CASE REPORT

PRIMARY EXTRASKELETAL MYXOID CHONDROSARCOMA DIAGNOSED BY FINE NEEDLE ASPIRATION CYTOLOGY A CASE REPORT

Mandakini M Patel, Bhavna Gamit, Pinal C Shah, Divya Kriplani
Department of Pathology, Government Medical College, Surat, Gujarat, India

Correspondence to: Pinal C Shah (drpinalshah@gmail.com)

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ABSTRACT
Extraskeletal myxoid chondrosarcoma (EMC) is a rare soft tissue sarcoma. We report one such case where a diagnosis of EMC was made based on fine needle aspiration cytology (FNAC). A 75 year-old male presented to our FNAC clinic with a mass in the right thigh. Magnetic resonance imaging was suggestive of a malignant soft tissue neoplasm and showed no bone involvement. A diagnosis of a myxoid sarcoma favouring an EMC was made. Subsequent excision of the mass followed by histopathological examination and immunohistochemistry confirmed this diagnosis. EMC has distinctive cytological features that are helpful in reaching the diagnosis but it has to be differentiated from other myxoid sarcomas.
Key Words: Extraskeletal Myxoid Chondrosarcoma (EMC); Myxoid Sarcoma; Fine Needle Aspiration Cytology (FNAC)

Introduction

Extraskeletal myxoid chondrosarcoma was identified as a distinct entity for the first time by stout and verner in 1953. Enzinger and Shikari in 1972 defined its clinicopathological features.[1] It is a rare neoplasm which mostly affects older patients with a peak incidence in the fifth and sixth decades, only a few cases have been encountered in children and adolescents. The male to female ratio is 2:1. A majority of tumors occur in the extremities especially the thigh and popliteal fossa.[2] Rarely tumors may occur in other anatomic locations.[3] Late recurrences and metastases are common.[2] We present a case which was diagnosed on fine needle aspiration cytology as a myxoid sarcoma most likely an extraskeletal myxoid chondrosarcoma and later confirmed by histopathological examination.

Case Report

A 75 year old male presented with a huge lobulated mass over the lower part of his right thigh. The mass had been noticed by the patient 8 years back and had gradually increased to its present size and now ulcerated for which the patient seeked medical care. On local examination the swelling was 10 x 10 cm in size and firm in consistency. An MRI scan showed the swelling to be in the intramuscular plane with no bone involvement. FNAC was done using a 22 gauge needle and smears prepared were stained with H&E, MGG and papanicolaou stains. The smears were moderately cellular and showed cells that were predominantly dispersed and some in cords and strands embedded in myxoid stroma.

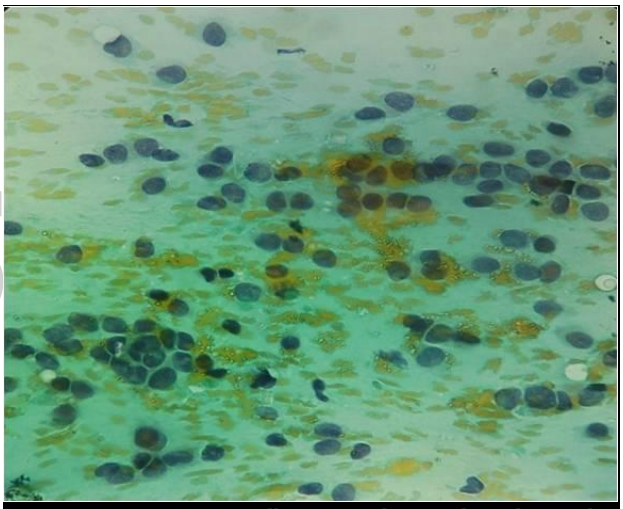

Figure-1: Monotonous cells arranged in cords and strands with a myxoid background.(Papanicolaou stain 400X)

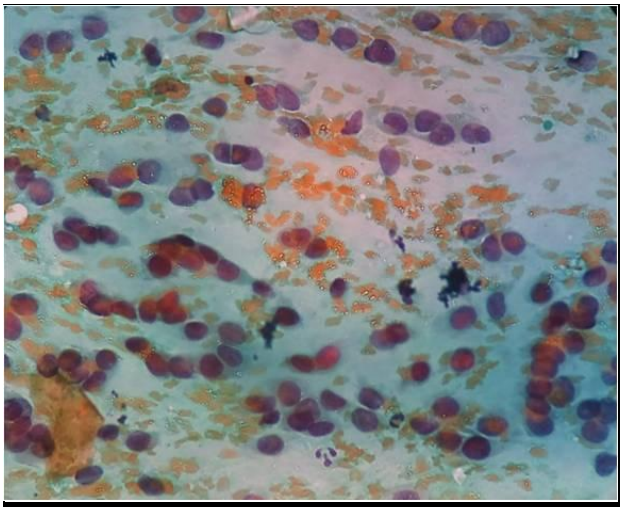

Figure-2: Cords and strands of small uniform cells with hyperchromatic nucleus embedded in myxoid stroma (Papanicolaou stain 400X)

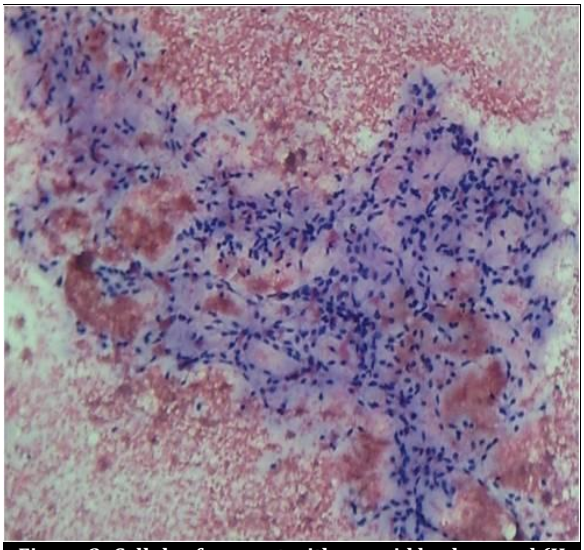

Figure-3: Cellular fragment with myxoid background (H & E 100X)

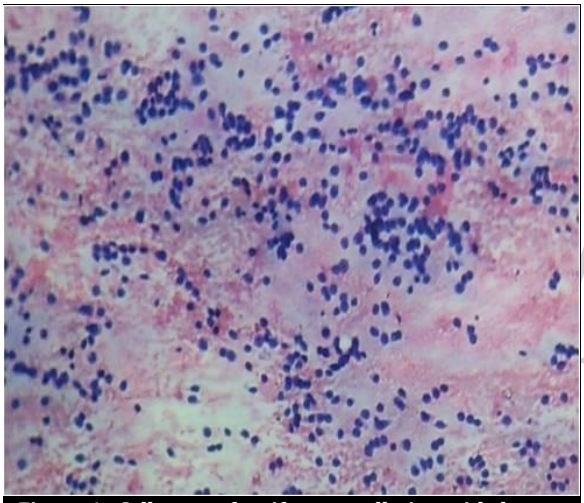

Figure-4: Cells are of uniform small size with hyper-chromatic nuclei dispersed haphazardly and in cords. Background shows blue myxoid stroma (H & E 100X)

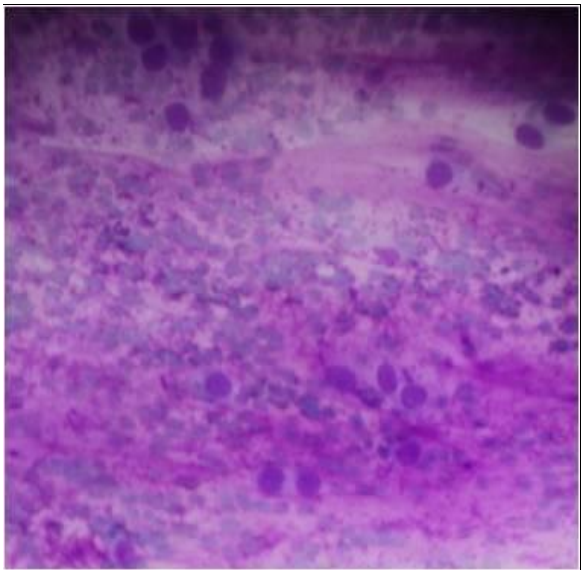

Figure-5: Cells embedded in myxoid material which appears magenta (MGG 400X)

The individual cells were small in size having scant cytoplasm. The nuclei were round to oval, hyperchromatic with small distinct nucleoli at places. Based on the clinical, radiological and cytological findings the diagnosis of EMC was made. Subsequent surgical excision of the mass and Histopathological examination confirmed the diagnosis.

Discussion

ECM is a rare slow growing neoplasm that occurs most commonly in the extremities. Though the term ECM was initially coined by Enzinger and Shikari[3] it has also been adopted by the WHO[4].

The cytological findings in this case were that of cells with small hyperchromatic nuclei mostly scattered and some arranged in a single file pattern, cords and embedded in abundant myxoid stroma. Few cells showed distinct prominent nucleoli. Background showed capillary fragments. Other reports have also described similar findings.[3,5] Other features that have been observed by pathologists are- cartilage like cells with lacunae formation[3], nuclear grooves[5] and intranuclear inclusions[6]. The presence of cytoplasmic vacuolization and cartilaginous differentiation has also been described in a single case report.[6] Mitoses are rare but maybe seen in less well differentiated and more cellular tumours.[2] Secondary changes like fibrosis and haemorrhage are common and the presence of hemosiderin laden macrophages is not an uncommon finding on cytological smears.[5]

EMC does not have any specific immunohistochemical markers. Nearly all cases are vimentin positive. S-100 and EMA show focal and weak immunoreactivity in 20% cases6. Some tumours may also show neuroendocrine differentiation and reactivity for NSE, chromogranin and synaptophysin. Ultrastructural examination of these tumours shows dense core neurosecretory granules. Cytokeratin maybe focally positive in few neoplasms.

EMC shows a balanced translocation t(9:22)(q22;q12). The breakpoint involves the EWSR1 gene on 22q12 and the TEC gene on 9q22. It can be detected with a EWRS1 probe using FISH technique.[6] Large tumor size, high cellularity, anaplasia, high Ki-67 expression, and high mitotic activity seem to be associated with adverse outcomes. EMC has to be differentiated from other myxoid soft tissue sarcomas.[7]

Myxofibrosarcoma the stroma forms a diffuse myxoid granular film in the background and nuclear atypia and pleomorphism are often marked. Myxoid/round cell liposarcoma is characterized by large cells, sometimes epithelioid and the presence of a network of arborizing blood vessels. Multivacoulated lipoblasts with scalloped nucleus maybe seen. Myxoid leiomyosarcoma is a rare diagnosis and is often misdiagnosed as other myxoid sarcomas especially myxoid fibrosarcoma. Skeletal myxoid chondrosarcoma is a more aggressive tumour. Chondromyxoid fibroma may occur in soft tissues due to secondary implantation. It is a highly pleomorphic tumor and shows multinucleated giant cells. Myxoid variant of chordoma is differentiated by its location in the sacrococcygeal region, base of skull, or the cervical spine and the presence of multinucleated, physaliphorous cells. Myxoma is less cellular, having cytologically bland cells that are separated by abundant myxoid stroma and show a dearth of vascular structures. Juxtacortical chondrosarcomas lack myxoid component and show bone involvement radiologically. Mixed tumors of the salivary gland (pleomorphic adenoma) and sweat gland origin (chondroid syringoma). Lesions are mostly superficial in location. The presence of epithelial component along with myxoid helps in reaching the diagnosis.

Conclusion

In the end we conclude that it is feasible to make a correct cytological diagnosis of extracellular myxoid chondrosarcoma based on its distinctive features. However the clinical and radiological data should be in concurrence.

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