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Case Report

Extraskeletal myxoid chondrosarcoma of the finger mimicking a glomus Tumour: A rare case scenario

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Abstract

Extraskeletal myxoid chondrosarcoma (EMC) is an extremely uncommon soft tissue neoplasm of chondroblastic origin, predominantly seen in middle-aged and elderly individuals and is more prevalent in males. It exhibits a predilection for the deep soft tissue of proximal extremities and limb girdles. It is a rare tumour with distinct histology and a characteristic chromosomal translocation, usually t (9;22) (q22;q12.2), fusing EWSR1 to NR4A3. Despite its nomenclature, EMC exhibits no genuine hyaline cartilaginous differentiation and is classified as a tumour of uncertain differentiation in the WHO classification. Histologically, it exhibits myxoid/reticular histology, necessitating immunohistochemistry to aid in its diagnosis. However, molecular methods like fluorescence in-situ hybridization (FISH) are necessary to obtain a definitive diagnosis. In this instance, we detail an uncommon example of an EMC developing in the finger of a middle-aged female that was clinically thought to be a glomus tumour. Owing to its rare presentation, aggressive nature and tendency to recur, a high index of suspicion by a pathologist is required to diagnose this tricky entity, which can otherwise be readily missed.

Keywords: Cartilagous, Extraosseous, Digits

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1. Introduction

Enzinger and Shiraki first identified extraskeletal myxoid chondrosarcoma (EMC) in 1972 as a low-grade sarcoma consisting of primitive chondroid cells.[1] It is a rare tumour with distinct histology and a characteristic chromosomal translocation, typically t (9;22) (q22;q12.2), fusing EWSR1 to NR4A3. A small proportion of EMCs have t (9;17) (q22;q11.2), which thus results in a RBP56-NR4A3 fusion gene and neuroendocrine differentiation.[2] Median age of presentation is 50 years however rare cases are reported in childhood and adolescence. They usually arise in the deep soft tissue of the proximal extremities and limb girdles, the thigh being the most common site. Less common sites are trunk, head and neck, abdomen, pelvis, etc. Rare sites are finger, cranium, retroperitoneum, pleura and bone.[2] Herein, we describe a case of an EMC arising in the finger of a middle aged female that was clinically thought to be a glomus tumour.

2. Case Details

A 40-year-old female patient came to the surgery OPD with a slowly growing mass in the ring finger of the right hand from the past 8 months. It was quite close to the nail bed, pea-sized, and painful to touch. Based on site and presentation, it was thought to be a Glomus tumour clinically. No radiological investigation was advised.

The mass was excised and sent to us for histopathology. The gross specimen measured 0.8 x 0.7 x 0.5 cm and was slightly fragile. On microscopy, multiple nodular tumour areas of variable cellularity were seen. Most bits were hypocellular and myxoid and comprised of bland-looking spindle cells forming cords and cribriform arrays. Focal areas showed high cellularity consisting of nests of round to ovoid cells with bland-looking nuclei, indistinct nucleoli and a scant amount of eosinophilic to vacuolated cytoplasm. Stroma was hypovascular and mitotic activity was low. No

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obvious cartilaginous or osseous differentiation was noted. No significant atypia or necrosis was seen either. (**Figure 1A-E)**

Based on the histopathology, differentials suggested were Myxoid Glomus Tumour, Cutaneous Myoepithelioma/ Myoepithelial Carcinoma, Extraskeletal Myxoid Chondrosarcoma and Ossifying Fibromyxoid Tumour. A comprehensive IHC panel comprising PanCK, Vimentin, EMA, SOX10, S100, SMA, INSM1 and INI 1 was accordingly advised for ascertaining the exact tumour nature. Only Vimentin and INSM1 were strongly positive, S100 was focally positive and INI1 was retained. Rest all the IHC markers were negative (**Figure 2A-F).** This suggested it to be an Extraskeletal Myxoid Chondrosarcoma. The patient was advised for molecular confirmation by FISH and it came out to be positive. The patient was not given any adjuvant therapy since the tumour size was small, but she has been kept in close and regular follow-up.

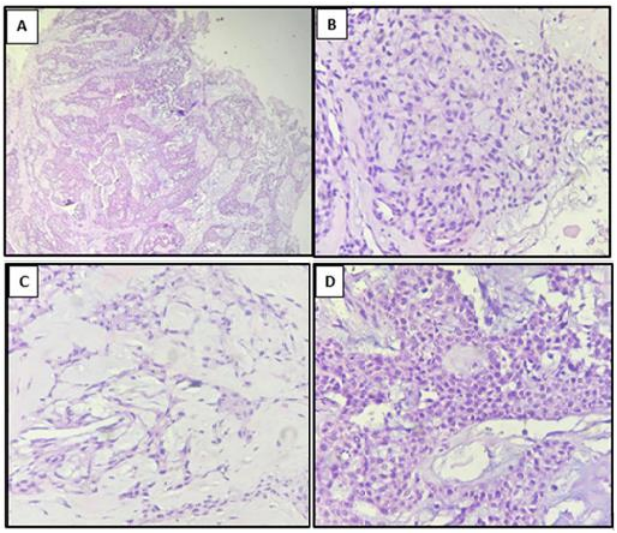

Figure 1: A) Tumor showing alternate hypercellular and hypocellular areas in a lobular tumor separated by dense fibrocellular septa [H&E x 10x]; **B**): Tumor cells were seen assuming a radial pattern of columns, cords or strands rendering a microcystic appearance [H&Ex40x]; **C)**: Hypocellular areas displaying myxoid matrix and spindled cells [H&E x40x]; **D**): Hypercellular areas display nests of round to ovoid tumor cells with bland looking nuclei, indistinct nucleoli and scant amount of eosinophilic to vacuolated cytoplasm. [H&Ex 40x]

3. Discussion

Of all soft tissue sarcoma's EMC are uncommon tumors that account for less than 1%.[2] They are considered malignant mesenchymal neoplasms of uncertain differentiation characterized by multilobular architecture, abundant myxoid matrix, uniform cells arranged in cords, clusters and reticular networks and the very characteristic EWSR1 – NR4A3 rearrangement.[2] The range of size is highly variable, the mean being around 7 cm, and they are often accompanied by pain and tenderness.[4] Because of these features, if present on fingers they can clinically mimic glomus tumours, which was the scenario of the present case too.

On histopathology, the absence of any obvious chondroid differentiation makes it a tricky entity to diagnose and the usual abundance of myxoid areas brings all the myxoid-rich soft tissue tumours in its differentials.[5] In our case, based on the age and site we considered the following differentials:

3.1. Myxoid glomus tumor

These soft tissue tumours originate from the glomus body in the reticular dermis and are most commonly found in the nail unit. Usually, these are well-circumscribed lesions containing capillary-sized vasculature encircled by glomus cells (monomorphous small polygonal cells with pale cytoplasm and typical round nuclei) and thus are straightforward to diagnose by histopathology.[6] However, rare variants such as myxoid and symplastic change, can cause diagnostic difficulty necessitating an IHC confirmation of Pan CK negativity and diffuse SMA positivity.

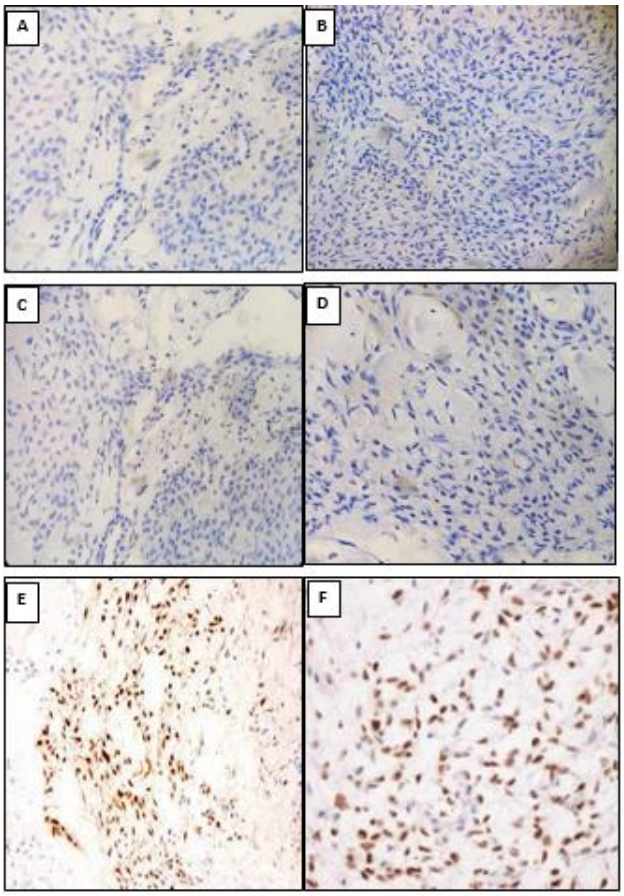

Figure 2: Immunohistochemical analysis- **A:** Pan CK expression negative; **B:** EMA negative; **C:** SOX 10 negative; **D:** S100 negative; **E:** Tumor cells display strong INSM1 expression; **F:** Tumor cells display retained INI expression [all at x40x]

3.2. Cutaneous myoepitheliomas / Myoepithelial carcinomas

These are tumours with only myoepithelial cells and are typically seen in extremities [lower> upper], trunk, head and neck.[2] Presentation age ranges significantly from 10 - 65 years and interestingly carcinomas are more common in children. These are well-circumscribed dermal lesions, composed of ovoid, spindled, plasmacytoid or epithelioid cells in abundant myxoid or hyalinized stroma. 10-15% show cartilaginous/osseous differentiation. EWSR1 gene rearrangement with POU5F1 on 6p21 or PBX1 on 1q23 is commonly seen. On IHC they are Pan CK, S 100, SOX 10 and Calponin positive.

3.3. Ossifying fibromyxoid tumor

These are a unique mesenchymal neoplasm of uncertain differentiation, presenting as lobulated or nodular growth patterns composed of cords, nests, clusters and sheets of uniform ovoid cells embedded in a variable myxoid, fibromyxoid or hyalinized stroma[7] often with an incomplete peripheral shell of bone and can occur in all parts of the body, lower extremities being the most common. In this case, though no bony areas were seen, since the mass was small, we could not rule it out completely. On IHC, these are Pan CK, MUC4, EMA and SMA positive and usually show loss of INI1. On the molecular level, PHF1 gene rearrangement has been observed in 80% of cases.

The IHC results in our case favoured it to be an EMC since these are Pan CK, EMA, SOX 10 negative, diffusely positive for Vimentin, only focally positive for S100 and SMA and usually positive for INSM1. INI 1 is also usually retained except for those EMCs that have rhabdoid features. Literature search suggested that the only confirmatory investigation for EMCs is molecular studies, so we advised FISH for EWSR1-NR4A3 fusion, which came out to be positive.

Compared to other soft tissue, EMC has a higher rate of local recurrence and distant metastasis than other soft tissue tumours, with recurrence rates ranging between 30 and 50% and with distant metastasis reaching upwards of 50%.[2] significantly poorer survival is reported for older age, larger tumour size, non-surgical status, and high tumour grade. Similar to other soft tissue tumours, conventional treatment for primary EMC is wide local excision with or without radiation therapy.[8,9] However, newer studies suggest a valuable role for radiation therapy in improving cancer-specific survival and reducing rates of local recurrence. Studies evaluating the role of chemotherapy in EMC have demonstrated poor response rates, thus relegating its use to a case-by-case approach.[10,11]

4. Conclusion

Extraskeletal myxoid Chondrosarcomas are rare tumors that may mislead pathologists because of their benign-looking cells and the usual absence of cartilaginous differentiation. Hence, knowing about them and keeping them in the differentials of myxoid-rich soft tissue tumors is very important to not miss them because they carry a comparatively worse prognosis.

5. Source of Funding

None.

6. Conflict of Interest

None.

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