Diagnostic role of fine needle aspiration cytology (FNAC) in a case of extraskeletal myxoid chondrosarcoma (EMC)

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Abstract

Introduction: Extraskeletal myxoid chondrosarcoma (EMC) is a rare cartilage tumor comprising 1% to 3% in all soft tissue malignancies. Limited literature available on its cytological features and the diagnostic modality of FNAC. Here, we report a rare case of extraskeletal myxoid chondrosarcoma on FNAC. 40 years female presented with swelling on right waist, rapidly increasing in size and pain since 2 months to present with size of 25x25x10 cm, firm to hard, for which FNAC done. X-ray showed in-homogenous opacity and calcification, bony cortex is intact without erosion. Aspirate showed uniform cells of small to medium size, arranged in chords and clusters embedded in abundant myxoid background. In view of clinical history and radiology, cytological diagnosis of extraskeletal myxoid chondrosarcoma was suggested and confirmed by histopathology. On light microscopy it shows obvious chondroid differentiation, so it needs to be distinguished from other chondromyxoid soft tissue swellings. This case emphasizes diagnostic role of FNAC at rural based hospital devoid of adequate radiological support, immunohistochemistry and other ancillary techniques.

Keywords: Extraskeletal myxoid chondrosarcoma, FNAC.

INTRODUCTION

Extraskeletal myxoid chondrosarcoma (EMC) is a rare soft tissue sarcoma that was recognized as a distinct pathologic entity by Stout and Verner in 1953.1 However, it was not until 1972 that Enzinger and Shiraki2 defined the clinicopathological features of EMC, showing a relatively protracted clinical course and a better prognosis than that with conventional bone chondrosarcoma. Extraskeletal myxoid chondrosarcoma present as deeply located soft tissue tumor in extremities or retroperitoneum and typically affect middle aged patients.3 It is an indolent tumor, however late recurrences and metastasis to lung are common. Tumors with differentiation are known to have aggressive clinical course.4 Since its re-emergence in the latter half of the 20th century, cytopathology and particularly fine needle aspiration (FNA) biopsy-performed cytodiagnostic methods have been increasingly applied to a variety of internal and external anatomic sites.5 Now a day it is very popular and plays a vital role in diagnosis of tumors. Here, in this case fine needle aspiration cytology (FNAC) played diagnostic role.

CASE REPORT

A 40 years female presented with swelling on right waist was referred to our FNAC clinic. It was rapidly increasing in size and pain since 2 months. Initially of 6x5x4 cm in size, to present with size of 25x25x10 cm, firm to hard in consistency. X-ray showed in-homogenous opacity and evidence of calcification, bony cortex is intact without erosion. For which FNA was taken under all aseptic precaution by using 24 gauge needle and aspirate stained with Papanicolaou and May-Grumwald-Giemsa (MGG) stain. In view of clinical history and radiology, cytological diagnosis of extraskeletal myxoid chondrosarcoma was suggested.
Biopsy advised and histopathology confirmed the FNAC diagnosis.

**Cytology Findings**

Highly Cellular smear showed uniform spindle shaped cells of small to medium size, arranged in clusters and chords embedded in abundant chondromyxoid background. Individual cells are having moderate amount of cytoplasm and round ovoid nuclei. At places cells with binucleation, multinucleation, unipolar and bipolar cytoplasmic processes are also evident. In view of clinical history and radiology cytopathological diagnosis of extra skeletal myxoid chondrosarcoma was suggested.

**Histopathology Findings**

H and E stained section showed tumor tissue composed of round uniform cells arranged in lobules and chords separated by myxoid material. Individual cells having hyperchromatic nuclei with eosinophilic cytoplasm, features characteristic of chondroblasts. Background is formed by plenty of myxoid material. At places cells show binucleation. Cytological features are supported by histopathology and final diagnosis of extraskeletal Myxoid Chondrosarcoma was given.

**Figure 1:** Uniform spindle cell of small to medium size are arranged in clusters and chords embedded in abundant myxoid background. (Papanicolaou Stain 10X)

**Figure 2:** Individual cells having round to ovoid nuclei with moderate amount of basophilic cytoplasm. (Papanicolaou Stain 40x)

**Figure 3:** Cells with unipolar or bipolar cytoplasmic processes (Papanicolaou Stain 40x)

**Figure 4:** Abundant chondromyxoid background (Papanicolaou Stain 10x)

**Figure 5:** Metachromatic colour due to chondromyxoid stroma. (MGG Stain 10x)

**Figure 6:** (Histopathology for confirmation) – Histopathologically, cells arranged in lobules and chords separated by myxoid material. (H and E Stain 10x)

**Figure 7:** Clinical presentation - 40 years female with swelling of size 25x25x10 cm

**Figure 8:** X-ray showed in-homogenous opacity and evidence of calcification, bony cortex is intact without erosion
DISCUSSION
Extraskeletal Myxoid Chondrosarcoma is a rare cartilage tumor comprising 1% to 3% in all soft tissue malignancies. The tumor most commonly affects patients older than 35 years and only few cases have been encountered in children and adolescents. Very limited literature available on its cytological features and a diagnostic modality of FNAC. Only few cases of extraskeletal myxoid chondrosarcoma have been published. Originally named chondroid sarcoma, extrasosseous chondrosarcoma was first described in 1953 by Stout and Verner. Extraskeletal myxoid chondrosarcoma histology appears chondroblastic in nature, similar to premature mesenchymal cell rather than mature cartilage. Extraskeletal Myxoid Chondrosarcoma cells remains in an early stage of differentiation, showing evidence of cartilage synthesis at varying stages of chondrogenesis in a matrix of sulfated acid mucopolysacharides similar to chondroitin sulphate. The distinct characteristic cytological features help us in a diagnosis were:

1. Cellularity of smear, uniform spindle cell of small to medium size arranged in clusters and chords.
2. Abundant chondromyxoid background.
3. Individual cells having round to ovoid nuclei with moderate amount of basophilic cytoplasm.
4. at places binucleation, multinucleation, unipolar or bipolar cytoplasmic processes.

First three findings confirm the cartilaginous nature, while the fourth, in the clinical context, indicates malignant behaviour. Most of our findings are similar with other reports and available literature. Our differential diagnosis includes other tumors of soft tissue and bone showing chondromyxoid material and chondroid differentiation on FNAC. It includes chondroblastoma, chondroma, myxoid variant of liposarcoma, chondrosarcoma, chordoma. Chondroblastoma, in most cases found in knee region in epiphysial cartilage of long bone and shows zonal deposits of poorly differentiated chondroblasts and chondrocytes, which may partially calcified and epulis type multinucleated giant cells. In chondroma, synovial tissue of knee and hands are affected mostly. The chondrocytes of chondroma are poorly outlined cells with indistinct cell borders, abundant clear or vacuolated cytoplasm and small pyknotic nuclei. Binucleation and mitotic figures are extremely rare with sclerosed matrix. Myxoid liposarcoma is characterised by chicken wire appearance of fine, interconnecting capillary vessels and monovacuolated lipoblasts. Myxoid stroma in liposarcoma and other myxoid tumours is not metachromatic. Chondrosarcoma has chondroid background and cartilage-like cells embedded within lacunae, where as in EMC chondroblast-like cells lying loose within a myxoid stroma. Chordoma is uncommon malignant neoplasm of axial skeleton typically showing characteristic vacuolated physaliphorous cells. The distinct cytological features of EMC, in the clinical context help to exclude all the differential diagnosis.

CONCLUSION
Cytomorphology of extraskeletal myxoid chondrosarcoma is quite distinct. So, FNAC plays vital role in diagnosis of extraskeletal myxoid chondrosarcoma and is a valuable tool for cytologist to make presumptive diagnosis without ancillary techniques in rural setup where histopathology is the only confirmatory tool.

REFERENCES