Case Report

Myxoid Liposarcoma- A case Report

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ABSTRACT

We present a case of 62 year old female with tumor mass excised from right calf muscle. Histopathology and immunohistochemistry findings are compatible with a diagnosis of Myxoid Liposarcoma. We present a brief review of Myxoid Liposarcoma.

Key words: Soft tissue Sarcoma - ML - Myxoid Liposarcoma; LS - Liposarcoma

INTRODUCTION

Myxoid Liposarcoma make up the major subset of Liposarcoma, which in the most series represent the 2nd and 3rd most common type of soft tissue sarcoma ,ML and its high-grade form together account for around 30-35 % of LS [1]. The incidence is approximately 1/769,000 per year. Myxoid Liposarcoma is the most common subtype of Liposarcoma, occurs predominantly in the extremities of adults and has a tendency either to recur locally or to metastasize to unusual soft tissue locations [2). Myxoid Liposarcoma is an intermediate to high grade tumour.

CASE HISTORY

This is a case of 62 year old female patient who presented with history of swelling over right calf muscle since 03 months, progressively increasing in size. On Gross examination, mass was 12x2 cm, partly skin covered, external surface was multinodular, solid and yellowish in colour. Cut surface was multilobular, solid and yellowish in colour separated by fibrous septae. It was soft to firm, greasy on touch. Areas of hemorrhage and necrosis were absent. (Fig 1)

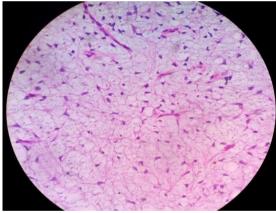
Fig 1: Gross photograph of Myxoid liposarcoma



Histopathological examination revealed, a tumour mass comprised of cells with vacuolated cytoplasm, compressed nuclei and showing nuclear pleomorphism. Lipoblast are noted at places.

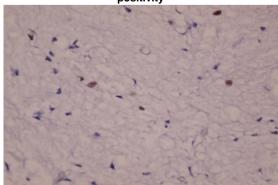
Stroma is Myxoid with thin walled delicate capillaries. Areas of necrosis are also seen at places. The tumour mass was seen invading surrounding muscle tissue. (Fig 2)

Fig 2: H&E Microphotograph of Myxoid liposarcoma



On Immunohistochemistry, it was positive for S - 100 protein. (Fig 3)

Fig 3: Immunohistochemistry showing S – 100 positivity



DISCUSSION

Liposarcoma is the single most common soft tissue sarcoma and accounts for at least 20% of all sarcomas in adulthood. It has three principal forms; well differentiated (of which the dedifferentiated type is a variant), Myxoid (of which the round cell type is a variant) and pleomorphic.

Myxoid tumour mainly affect somewhat younger adults (in the second to fifth decades) than other types. Myxoid Liposarcoma show an overwhelming predilection for the limbs. A tendency for multicentricity in Liposarcoma has been described but this reflects indolent soft tissue metastasis, a characteristic feature of Myxoid subtype and most such patients eventually develop disseminated disease [3].

A history of large long standing mass showing recent rapid growth of a Myxoid lesion usually indicates either dedifferentiation or else progression of a Myxoid lesion to its high grade (round cell) form. Around 5% of case is subcutaneous in location. The cardinal diagnostic feature has traditionally been the presence of lipoblast.

Special stain for lipid and immunohistochemistry generally play no essential role, except that on occasion S - 100 protein staining may highlight multivacuolated Lipoblast and tumour cell in Myxoid Liposarcoma are also often S - 100 positive [4].

MLs are characterized by a specific reciprocal chromosome translocation, either (12:16) (q13: p11) or less often (12:22) (q13:q11-12) both of which result in rearrangement of the DDTT3 gene, a transcription factor involved in adipocyte differentiation [5]. In it low grade from this tumour is gelatinous somewhat reddish and often contains areas of seemingly infarcted more mature adipose tissue [6].

Myxoid liposarcoma can be distinguished from intramuscular myxoma by its greater vascularity and and the presence of lipoblast, from myxofibrosarcoma by the fact that the latter shows notably grater nuclear hyperchromasia and pleomorphism and tend to have more curvilinear vessels. Predominantly round cell lesions may be hard to distinguish from almost any anaplastic round cell malignancy (including metastatic carcinoma), but useful cluesretained even in most high grade cases are the presence of crow's feet vessels and small areas of mucin pooling.

A careful search for Lipoblast usually should have fruitful. It is of interest that, whereas the non -

lipogenic spindled or pleomorphic cells are negative for S - 100 in most types of fatty tumour, the undifferentiated round cells and some of the blander spindle cell in Myxoid and round cell is often show cytoplasmic (and nuclear positivity for S - 100 protein in the same way as adipocytes and Lipoblast [7].

The primary treatment of all types of LS is surgical, in as much as radiation therapy & chemotherapy are generally of only dubious value. In our case patient has undergone above knee amputation because swelling has recurred, again at the ankle joint and was showing similar histomorphological features.

Tumour location, size & histologic subtype are the most important prognostic indicators [8]. Both the pure Myxoid Liposarcoma and the atypical lipomatous tomour (particularly the latter) tend to recur locally rather than to metastasize [9].

By contrast, the pure round cell pleomorphic and dedifferentiated types often give rise to wide spread metastases [10]. In the classic series of Enzinger and Winslow [11], the 5 year survival rate of patients with Myxoid and well differentiated forms exceeded 70% whereas for the round cell pleomorphic verities it was only 18%.

After surgery, patient's condition is good without development of recurrence and metastasis.

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