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Case Report

Atypical lipomatous tumour of the axilla– A case report with histological correlation

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ABSTRACT

Liposarcoma is the most prevalent soft tissue malignant tumour. It mostly arises in the subcutaneous tissue of shoulders, limbs and neck, and retroperitoneal space. Atypical lipomatous tumour (ALT) is a well-differentiated liposarcoma (WDLPS) and constitutes 40–45% of all liposarcoma cases. They are locally invasive mesenchymal soft tissue tumours. Most of them are large in size. They have heterogeneous histomorphology, molecular and genetic characteristics, and clinical prognosis, making the diagnosis and treatment of difficult for surgeons.

A case of 45-year-old woman with a tumour of the right axilla, causing a discreet asymmetry is presented. The tumour was soft and caused no tenderness. The patient reported no previous injury to the region. Fine needle aspiration cytology (FNAC) revealed atypical lipomatous cells suspected of liposarcoma. The patient was operated under general anaesthesia. The postoperative course was uneventful. The final histopathological diagnosis – was atypical lipomatous tumour/well-differentiated liposarcoma (ALT/WDLPS). ALT/WDLPS is a locally invasive tumour with a good prognosis. However delayed treatment is associated with an increase in tumour size, which can affect mobility. Therefore, to avoid possible local recurrence, extensive surgical resection of tumour tissue with wide margins is a desirable treatment for all ALT/WDLPS cases. Besides, for ALT/WDLPS tumours that are difficult to extensively excise, long-term follow-ups are necessary due to the possibility of recurrence.

1. Introduction

Soft tissue sarcoma is a heterogeneous lesion comprising of more than 50 histological subtypes with different biological behaviours. Liposarcoma is a malignant mesenchymal tumour, and it is the most frequent subtype of soft tissue sarcomas. Histologically diverse subtypes of liposarcoma help in prognosticating patient outcomes. The World Health Organization classification of soft tissue and bone tumours identified the following four major liposarcoma subtypes: atypical lipomatous tumour/well-differentiated liposarcoma (ALT/WDLPS), dedifferentiated liposarcoma (DDLPS), myxoid liposarcoma, and pleomorphic liposarcoma. ALT/WDLPS and myxoid liposarcomas are low-grade tumours and have a more indolent clinical course. Dedifferentiated and pleomorphic liposarcomas are highly aggressive tumours with potential for metastasis. Term Atypical lipomatous tumour (ALT) is synonymous with Well-differentiated liposarcoma (WDLPS), and the use of these terms is decided according to the tumour location and resectability.[1] WDLPS occurring in a limb is usually easily treated and does not metastasise, therefore often referred to as ALT. On the other hand, WDLPS occurring in the retroperitoneum have a higher probability of recurrence or dedifferentiation. Thus, the term WDLPS is used to highlight the malignant features.[2] The most frequent sites affected are soft tissues of the limbs, retroperitoneum, thigh, mediastinum, and para-testicular tissues, and subcutaneous tissues.[3] ALT/WDLPS commonly affect middle-aged or older people. It is histologically characterised by lipoblasts in mature fat, atypical stromal cells and, a prominent sclerotic component.[4,5]

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2. Case History

Female aged 45 years presented with a well-defined lump in the right axilla. The tumour measured approximately 8x6 cm, the tumour was soft and caused no tenderness. The patient reported no previous injury to the region. An FNAC was done and smears prepared. FNA showed fragments of mature fat tissue and numerous dispersed, large, hyperchromatic, often with bizarre multinucleated nuclei. A cytological diagnosis of atypical lipomatous tumour / well-differentiated liposarcoma was made.

The patient was operated under general anaesthesia. The postoperative course was uneventful A biopsy of the mass resected was performed and showed grossly multilobulated and well-circumscribed tumour with a marbled yellow cut surface, with focal more firm/fibrotic white areas grossly. Appropriate sections were taken, and microscopic examination revealed mature adipose tissue intermingled with fibrous zones containing hyperchromatic stromal cells with nuclear atypia, many of which were multinucleated [Figure 1]. This collaborated with preoperative cytological findings.

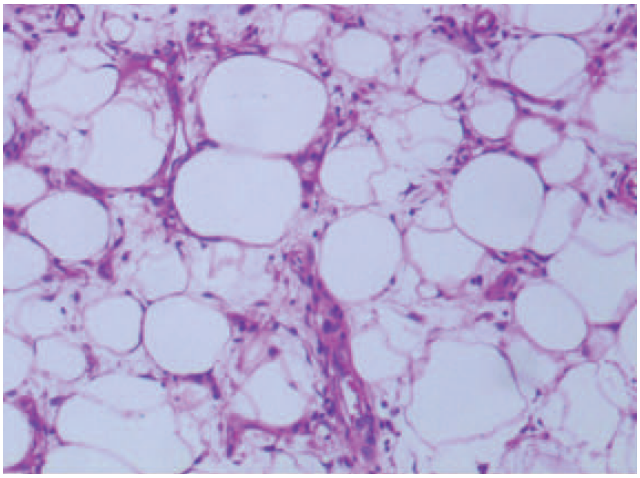

Fig. 1: (H&E 100x) Histopathological features of the ALT/WDLPS. The section shows mature adipose tissue with some fibrous zones. Fat cells vary in size, intermingled with atypical and hyperchromatic stromal cells and an occasional lipoblasts.

3. Discussion

Differentiating large lipomas from atypical lipomatous tumours (ALT) is challenging and preoperative management guidelines are not well-defined, but the appropriate classification is essential, as it may allow for observation, alter the surgical approach or prompt referral to a specialist. Most deep-seated, adipose tissue-derived large tumours are usually liposarcomas; although, lipomas can also reach similar sizes in some cases.[6] Lipomas exhibit mature adipose tissue only and do not show lipoblasts or atypical stromal cells. Most of the lipomas are small in size,[7] but giant lipomas as large as 55x38 cm have also been reported.[8] Differentiating between lipoma and liposarcoma on the basis of radiological findings is very difficult. Histopathological evaluation to assess cellular atypia, necrosis, mitotic activity and invasion in necessary for the differential diagnosis.[9]

Well-differentiated liposarcoma (WDLPS)/atypical lipomatous tumour (ALT) is deemed a low-grade malignant tumour that rarely metastasises but should be strictly followed because recurrence or dedifferentiation may occur. It is recognised that WDLPS and ALT are virtually synonymous, describing lesions that are identical both morphologically and karyotypically, and that site-specific variations in behaviour relate only to surgical resectability. The term WDLPS is now used for tumours of the retroperitoneum, mediastinum, and deep pelvis, whereas the term ALT includes tumours of the extremities and superficial sites.

Tumours located in the periphery usually do not metastasise. Complete resection is generally curative for such lesions. Therefore, designating the term sarcoma is inappropriate for these tumours, and the term Atypical Lipomatous tumour (ALT) is preferred. In deep-seated tumours (mediastinum, retroperitoneum, spermatic cord), the possibility of achieving negative margins is considerably diminished, and the threat of local recurrence, dedifferentiation (DDL) and death are more. These lesions are regarded as sarcomas, and the terminology of well-differentiated liposarcoma (WDLPS) is more appropriate.

Immunohistochemical and molecular analyses play a vital role in accurate diagnosis.

The cytogenetic characteristics of ALT/WDLPS are identified as surplus circular macrochromosomes with amplification of 12q13-15 sequences. CDK4 and MDM2 genes are persistently amplified in this region.[10] The MDM2 gene is responsible for the control of differentiation and proliferation of normal cells in humans.[11] MDM2 encoded proteins can promote tumorigenesis by negative regulation of the p53 protein. CDK4 regulates the G1/M cell cycle transition, and its inhibition can lead to the arrest of tumour proliferation.[12] The diagnosis of ALT/WDLPS depends upon histopathological examination combined with associated clinical information. In the absence of certain histological features, immunohistochemical staining for CDK4 and MDM2 can be done to identify ALT/WDLPS. High sensitivity and specificity in diagnoses of ALT/WDLPS can be achieved by using MDM2 and CDK4 immunohistochemistry or FISH simultaneously.[13]

The most crucial factor in prognosis and treatment is the anatomic location. ALT/WDLPS is mainly treated by surgical resection because radiotherapy and chemotherapy are not sensitive. For nonmetastatic ALT/WDLPS of limb,

the success of local excision is unclear. However, it is suggested that a margin of >1cm should be maintained for most soft tissue sarcomas. Some authors have also suggested that because ALT/WDLPS in the limb is nonmetastatic, a local excision with <1 cm margin can also be planned by the surgeon to optimise function and aesthetics. In the present case, extensive resection was difficult due to large tumour. Therefore excision was done along the tumour edge.

Absence of molecular testing for CDK4 and MDM2 expression by immunohistochemistry /or FISH present as a limitation to the present study.

4. Source of Funding

Nil.

5. Conflicts of Interest

None.

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