Round cell variant of myxoid liposarcoma in a young female at elbow: A case report

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Abstract

Round cell variant of myxoid liposarcoma (MRCLS) is a type of liposarcoma though classified separately (WHO), shares common clinical & morphological features. As compared with other types, this variant has younger onset with peak at 35-55 years with male predominance. It predominately affects limbs particularly thigh & rarely arises from retroperitoneum & subcutaneous tissue. Presence of more round cells in myxoid background (hypercellularity) pointed towards bad prognosis. We are here reporting a case of round cell variant of myxoid liposarcoma in 30 years female, diagnosed in histopathology, composed of uniform spindle cells, round cells with signet ring lipoblasts in prominent background of myxoid stroma with plexiform vasculature, which was negative in the cytology. CT scan shows, soft tissue density mass in the subcutaneous area of the right elbow. The presentation of our case is unique in the regard that, age, sex & the site (extensor surface of elbow), where minimal fatty tissue presents.

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Introduction
Liposarcomas are deep-seated soft tissue tumors in adults that originates in primitive mesenchymal cells.[1] Liposarcomas represent 15% to 20% of all soft tissue sarcomas and out of that myxoid and round cell liposarcoma accounts for 30% to 35%. As compare to other types, this variant has younger onset with peak at 35-55 years. [2] The diagnosis of liposarcoma is based on both clinical and morphologic features. The round cell liposarcoma is a variant of myxoid liposarcoma (MLS) and is a high grade counterpart of it with poorer outcome. Myxoid liposarcoma has marked predilection for lower extremity particularly the thigh with male predominence.

Here we are describing a unique case of round cell variant of myxoid liposarcoma in a 30 years young female arising from right elbow.

Case Report
A 30-year-old female presented with a rapidly growing mass located on her right forearm near the elbow on extensor surface since nine months. The patient had history of trauma a month back followed by rapidly increasing the size. On examination, a subcutaneous huge mass (7 cm diameter), firm, tender, tensely stretched the skin & reddish in colour. (Fig 1) The fine needle aspiration cytology done thrice but no malignant cells seen, except extensive haemorrhagic smear. All the haematological and biochemical investigations were within normal limit except for low HB (6 gm%). Computed tomography (CT) revealed a well-defined, approximately 6x6 cm sized soft tissue density mass in the subcutaneous area on the right forearm near elbow, without significant lymphadenopathy. Excision of lesion done & tissue sent for histopathology. Grossly we received multiple tissues (aggregating 6x5cms) friable, greyish white, attached to skin. Cut section shows area of haemorrhage. (fig 2) Histopathologic examination showed primitive mesenchymal spindle cell, round cell, variable number of mono and multinucleated lipoblasts in myxoid background with occasional area of chicken wire vasculature. (fig 3) Mild pleomorphism, mitotic activity & necrosis also seen, suggestive of round cell variant of myxoid liposarcoma (MRCLS). Patient sent to higher oncology centre for the further management.

Discussion
Soft tissue sarcomas are relatively rare tumor representing only 1% of all human neoplasms.[3] Liposarcoma itself constitutes about 9.8% to 18% of soft tissue sarcomas.[4] The common sites of liposarcoma are the extremities, particularly the thigh, buttocks constitutes 50% of cases. Other sites are retroperitoneal region, perirenal & mesenteric region. Upper extremity involve in only 10% of cases.[5,6] Liposarcoma is a tumor derived from primitive cells that undergoes
Adipose differentiation. In extremities it is slow growing, painless mass, often first noticed after the patient has sustained a minor trauma to the area.\(^7\) In our case patients had preceding history of trauma.

Fig 3: A) Lipoblast with scanty myxoid background. (H&E, 40X); B) Hypercellularity with prominent vascularity. (H&E, 40X); C) Persistence of chicken wire. (H&E, 40X); D) Mono and multivacuolated lipoblasts. (H&E, 40X)

In WHO classification of liposarcomas, Well-differentiated and dedifferentiated liposarcoma comprises the vast majority (approximately 90%) of liposarcoma subtypes while myxoid, round cell and pleomorphic account for less than 10% of liposarcomas.\(^8,9\)

Grossly MLS are well circumscribed, non encapsulated, multinodular, intramuscular, showing glistening, gelatinous or slimy on cut surface in low grade tumors. In high grade it is fleshy tan appearance.\(^9\) Microscopically the characteristic cell is mononucleated and multinucleated lipoblast having fat vacuoles either replacing nucleus aside or with scalloped appearance, along with myxoid stroma and anastomoting capillary network (chicken wire). Cytomorphology of round cell component may resemble that of the relatively small cell seen in myxoid areas. Increased cellularity or appearance of round cell morphology, denotes poorer prognosis. Progression from myxoid towards round cell type is denoted by reduction in myxoid component and vascularity with increase in mitotic activity.\(^11\) The FNAC finding in round cell
variant of myxoid liposarcoma is presence of scanty myxoid matrix and capillaries with round cells and lipoblasts.\textsuperscript{[10]} We have done thrice FNAC but could not get any positive finding except the bloody smear.

As immunohistochemistry is not needed to establish diagnosis of MRCLS, we have not done it .\textsuperscript{[9]} Though the diagnosis can be supported by variable positivity of S100 protein in high grade cases.

Ninety percent of Myxoid round cell liposarcoma lesions have a characteristic chromosomal translocation leading to the fusion of the \textit{DDIT3} and \textit{FUS} genes on chromosome regions 12q13 and 16p11. The consequence is the creation of a FUS-DDIT3 hybrid protein that promotes malignant transformation by dysregulating RNA transcription and thereby dysregulating adipocyte differentiation and cell-cycle control. The diagnosis can be confirmed by evidence of the \textit{DDIT3-FUS} translocation, presence of which is highly sensitive and specific for the diagnosis of MRCL, detected by fluorescence in situ hybridization (FISH) or RT-PCR .\textsuperscript{[9]}

High levels of the round cell component predict a poorer outcome, so it must be determined whether the tumor is more myxoid/low grade (<5% round cell component) or round cell/high grade (>5% round cell component). Another unfavourable predictors are necrosis, TP53 and CDKN2A in localized MLS. Prognosis of the disease depends upon the histological subtypes. Five-year disease specific survival rates are , 100% in well-differentiated liposarcoma, 88% in myxoid liposarcoma, and 56% in pleomorphic liposarcoma.\textsuperscript{[7]}

The primary treatment of liposarcoma is surgical along with radiotherapy whenever needed. The response of treatment by surgical removal is excellent in extremity lesion (13% recurrence), as compare to abdominal lesions (43%).\textsuperscript{[7]}

Conclusion

In liposarcomas when FNAC is negative, one should correlate clinically, radiologically & histopathologically. Histopathology suggest presence or absence of hypercellularity & round cell component, which decides its prognosis. More the cellularity worst the prognosis. Round cell variant of myxoid liposarcoma is not a separate entity as described by WHO, but it is a continuum of one lesion, where round cell variant has poorer outcome than pure myxoid liposarcoma.

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Competing Interests

None declared.

References