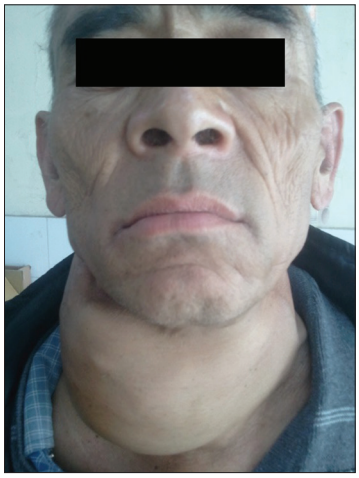

Figure 1: Patient with anterior neck swelling

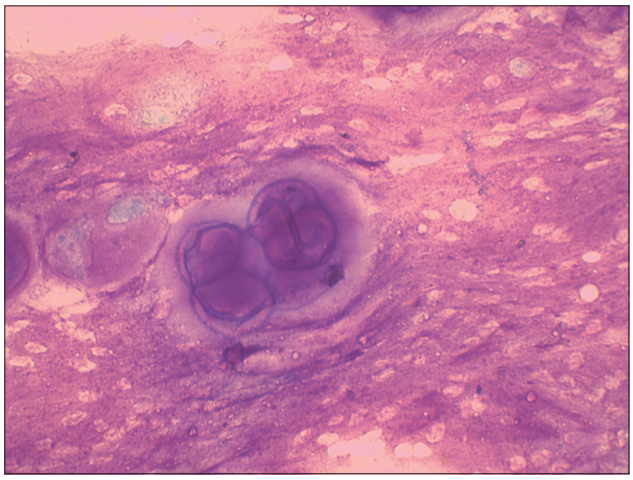

Figure 3: Tumor cells appearing to be in lacunae, ×100

DISCUSSION

EMCs is an unusual soft tissue sarcoma with ultrastructural and molecular features distinct from that of skeletal myxoid chondrosarcoma.[2] EMCs typically affects patients in the fifth and sixth decade of life; however, cases have been reported in children.[7] This tumor arises principally in the proximal extremities and limb girdles.[8] However, occurrences in synovium, pleura, maxillary sinus, epiglottis, soft tissue of the chin, and retroperitoneum have been reported.[1]

Diagnostic cytomorphologic criteria for EMCs have been described in the literature among few case reports and

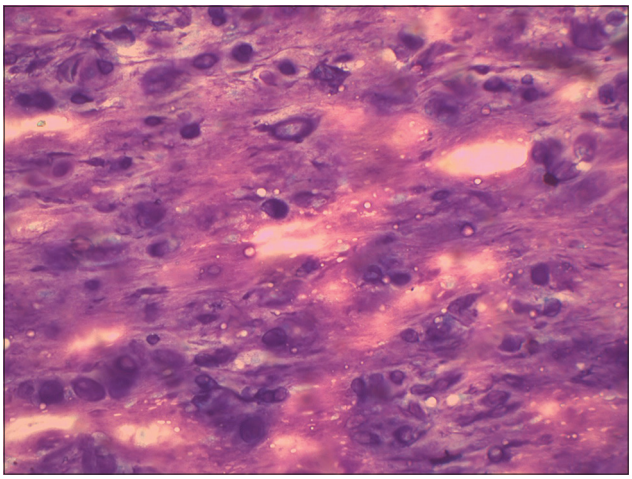

Figure 2: Low power view showing cellular smears with abundant chondromyxoid stroma

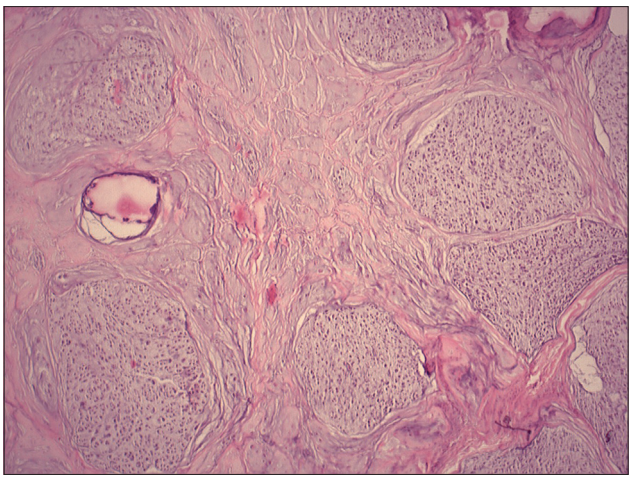

Figure 4: Low-power view of an extraskeletal myxoid chondrosarcoma with a characteristic nodular arrangement

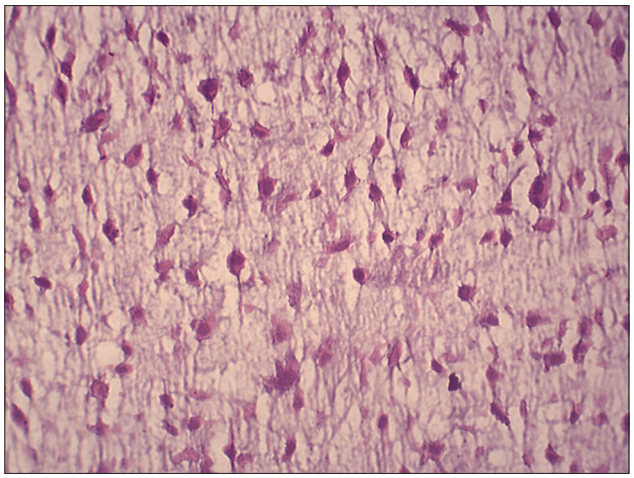

Figure 5: High-power view of tumour cells in extraskeletal myxoid chondrosarcoma

small case series. Common features include a background myxoid matrix with embedded tumor cells, anastomosing cords and lace-like arrangements or clusters of the uniform round to spindle-shaped cells with bland appearing nuclei and inconspicuous nucleoli, and grooved or cleaved nuclei suggesting chondroblast-like derivation; all of which were identified in our case. Other cell types have been reported to occur but only in rare cases, including epithelioid cells with vesicular nuclei, prominent nucleoli, and abundant cytoplasm.[9]

On histological examination, EMC has a multinodular pattern consisting of round or slightly elongated cells of uniform shape and size, in short anastomosing cords or strands, and less frequently in small, loose aggregates set in variable amounts of mucoid material. The individual cells have small hyperchromatic nuclei and a narrow rim of deeply eosinophilic cytoplasm, features characteristic of chondroblasts. Unlike chondrosarcoma of bone, differentiated cartilage cells with distinct lacunae are rare.[8]

EMCs may be difficult to distinguish from a number of benign or malignant chondroid-like or myxoid lesions. EMCs may easily be misdiagnosed as the myxoid variant of some other soft tissue sarcomas. Pathological considerations in the differential diagnosis of EMCs should include chondromyxoid fibroma, juxtacortical (parosteal) chondrosarcomas, chordoma, and myxoid liposarcoma, as well as predominantly myxoid lesions such as aggressive angiomyxoma, low-grade myxofibrosarcoma, and low-grade fibromyxoid sarcoma.[8]

The t(9;22)(q22;q12) translocation appears to be pathognomonic for EMC and has not been found in other mesenchymal tumors.[10] In terms of immunohistochemistry, the tumor cells are positive to vimentin.[11] The expression of S-100, the epithelial membrane antigen and neurofilament have been found to be inconstantly positive while synaptophysin positivity has also been reported.[12]

These low-grade sarcomas have an estimated 10-year survival of 70%, 48% local recurrence and 46% metastases.[1] Metastatic lesions spread hematogenously, with a predilection for the lungs.[13]

The treatment of patients with localized EMCs should include excision of the primary tumor with a wide surgical margin. EMCs have been reported to respond to high-dose radiation while being poorly responsive to adjuvant chemotherapy. When a wide margin cannot be obtained, the benefits of marginal surgery with adjuvant radiation must be weighed against the risk of recurrent disease, given the relative resistance of this tumor to radiation therapy. Amputation should be considered more strongly in such patients.[1] However, in our case surgery was deemed impossible because of extensive tissue involvement. Furthermore, patient, a migratory laborer was lost to follow-up.

CONCLUSION

EMC itself is a rare sarcoma with predilection for the limbs but may present with diagnostic challenge when present in unusual anatomic location. Detection of the EWS-CHN gene fusion resulting from the t(9;22) is a helpful diagnostic tool because it is positive in at least 75% of cases. Even more rarely it may originate in the region of the neck. The only satisfactory chance of cure of these tumors is early diagnosis and surgical intervention in patients with the localized disease and long-term follow-up.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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