

## Case Report

# Extraskelatal myxoid chondrosarcoma of thigh: a rare case report

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### ABSTRACT

Extraskelatal myxoid chondrosarcoma (EMC) are a rare entity of soft tissue tumors that occur predominantly in soft tissue of lower extremities. Here we present a case of 45-year-old female presented with left thigh swelling. MRI finding suggested primary neoplastic lesion. Fine needle aspiration cytology (FNAC) suggested myxoid soft tissue neoplasm/sarcoma. Morphological examination revealed typical extraskelatal myxoid chondrosarcoma with strong immunoreactivity for vimentin and focal immunoreactivity for epithelial membrane antigen (EMA).

**Keywords:** Extra-skelatal myxoid chondrosarcoma, Chondrosarcoma, Thigh

### INTRODUCTION

Extraskelatal myxoid chondrosarcoma (EMC) is rare soft tissue sarcoma that was first describe by Stout and Verner in 1953; however, it was not until 1972 that EMC was histopathologically defined as its own entity by Enzinger and Shiraki.<sup>1</sup> Because there is paucity of convincing evidences of cartilaginous differentiation, EMC is categorized as tumour of uncertain differentiation by World Health Organization (WHO).<sup>2</sup>

EMC has been known for its well distinguished histological, immunohistochemistry and cytogenetic features. The tumor has a slow growth curve but it does carry a high risk of metastasis.<sup>3</sup>

### CASE REPORT

A 45-year-old female presented with left thigh swelling and difficulty in walking since 6 years. On examination, approximately 15×20 cm<sup>2</sup> sized swelling was present over posterior and postero-lateral aspect on mid and lower thigh on left side. Swelling was gradually increasing in size, non-tender, fixed and firm to hard in consistency. Patient had history of trauma to same side.

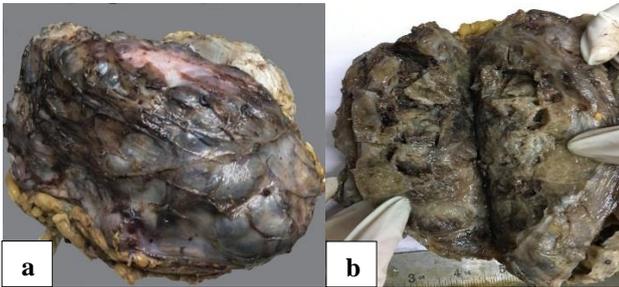
On magnetic resonance imaging (MRI), findings represent primary neoplastic lesion. Fine needle aspiration cytology (FNAC) of left thigh swelling was done and findings were suggestive of myxoid soft tissue neoplasm/sarcoma.

Excision was performed, we received well encapsulated soft tissue specimen of left thigh mass which measured 20×18×11 cm<sup>3</sup> in size. External surface was glistening and multinodular. Tumour was limited by capsule. On serial cutting, multiple solid and cystic areas were seen with abundant myxoid material. Areas of haemorrhage and necrosis were also present (Figure 1a and b).

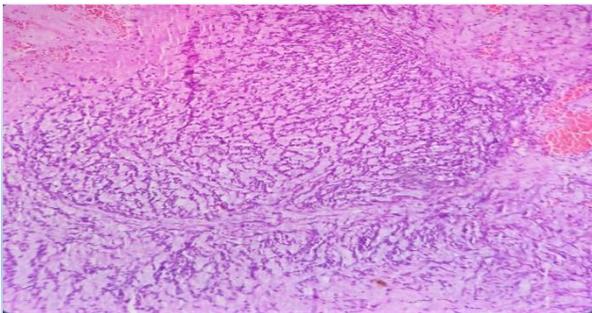
Histologically examination revealed multiple nodules containing tumour cells embedded in abundant myxoid and chondromyxoid matrix separated by fibrous septa. Tumour cells predominately arranged in cords, strands and pseudoacinar pattern. Tumour cells were round/spindle in shape, having high N: C ratio, hyperchromatic nuclei, inconspicuous nucleoli, and scant amount of eosinophilic cytoplasm. Areas of haemorrhage, vascular proliferation and occasional necrosis were seen (Figure 2-6).

Immunohistochemistry showed that tumour cells were immunoreactive for vimentin and focally positive for epithelial membrane antigen (EMA) (Figure 7).

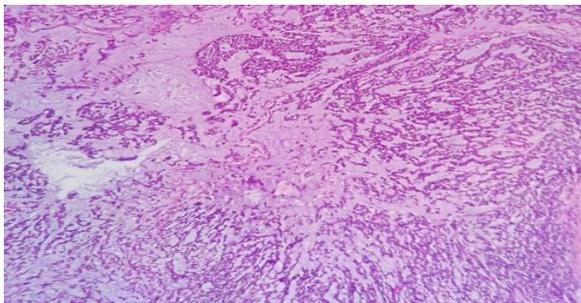
On the basis of morphology and Immunohistochemical features, histopathological diagnosis was extra skeletal myxoid chondrosarcoma, French federation of cancer centers sarcoma group (FNCLCC) grade II (2+2+1=5).



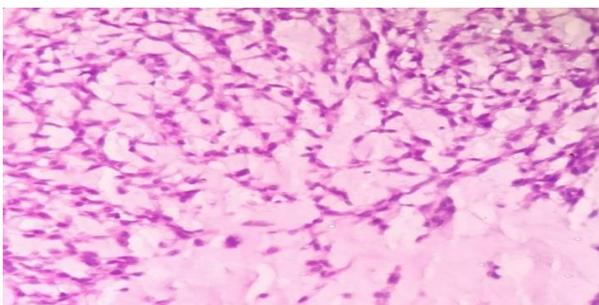
**Figure 1:** a) Encapsulated mass showed external surface glistening and multinodular; and b) cut surface: multiple solid cystic areas were seen with abundant myxoid material.



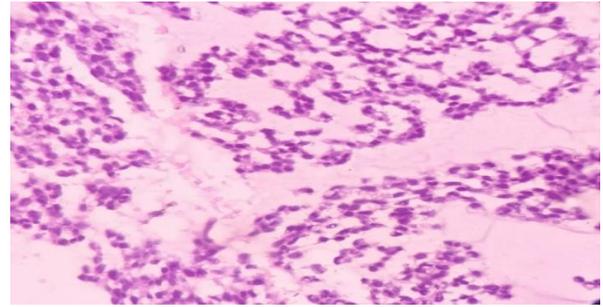
**Figure 2:** Nodular arrangement (H&E, 10X).



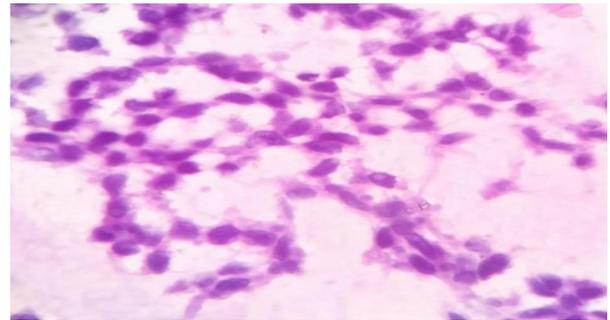
**Figure 3:** Tumour cells arranged in cords and strands, embedded in abundant myxoid matrix (H&E, 10X).



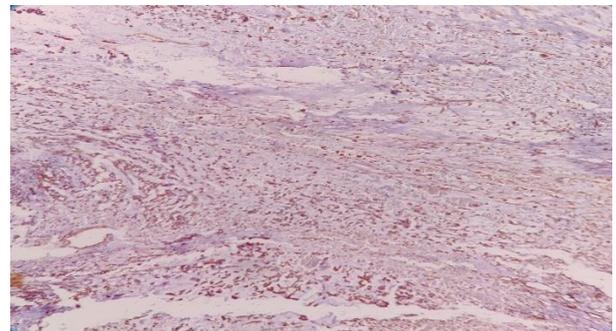
**Figure 4:** Tumour cells arranged in cords and strands with abundant myxoid matrix (H&E, 40X).



**Figure 5:** Tumour cells arranged in pseudoacinar pattern (H&E, 40X).



**Figure 6:** Tumour cells were round/spindle in shape with scant amount of eosinophilic cytoplasm (H&E, 100X).



**Figure 7:** Tumour cells were immunoreactive for vimentin (10X).

## DISCUSSION

EMC are rare soft tissue tumour and represent less than 3% of all soft tissue sarcomas.<sup>3</sup> It occurs throughout the adulthood, with peak in 5<sup>th</sup> or 6<sup>th</sup> decade. Male individuals are affected more commonly than female individuals (male to female ratio is 2:1).<sup>4</sup> The tumour has high rates of metastasis (approximately 50%), although characteristic findings are long term survival after diagnosis, even in the presence of metastasis. Most frequent sites of metastasis are lung, lymphnodes, bone and soft tissue sites.<sup>5</sup> EMC primarily occur deep in extremities especially skeletal muscle and tendon.<sup>6</sup> Limbs and limb girdles are most commonly affected sites followed by upper extremities-trunk, head, and neck. In 10% cases retroperitoneum, abdominal cavity or pelvis affected.<sup>6</sup>

EMC is a morphologically distinctive neoplasm characterized by multinodular architecture which is separated by fibrous septa.<sup>7</sup> Tumour cells forming interconnecting cords or cluster of chondroblast like cells in abundant myxoid stroma.<sup>8</sup> Characteristically, tumour cells of EMC typically present with eosinophilic granular, frequently vacuolated cytoplasm with round to oval nuclei with inconspicuous nucleoli.<sup>8</sup> The present case showed the classical morphology of EMC, which facilitated the diagnosis.

EMC may show typical, alternating hypercellular and hypocellular areas, and these aspects should be considered in differential diagnosis.<sup>7</sup> As for cases of typical EMC, histological differential diagnosis included in our case is myxoid liposarcoma, myxoma, myxofibrosarcoma, chordoma myxoid form, myoepithelioma, extraskeletal soft tissue chondroma, chondromyxoid fibroma, ossifying fibromyxoid tumour and myxopapillary ependymoma.

Myxoid liposarcoma composed of monomorphic small, spindled cells embedded in a prominent myxoid stroma include thin-walled branching (crow's feet) capillaries, uni- or bivacuolated lipoblasts, and mucin pooling, which are not seen in EMC.<sup>5</sup>

Similar to EMC, myxoma having low cellularity and abundant myxoid matrix. The presences of nuclear hyperchromasia and typical separated nodular growth pattern help in distinguishing EMC from myxoma.<sup>5</sup>

EMC arise in subfascial location where as myxofibrosarcoma most often arise in subcutaneous plane and it contain curvilinear and scattered pleomorphic cells.<sup>5</sup>

Chordoma-myxoid form excluded by its usual location in sacrococcygeal region, base of skull and cervical spine.<sup>8</sup>

Mixed tumour/myoepithelioma shows lobulated architecture with cords of epithelioid or spindled cells embedded in myxoid stroma. However, it shows intratumoral architectural and cytogenetic heterogeneity, where EMC is usually uniform. Myoepithelioma always positive for cytokeratin, EMA and S-100, where EMC rarely positive for S-100 and negative for other markers.<sup>5</sup>

Extraskeletal soft tissue chondroma usually occur in the soft tissues of the hands or feet which is unusual locations for ECM. Chordoma shows lobular architecture and cells are epithelioid with abundant clear to eosinophilic cytoplasm that have a bubbly/vacuolated appearance (physaliphorous cells) in myxoid matrix.<sup>8</sup>

Chondromyxoid fibroma having greater degree of cellular pleomorphism and condensation of the tumor cells underneath a narrow, richly vascularized fibrous band that borders the individual tumour nodule. Presence of multinucleated giant cells and foci of calcification or ossification, features rarely seen in myxoid chondrosarcoma.<sup>8</sup>

Ossifying fibromyxoid tumour is lobulated and is composed cords of uniform small ovoid cells in a variably fibromyxoid stroma and the shell of lamellar bone. However, OFMT generally lacks the abundant myxoid matrix of EMC and lamellar bone.<sup>5</sup>

The cells of myxoid chondrosarcoma stain strongly for vimentin, but this is the only marker that is consistently positive. Majority of these tumors show an absence or only focal staining for S-100 protein, neuron-specific enolase (NSE) and synaptophysin. 30% of cases show scattered cells that are epithelial membrane antigen (EMA) - positive.<sup>8</sup>

Cytogenetically, EMC associated with several chromosomal translocation including t (9:22) (922; q12), t (9, 17) (922: q11), t (9:15) (922; 921). The t (9: 22) (922; q12) is most common translocation fuse EWS R1 with NR4A3.<sup>8</sup> EMC is typically associated with a protracted clinical course, even when metastases develop, hence thought to be best classified as low-grade sarcoma.<sup>9</sup>

## CONCLUSION

EMC tumour behaves different from other sarcoma and also carries a high local recurrence rate and sometimes, metastasis. Characteristic findings of having long term survival even after presence of metastasis, necessitated the reporting of such cases by early histopathological examination to decrease the mortality rate.

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