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CASE REPORT

ATYPICAL LIPOMATOUS TUMOR/WELL-DIFFERENTIATED LIPOSARCOMA ---- A RARE ADIPOCYTIC NEOPLASM

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ABSTRACT

Soft tissue tumors constitute a heterogeneous and large group of neoplasm and always have fascinated clinicians and pathologists for many years. Adipose tumors formed single most common histological group in soft tissue tumors and liposarcoma constitute 20% of all malignant mesenchymal tumors. Atypical lipomatous tumor/Well-differentiated liposarcoma (ALT/WDLPS) forms a rare adipose tissue malignant neoplasm. This is a case of a 31-year-old female, presented in surgical op with complaints of painless mass in the left thigh. Physical examination revealed a nodular swelling in the superficial soft tissue of the left thigh m/s 9x8cm and a pre operative diagnosis of lipoma was made. The mass was removed surgically and the microscopic sections showed a cellular adipocytic neoplasm arranged in lobules separated by fibrous strands. The fibrous septae show atypical cells with irregular hyperchromatic indented nucleus and cytoplasm showing multiple small vacuoles. Since the tumor was located in the superficial soft tissues, a diagnosis of atypical lipomatous tumor/Well-differentiated liposarcoma with low chance for recurrence was made and patient was referred to higher centre for further management.

INTRODUCTION

Soft tissue tumors constitute a heterogeneous and large group of neoplasm. Adipocytic tumors form most common soft tissue neoplasms. Malignant soft tissue tumors constitute about 1% of all malignancy and liposarcoma constitute 20% of these tumors. Atypical lipomatous tumor/Well-differentiated liposarcoma (ALT/WDLPS) is a rare soft tissue tumor first described by Virchow and forms the most common adipose tissue malignant neoplasm. This tumor usually seen in adult with higher predilection for men[1]. These tumors commonly originate from deep soft tissues and present as slowly growing painless mass. The single most important prognostic factor in these tumors is the original site of the lesion. These terms (ALT/WDLPS) are often used interchangeably because the lesions are histological indistinguishable. In this article we report a case of atypical lipomatous tumor/Well-differentiated liposarcoma originating from the soft tissues in the thigh.

Case report

A 31-year-old female, presented in surgical op in our hospital with complaints of painless mass in the left thigh. She noticed the swelling six months back, which increased gradually in size. She had no other symptoms referable to any other systems. She had full term normal delivery one month back. Her post partum period was uneventful and she was lactating.

Examination revealed a nodular swelling in the superficial soft tissue of the left thigh m/s 9x8cm .It was firm in consistency and was non tender. The skin over the mass was normal. No other abnormalities in the thigh were noticed. The regional lymph nodes were not palpable. Computerized tomography showed a well circumscribed localized soft tissue mass without significant regional lymphadenopathy. All routine investigations were all within normal limits. A pre operative diagnosis of lipoma was made. The mass was excised under general anesthesia. Peroperativly bleeding was more than expected and hemostasis achieved with difficulty .The excised mass was sent to the department of pathology.

Gross examination showed a nodular yellowish mass measuring 7x5x1.5cm. Cut surface was yellowish, firm and myxoid. No hemorrhagic /necrotic areas seen. The specimen was sequentially processed, and stained with hematoxylin and eosin. Microscopically sections showed fat and a cellular neoplasm arranged in lobules separated by fibrous strands. The fibrous strands show atypical cells with irregular

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hyperchromatic indented nucleus and cytoplasm showing multiple small vacuoles-lipoblasts. [fig.1]

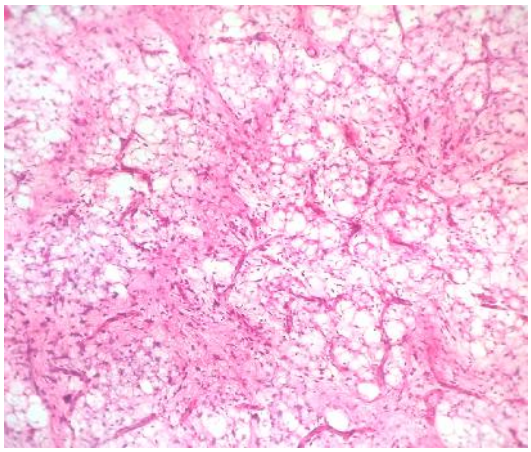

Fig.1 A cellular adipocytic neoplasm showing fibrous strands with atypical cells having irregular hyperchromatic indented nucleus (LP, 10X)

Lobules of tumor show mature adipocytes and occasional scattered cells showing centrally located nucleus with indentation by cytoplasmic fat vacuoles. Also seen lipoblasts with signet ring morphology. [fig.2]

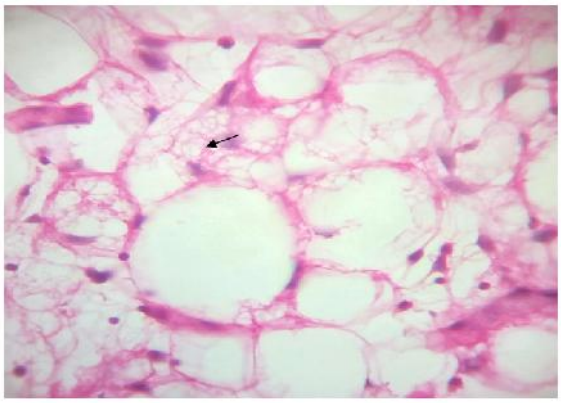

Fig.2 Lobules of tumor show lipoblasts (arrow) with centrally located indented nucleus and multiple cytoplasmic fat vacuoles. (HP, 40X)

Tumor showed marked vascularity with capillary proliferation. No mitosis/necrosis seen. No spindle cell/ myxiod areas/dense inflammatory infiltrate seen. The tumor cells seen reaching up to the surgical resected margins. From the above-mentioned gross and microscopic features a diagnosis of atypical lipomatous tumor/ Well-differentiated liposarcoma was made. Patient was referred to the higher centre for management.

DISCUSSION

Soft tissue tumors always have fascinated clinicians and pathologists for many years. This heterogeneous group include a large variety of tumors that show close histopathological similarities with only very few difference which is revealed only on accurate and careful microscopic examination. By definition Soft tissue represents the nonepithelial- extra skeletal tissue of the body exclusive of glia, reticuloendothelial system and supporting connective tissue of parenchymal organs. Adipose tissue, voluntary muscles, vessels and fibrous tissue form the soft tissue structures of the body. When the whole group of soft tissue tumors are considered, adipose tumors formed single most common histological group and liposarcoma constitute 20% of all malignant mesenchymal tumors [2]

The classification of liposarcoma includes the category of atypical lipomatous tumor/Well-differentiated liposarcoma. These commonly occur in middle aged adults with a peak incidence in the sixth decade of life and equally affect males and females. Common sites of ALT/WD liposarcoma are deep soft tissue thigh, retroperitoneum, mediastinum and the paratesticular area. These tumors can also arise very rarely in subcutaneous tissue and skin. These tumors when arise from extremities and superficial soft tissues are called as atypical lipomatous tumor since they are resectable and low recurrence rate are low (50%). On the contrary, retroperitoneal and groin tumors, because of their deep location, are usually less amenable to complete resection leading to higher recurrence rate (90%). and also in these deep tumors chances for transformation is also high (28% vs. 5%). So it is proposed that the term atypical lipoma can be used for tumors located in the superficial soft tissues and extremities and well differentiated liposarcoma for the latter. Four variants of WDL/AL have been described by the WHO: lipoma-like, sclerosing, spindle cell and inflammatory.[3,4]

Atypical lipomatous tumor/Well-differentiated liposarcoma resemble ordinary lipoma both grossly and on low power examination. But on closer examination we can see scattered atypical cells with large deep staining nucleus. These cells may be scattered among mature adipocytes (lipoma-like) or may be seen concentrated on the fibrous strands traversing the lobules of adipocytes (sclerosing). scattered in between we can also see classical lipoblast with eccentric /indented nucleus and multiple cytoplasmic vacuoles with fat. Some tumors show dense neutrophilic or lymphoplasmacytic inflammatory infiltrate (inflammatory). bland spindle cells arranged in fascicles or whorls in a myxoid stroma form the spindle cell variant.[5]

Ancillary tests like immunohistochemistry don't have much role in the diagnosis of these tumors. The tumor cells show positivity for Vimentin and S-100. Cytogenetically most of atypical lipomatous tumor/Well-differentiated liposarcoma show giant and marker ring chromosomes at the molecular level. These tumors are characterized by alternations of p53/mdm2 pathway. A link between ALT/WDLPS and q13-15/q14-15 mutation in chromosome 12 has been proven [6,7]

The single most important prognostic factor in ALT/WD liposarcoma is the original anatomical location. Tumors located in surgically accessible soft tissue do not usually recur following complete resection. But those located in deep sites such as retro peritoneum, mediastinum and spermatic cord are known for repeated recurrences. The risk of dedifferentiation is approximately more than 20% in the retro peritoneum but it is less than 2% when the tumor is located in the extremities

CONCLUSION

Atypical lipomatous tumor/Well-differentiated liposarcoma (ALT/WDLPS) is a rare soft tissue tumor. These lesions commonly occur in middle aged adults equally affecting males and females. Common sites are deep soft tissue thigh, retroperitoneum, mediastinum and the paratesticular area and they resemble ordinary lipoma both grossly and on low power examination. But on high power examination scattered atypical cells resembling the lipoblasts can be seen. The single most important prognostic factor being the original anatomical location lesions located on the superficial surgically amenable sites will have lower chance of recurrence and high disease free survival rates.

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Declarations

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