

Myxoid Liposarcoma with Cartilagenous Differentiation

Hina Tariq¹, Tariq Sarfraz² and Iqra Saeed¹

¹Department of Histopathology, Army Medical College, Rawalpindi, Pakistan

²Department of Pathology, Army Medical College, Rawalpindi, Pakistan

ABSTRACT

Myxoid liposarcoma (MLPS) is an adipocytic tumour and second commonest among the types of liposarcoma. It exhibits a wide histological spectrum. It is important to be aware of this diversity of morphology in this tumour to prevent a wrong diagnosis. Heterologous differentiation in myxoid liposarcoma is reported; however, this is rare.

To date, seven cases of MLPS with cartilagenous differentiation have been reported. Only five of these were further confirmed by cytogenetic analysis. We are reporting the 8th case of MLPS with cartilagenous differentiation.

A 66-year female presented with posterior left thigh mass. Excision was done. Microscopic examination showed sheets of bland spindle cells containing round to oval nuclei and fine, delicate chicken-wire vessels. The firm blue appearing areas, which accounted for about 20% of the tumor volume, were characterised by mature hyaline cartilage. The final diagnosis was MLPS, showing prominent cartilagenous differentiation. Anterior and lateral margins were involved. Tumour was very close to the rest of the resection margins. FNCLCC grade was 1. Provisional pathological stage, as per staging system of the Union for International Cancer Control for extremity, was pT2.

Patient was examined by oncologists. Wide local excision is in plan as two margins were involved by the tumour, while rest of the margins were very close to tumour.

Key Words: *Liposarcoma, Myxoid, Cartilagenous differentiation.*

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INTRODUCTION

Liposarcoma is one of the most common sarcomas of adults. On the basis of microscopic features, it is classified as well differentiated/de-differentiated liposarcoma, myxoid liposarcoma (MLPS), and pleomorphic liposarcoma. MLPS makes up more than half of all reported liposarcomas. Recently, round cell differentiated pattern has been included in MLPS. Majority of patients present in middle age. Out of all liposarcomas, MLPSs occur in adolescents and young adults. It shows predilection for thigh. Grossly, it shows gelatinous surface and exhibits multinodularity.¹ Diagnosis of MLPS is primarily based on the characteristic histology. It is well documented in literature that MLPS exhibits a broad histopathological spectrum. Majority of the cases exhibit classic morphology, *i.e.*, well-demarcated lobulated tumours containing even distribution of relatively bland appearing fusiform cells and lipoblasts in abundant myxoid stroma.²

Diversity of histologic patterns in MLPS can lead to misdiagnosis. Fritchie *et al.* described various microscopic appearances in 46 cases of MLPS.¹ They identified variety of patterns, including chondroid metaplasia in 4% of their total cases.

Other common patterns were classic myxoid, round cell and pseudoacinar. DDIT3 rearrangement was identified in cases with cartilagenous areas. In these cases, mature hyaline cartilage showed a sudden transition to conventional MLPS. Overt malignant features in the cartilagenous foci were not recognised.¹

A total of nine MLPSs with heterologous elements have been described in literature. In addition to chondroid differentiation, osseous and leiomyomatous differentiation have been reported.³ Enzinger and Winslow and Evans had described cartilagenous differentiation as a metaplastic phenomenon. In some studies, distinct distribution of both cartilagenous and myxoid liposarcomatous areas has been reported; while in other cases, gradual transition between the two were documented.⁴⁻⁷

Cartilagenous and rhabdomyoblastic differentiation was reported by Kim *et al.* in a case report.² Until now, hallmark fusion gene has been detected in only five cases. Details of mechanism for production of cartilage in MLPS are still unclear.⁶ Although FUS-DDIT3 fusion has been documented in both the liposarcomatous and cartilagenous areas in MLPS in various studies, there is difference in prognosis and proliferation activity from typical MLPS. These findings point to some other molecular mechanism for production of cartilagenous differentiation. This mechanism will become clearer, as more and more cases are reported.⁸

Round cell component can be found in few cases of MLPS. It is linked to aggressive behaviour. According to latest reports, even 5% of round cell component upgrades the tumour grade.⁹

Correspondence to: Dr. Hina Tariq, Department of Histopathology, Army Medical College, Rawalpindi, Pakistan
E-mail: dr.hina2013@gmail.com

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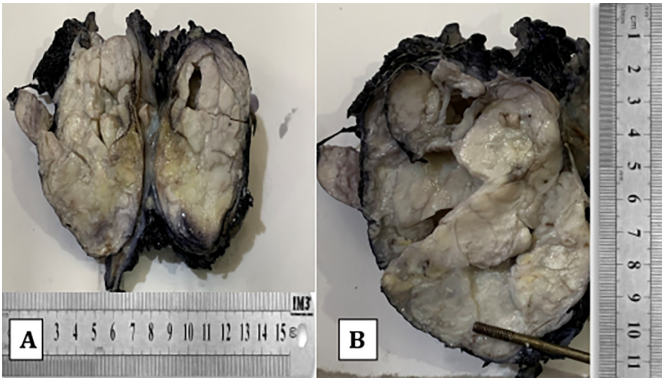


Figure 1: Gross specimen of myxoid liposarcoma. (A) Cutsurface reveals a lobulated solid myxoid surface with yellow white areas. (B) Focus of cartilage pointed by probe.

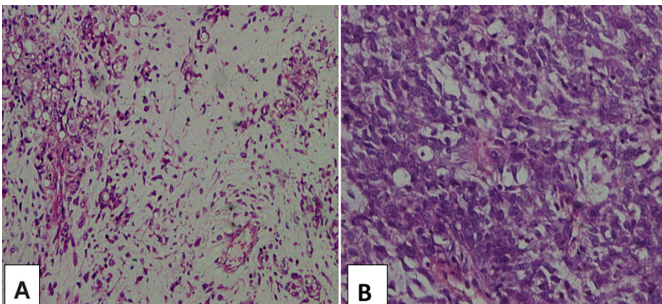


Figure 2: Myxoid liposarcoma. (A) High power (40x) reveal fusiform cells and lipoblasts with few blood vessels. Stroma is myxoid. (B) Myxoid liposarcoma with round cell component. 40x.

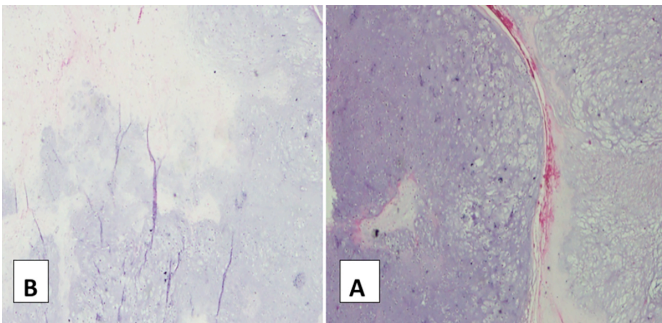


Figure 3: Myxoid liposarcoma with chondroid differentiation. (A) & (B) 4x.

CASE REPORT

A 66-year female presented with left thigh posterior aspect mass. Plain MRI thigh revealed a large, well defined, ovoid, heterogeneously enhancing mass lesion in subcutaneous tissue at posterolateral aspect of left thigh, causing marked anteromedial displacement of left biceps femoris muscle. It measured 9.3 x 4.7 x 2.4 cm. Ultrasound-guided biopsy from left thigh was reported as myxoid spindle cell neoplasm, favour MLPS. Excision was done. The specimen received was an oriented lobulated and encapsulated mass partly covered by fat posteriorly and by thin flap of muscle inferoanteriorly. Cut surfaces showed a solid gelatinous tumour with yellow white areas and hemorrhagic foci (Figure 1). Some areas also showed cartilage and focal calcification. It was reaching the resection margins. Microscopic examination showed sheets of bland spindle cells with round to oval nuclei and fine, delicate chicken-wire vessels, myxoid areas characterised by a myxoid

matrix with individual bland spindle shaped cells and lipoblasts (Figure 2). The firm shiny blue areas, about 20% of the tumour volume, were characterised by mature hyaline cartilage (Figure 3). It appeared as a distinct and different component than typical areas. Less than 5% round cell tumour was noted. The final diagnosis was MLPS showing prominent cartilaginous differentiation. Anterior and lateral margins were involved. Tumour was very close to the rest of the resection margins. FNCLCC grade was 1. Provisional pathological stage, as per staging system of the Union for International Cancer Control for extremity, was pT2.

Patient was examined by oncologists. Wide local excision is in plan as two margins were involved by the tumour, while rest of the margins were very close to tumour.

DISCUSSION

MLPS presents after 4th decade. It characteristically appears as a nodular swelling on thigh. Histologically, most of the tumours contain monomorphic spindle cells and lipoblasts with minimal atypia. In the background, abundant myxoid stroma is present. Chicken-wire capillary vasculature is characteristic of this tumour. Cells with cytoplasmic vacuolations with eccentric appearing nuclei are also seen. Few cases with discrete cartilaginous, leiomyomatous and osseous differentiation in MLPS have been described in literature. Till date, five cases of MLPS with cartilaginous differentiation have been reported. If prior case reports of Enzinger and Winslow and Evans are taken into account, then the total number of cases reach seven. However, they initially described it as a metaplastic phenomenon. Distribution of cartilaginous areas can be distinct from typical myxoid areas or there can be gradual transition between both patterns^{4,6,7} Siebert *et al.* discussed that cartilaginous areas can be misinterpreted as various other soft tissue tumours.⁷ Mature cartilage is rarely reported in MLPSs.⁵ Cytogenetic evaluation of the conventional histology and the cartilaginous area can be used to support the diagnosis.¹ Only few cases of heterologous components have been reported so far and often without molecular studies. Concept of "de-differentiated" MLPS is also still controversial.¹⁰ Distinct translocation t(12; 16) (q11; p11) occurred consistently throughout various reported studies and many reports showed it in more than 90% of the MLPSs.¹

In literature, the quantity of round cell component, tumour necrosis and p53 over expression are well documented prognostic factors.⁴ Importance of round cell component as a prognostic factor was first identified by Enzinger and Winslow, which was further confirmed in follow-up studies.^{4,5} Microscopic details of cartilaginous areas were described in only a small number of cases. So far, cartilage has not been classified as benign or malignant. Ossification along with cartilage formation has also observed in one study and this phenomenon was designated as enchondral ossification rather than as osteosarcomatous differentiation.² Authors of various previous case reports have speculated that progenitor cells might have resulted in generation of heterologous component.¹ Additional studies are required to further evaluate the true origin of cartilage in a MLPS.

CONFLICT OF INTEREST:

No conflict of interest was reported.

AUTHORS' CONTRIBUTION:

HT: Conception of idea, draft writing.

TS: Finalised the draft, critical review.

IS: Worked on idea, finalised the draft.

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