



Case Report

Extra skeletal myxoid chondrosarcoma: A rare case report

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ARTICLE INFO

Article history:

Received 09-10-2018

Accepted 29-04-2019

Available online 22-11-2019

Keywords:

Chondrosarcoma

Myxoid neoplasm

ABSTRACT

EMC is an uncommon malignant neoplasm. In spite of its distinctive clinicopathological features, its clinical features remain debated.¹ Few cases with EMC did not show any apparent enhancement in enhanced CT scan.² we report a case of 82 years old male, who presented with elbow swelling and was diagnosed clinically as ganglion or bursitis, which turned out to be extra skeletal myxoid chondrosarcoma. This case is reported for its deceptive appearance clinically and for its rarity.

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1. Case Report

1.1. Clinical features

82 year old male patient complains of pain and swelling in right elbow and since 31/2 months. Swelling was progressive in size. On examination swelling of size 3.5x3cm was located in right elbow, tenderness, inflammation and mild restriction of movement noted. Right elbow X-ray was taken which was reported as normal. Lesion was excised and sent for histopathological examination.

1.2. Pathology

1.2.1. Gross

Received grey white cystic soft tissue measuring 3x2.5x1.5 cm. Cut surface shows multiple cystic spaces filled with grey white gelatinous material.

1.2.2. Microscopy

H/E Section showed a neoplasm composed of multiple lobules showing extensive myxoid area with tumor cells arranged in sheets and trabecular pattern having round to oval nuclei showing moderate pleomorphism and

eosinophilic cytoplasm. Increase in mitosis noted. It was reported as extra skeletal myxoid chondrosarcoma.

To confirm following panels of immunohistochemistry was employed which includes S100, Vimentin, NSE, Synaptophysin, CK and EMA. Of which vimentin showed positivity. All other markers were negative.

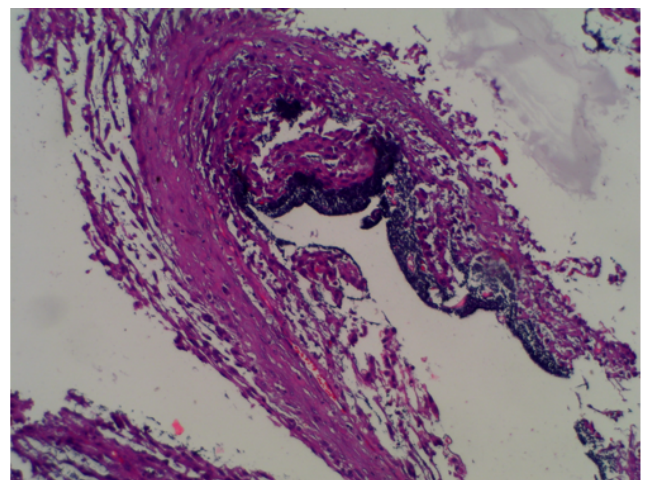


Fig. 1: Section shows a cystic space with tumor cells and myxoid areas (LP)

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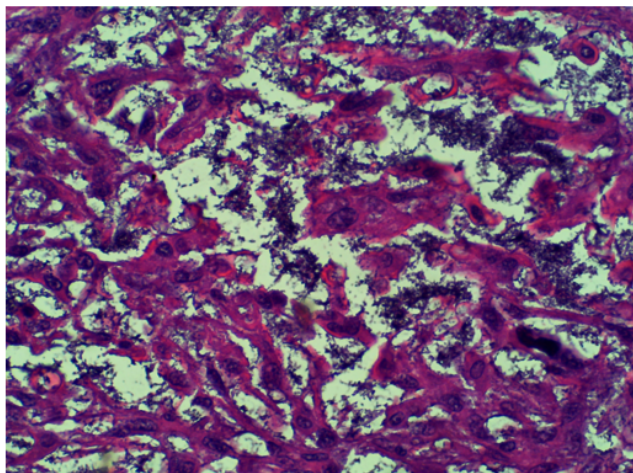


Fig. 2: Tumor cells arranged in sheets with round to oval nuclei showing moderate pleomorphism and eosinophilic cytoplasm

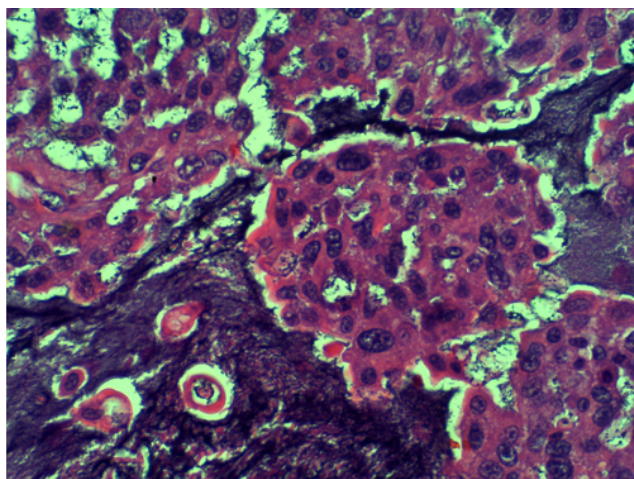


Fig. 3: Section shows tumor cells with extensive myxoid changes

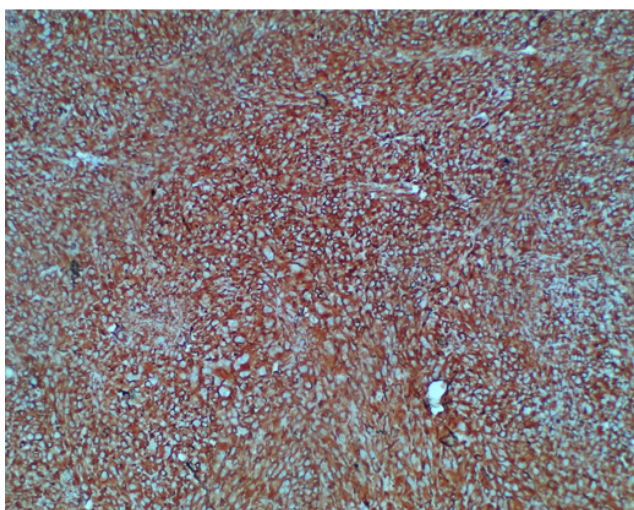


Fig. 4: Section shows strong positivity with vimentin

2. Discussion

EMC is an uncommon soft tissue sarcoma with chondroblastic origin but positivity of neuroendocrine or neural markers suggest likely neuroendocrine or neural differentiation in some Extraskeletal myxoid chondrosarcoma.^{3,4} Regardless of the name there is no substantial indication of cartilaginous differentiation.

It mostly occurs in adults, about 54 years. Males are affected more than female.⁵ It is more common in extremities.³ Occasionally occur in foot, trunk, head and neck and paraspinal, and region.⁶

Clinical features vary depending on their locations, EMC tumors present with nerve compression symptoms.⁷ Swelling, decreased mobility, pain tenderness.⁶

Radiologically shows a soft tissue mass with clear border and even density. The mass will have a lobulated appearance, and in T1WI it is hypoechoic to muscle, but on T2WI hyperechoic, and it is separated into several lobules by numerous septa which appear hypoechoic.⁷

Grossly it is a large mass with pseudocapsule, C/S shows a well-defined multiple nodules with gelatinous areas separated by fibrous septa.

Microscopy shows a multinodular architecture with malignant chondroblast-like cells arranged in clusters or cords in a myxoid matrix. There is no definite cartilaginous differentiation.

The neoplastic cells have moderate eosinophilic, finely granular to vacuolated cytoplasm and uniform round to oval nuclei and inconspicuous nucleoli. Mitosis is usually low.

Cellular variant – shows epithelioid cells with high mitosis and geographic necrosis along with that conventional EMC noted.

It is essential to differentiate EMC from other myxoid tumors and extraskeletal mesenchymal chondrosarcoma. Immunohistochemistry shows reliable positivity for vimentin and shows positivity for S100, CK, EMA focally. Few cases show positivity for NSE, Synaptophysin.⁵

Translocation t (9;22)(q22;q12) consistently has been identified in EMC.^{8,9}

3. Conclusion

EMC shows increased incidence of tumor recurrence and distant spread in spite of a prolonged clinical course.¹⁰ The only therapeutic option for EMC is early wide local resection with or without radiation for localized disease.¹¹ MRI alone cannot distinguish EMC from common extraskeletal chondrosarcoma. Therefore, diagnosing EMC with both CT and MRI scan is suggested.²

4. Source of funding

None.

5. Conflict of interest

None.

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Cite this article: Janaki A, Kavitha A , Sindhura . Extra skeletal myxoid chondrosarcoma: A rare case report. *Indian J Pathol Oncol* 2019;6(4):717-719.