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Lipoma or Liposarcoma? - The Diagnostic Challenges

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Abstract

Atypical lipomatous tumour / well-differentiated liposarcoma (ALT/WDLPS) is a locally aggressive mesenchymal neoplasm composed either entirely or partly of an adipocytic proliferation showing at least focal nuclear atypia in both adipocytes and stromal cells. This case report details a case of a 45-year-old female who presented to OPD with swelling over the medial aspect of the thigh for 3 years. Clinically she was diagnosed with lipoma followed by an excision and histopathological examination. Histopathology revealed features suggestive of Atypical lipomatous tumour / well-differentiated liposarcoma (ALT/WDLPS) which showed the presence of striking variation in adipocyte cell size. This study is done to emphasise the importance of histopathological examination as an important diagnostic tool in differentiating lipoma from liposarcoma.

Keywords: *Atypical Lipomatous Tumour (ALT), Lipoblast, Lipoma, Liposarcoma, Well-Differentiated Liposarcoma (WDLPS).*

Introduction

Liposarcomas account for 20 per cent of all sarcomas. Liposarcoma is a diverse set of distinct lesions that provide many diagnostic challenges. Atypical lipomatous tumours are the most common kind of liposarcoma, accounting for 40-45% of all cases. Its synonym is well-differentiated lipomatous tumour. Atypical lipomatous tumour/well-differentiated liposarcoma (ALT/WDLPS) is a locally aggressive mesenchymal neoplasm made up totally or mostly of adipocytic proliferation with at least localized nuclear atypia in both adipocytes and stromal cells [1].

Its histological subgroups include lipoma-like liposarcoma, inflammatory liposarcoma, and sclerosing liposarcoma [2]. It is most typically seen as a deep-seated lesion in the proximal extremities and trunk. Other typical sites are the retroperitoneum and Para testicular regions [1].

Lipoma-like ALT/WDLPS is made up of mature adipocytes that, unlike benign lipomas, show significant cell size fluctuation as well as nuclear atypia in fat cells or stromal spindle cells. Lipoblasts can be found in variable numbers (from many to none). Importantly, the existence of lipoblasts does not constitute a diagnosis of liposarcoma. As a result, histological testing is critical in determining the diagnosis [1,2]. The MDM2 and CDK4 genes are always amplified. Atypical lipomatous tumour/well-differentiated liposarcoma (ALT/WDLPS) does not metastasize unless it develops dedifferentiation [2].

Case Report

A 45-year-old female came to OPD with complaints of swelling over the medial aspect of the left thigh for 3 3-year duration. Clinically the swelling was evaluated for lipoma and planned for removal by excision. The excised specimen was sent for histopathological examination.

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We received a single grey-yellow fibrofatty mass measuring 12 cm in greater dimension (Fig 1) with soft to firm consistency and its cut surface showed areas of calcification. The tissue was processed and the sections were examined. The examined sections showed a lipomatous tumour with striking variation in adipocyte size (Fig 2) with atypical stromal cells (Fig 3) in a collagenous background (Fig 4). Occasional lipoblast (Fig 5,6) and focal coagulative necrosis (Fig 7) also present.

Scattered chronic inflammatory cells were seen in the background stroma. Areas of dystrophic calcification (Fig 8) and haemorrhage were also noted. It was graded as Grade 1 according to the French Federation of Cancer Centres Sarcoma Group (FNCLCC). Hence it was reported as Grade-1 Atypical lipomatous tumour / well-differentiated liposarcoma (ALT/WDLPS). However, gene amplification of MDM2 and CDK4 is required for confirmation.

Figure 1. Gross Image of Specimen

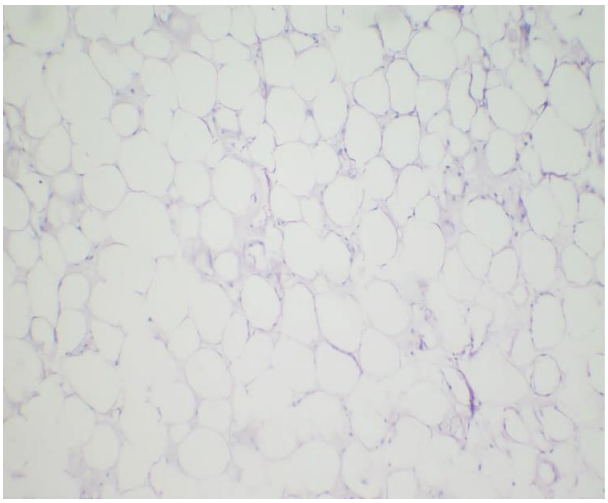

Figure 2. Low Power (10x) Microphotograph Showing Striking Various Size Adipocytes

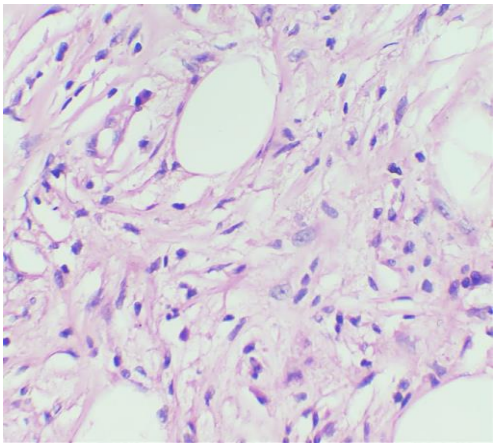

Figure 3. High Power(40x) Microphotograph Showing Atypical Stromal Cells

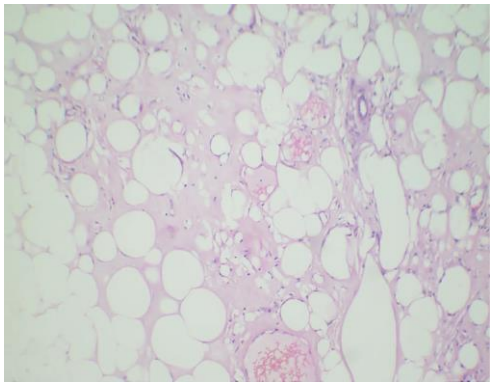

Figure 4. Low Power (10x) Microphotograph Showing Collagenous Background

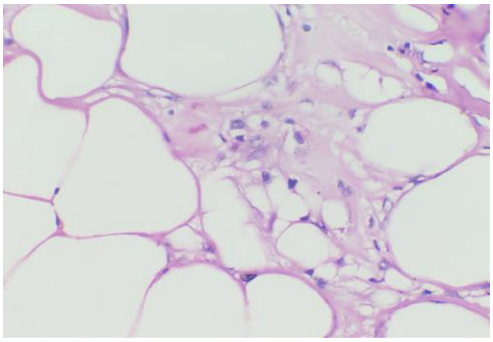

Figure 5. High Power(40x) Microphotograph Showing Lipoblast

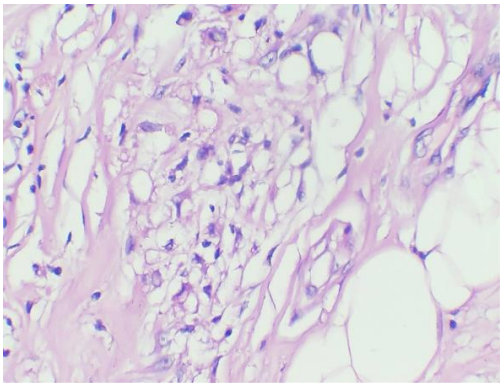

Figure 6. High Power(40x) Microphotograph Showing Lipoblast

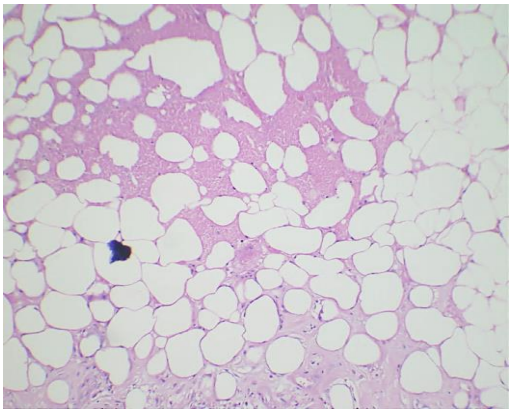

Figure 7. Low Power(10x) Showing Area of Coagulative Necrosis

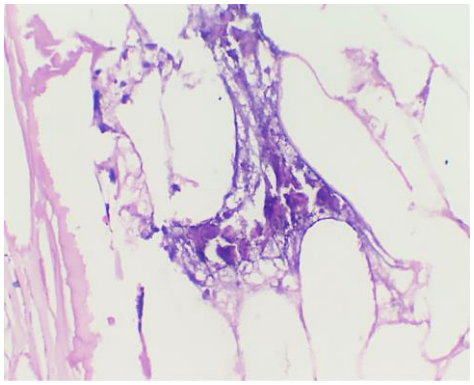

Figure 8. High Power(40x) Showing a Focal Area of Calcification

Discussion

Atypical lipomatous tumour/well-differentiated liposarcoma (ALT/WDLPS) is the most common malignant adipocytic neoplasm, accounting for 40-45% of all liposarcoma cases. It happens in the fourth to fifth decade of life. It is a locally aggressive malignant tumour that affects the retroperitoneum, limbs, Para testicular area, and mediastinum. ALT/WDLPS may be associated with Li-Fraumeni syndrome, however almost all instances are sporadic, and the cause is unclear [2].

The presence of a unique ring and/or large marker chromosomes indicates a genetic abnormality. Ring chromosomes have amplified sequences from the 12q13-15 chromosomal region, which contains multiple proto-oncogenes such as MDM2, CDK4, and HMGA2 [3]. As a result, molecular or immunohistochemical approaches can identify the amplification of HMGA2, MDM2, and CDK4 (as well as overexpression of the respective proteins) in atypical lipomatous tumour/well-differentiated liposarcoma [4].

Hence, in rare complicated situations, identification of MDM2 and CDK4 by FISH is the gold standard for supporting the diagnosis.

ALT generally manifests as a deep-seated, painless lump that can gradually grow to a very big size. ALT often consists of a big, well-defined, lobulated mass. Variable consistencies exist, ranging from hard grey to gelatinous regions, depending on the amount of fibrous and myxoid components. The tumour is made up of mature adipocytes with varying cell sizes and nuclear atypia. Furthermore, as their names suggest, distinct subtypes exhibit morphological differences [5].

Lipoma-like ALT/WDLPS exhibits varying adipocytic sizes that are linked with nuclear atypia in stromal and/or adipocyte cells. Sclerosing ALT/WDLPS is characterized by hyperchromatic bizarre stromal cells placed against a fibrillary sclerotic backdrop. Inflammatory ALT/WDLPS: dispersed atypical

stromal cells in a chronic inflammatory setting. The presence of lipoblasts is not necessary for diagnosis. The appearance of many morphological patterns in a single lesion is typical, particularly in the retroperitoneum [6].

ALT has no potential for metastasis until it undergoes dedifferentiation. The eventual risk of dedifferentiation varies with place and lesional duration. The most significant prognostic factor is anatomical location [7,8]. Lesions found in surgically accessible anatomical locations do not resurface after full excision. Patients with extremity tumours had much higher survival rates than those with retroperitoneal or scrotal malignancies [9].

Conclusion

This case report is done to emphasise the importance of histopathology in distinguishing the liposarcoma from lipoma. Histopathological examination of the specimen aids in diagnosing the liposarcoma as an important diagnostic tool.

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