ISSN: 2230-9926

Available online at http://www.journalijdr.com

IJDR

International Journal of
DEVELOPMENT RESEARCH

International Journal of Development Research
Vol. 5, Issue, 01, pp. 3034-3036, January, 2015

Full Length Research Article

DEDIFFERENTIATED LIPOSARCOMA WITH LOW GRADE DIFFERENTIATION - A CASE REPORT

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ARTICLE INFO

Article History:
Received 24th October, 2014
Received in revised form
26th November, 2014
Accepted 30th December, 2014
Published online 26th January, 2015

Key words:
Dedifferentiated Liposarcoma,
Low Grade Differentiation,
Reteroperitoneum.

ABSTRACT

Introduction: Dedifferentiated liposarcoma (DDLS) is a rare sarcoma accounting for only 2% of all soft tissue sarcomas.It is the most common liposarcoma of the retroperitoneum. It is defined as the morphological progression of a well-differentiated liposarcoma to a nonadipocytic sarcoma.

Case Presentation: A 61 years old male presented to the outpatient department with symptoms of abdominal pain associated with distension for a duration of 2 months. A CT abdomen performed outside hospital showed a retroperitoneal mass for which he underwent a retroperitoneal tumor excision. Histopathological report of the excised tumor was given as pT2b Nx cM0- Grade 2, Dedifferentiated liposarcoma with low grade differentiation towards smooth muscle.

Discussion: The non adipocytic component of DDLS is 90% of times a high grade sarcoma. This paper presents a case of dedifferentiated liposarcoma of the retroperitoneum showing a low grade differentiation towards smooth muscle. This case has been chosen here for discussion here to the infrequent occurrence of low grade differentiation.

INTRODUCATION

Dedifferentiated liposarcoma (DDLS) is a rare sarcoma accounting for only 2% of all soft tissue sarcomas. It is the most common liposarcoma of the retroperitoneum. It is defined as the morphological progression of a well-differentiated liposarcoma to a nonadipocytic sarcoma. The non adipocytic component of DDLS is 90% of times a high grade sarcoma. This paper presents a case of dedifferentiated liposarcoma of the retroperitoneum showing a low grade differentiation towards smooth muscle.

Case Presentation

A 61 years male came to the OPD with complains of abdominal distention and vague abdominal pain. A CT scan taken outside hospital showed a mass with adherence to the appendix on one end and extending up to the right testis on the other end. Patient underwent surgery and the removed specimen was sent for histopathology. On gross examination, the specimen was an irregular nodular soft tissue mass measuring 30x23x14cm and weighing 2800 grams.

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The cut surface of the tumor showed predominantly yellow areas with focal myxoid change. There was also a nodular solid grey white lesion measuring 7.6x5x5 cm which was firm in consistency with whorled appearance (Fig. 1).

Histopathology

Hematoxylin and eosin stained sections showed sheets of adipocytes with lipoblasts (Fig. 2) which had moderate to scant amount of cytoplasm and multiple vacuoles indenting the pleomorphic hyperchromatic nucleus. Gradual transition to fascicles of spindle cells with mild nuclear pleomorphism and occasional mitotic figures - 1/10 high power field (HPF) was noted (Fig. 3). There was no necrosis noted in any of the sections studied. On immunohistochemistry, the dedifferentiated areas showed Smooth muscle actin (SMA) positivity (Fig. 4) which confirmed smooth muscle differentiation.

DISCUSSION

Adipocytic tumors are classified into benign and malignant adipocytic tumors (Table1) among which malignant adipocytic tumors constitute approximatly 20% of all soft tissue sarcomas of adulthood.

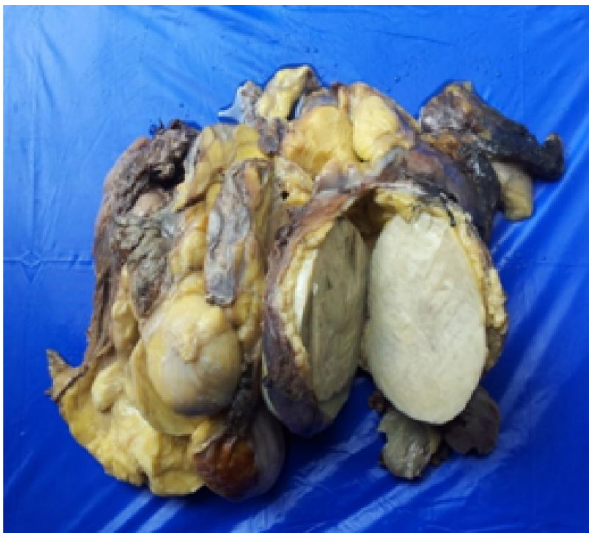

Figure 1. Gross: Cut surface: Predominantly yellow areas with focal myxoid changes with a nodular solid grey white lesion with whorled appearance

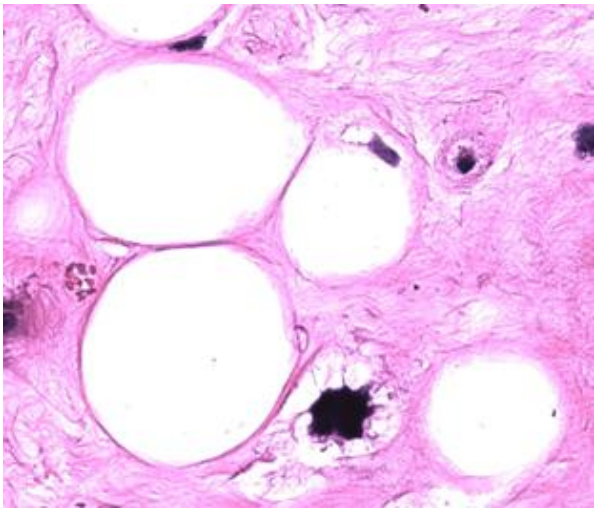

Fig. 2. H and E 10X: Lipoblast with pleomorphic hyperchromatic nucleus. Cytoplasm shows multiple vacuoles indenting the nucleus

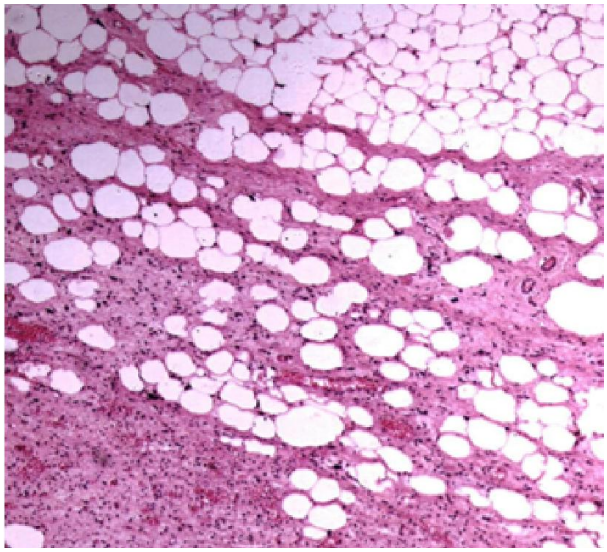

Fig. 3. H and E 4X: Indistinct zone of transition between the adipocytic and non adipocytic component

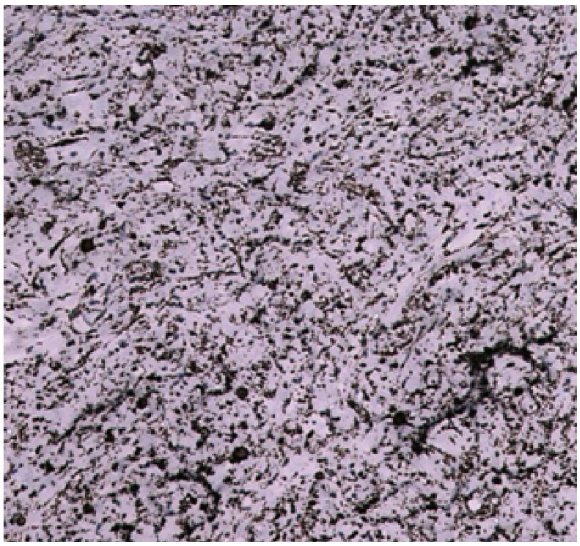

Fig. 4. IHC 200 X: Smooth muscle actin positive

Table 1.

WHO Classification of Adipocytic tumours (Fletcher *et al*., 2013)
Benign
Lipoma
Lipomatosis
Lipomatosis of nerve
Lipoblastoma / Lipoblastomatosis
Angiolipoma
Myolipoma of soft tissue
Chondroid lipoma
Spindle cell lipoma / Pleomorphic lipoma
Hibernoma
Malignant
Atypical lipomatous tumour /Well differentiated liposarcoma
Dedifferentiated liposarcoma
Myxoid liposarcoma
Pleomorphic liposarcoma

Dedifferentiated liposarcoma accounts for only 10% of all the liposarcomas. It is a relatively new tumor introduced by Evans in 1979 (Henricks *et al*., 1997). DDLS is a tumor composed of a highly cellular non lipogenic component along with a well differentiated liposarcomatous component. In most of the instances the non lipogenic component of DDLS shows high grade differentiation mostly resembling a fibrosarcoma or undifferentiated pleomorphic sarcoma.

A very small number of cases only have showed low grade areas of dedifferentiation (Henricks *et al*., 1997; Elgar and Goldblum, 1997 and Huang *et al*., 2005). DDLS is much less aggressive than other types of high-grade pleomorphic sarcoma. DDLS occur more commonly in males over 50 years of age. 75% of DDLS occur in the retroperitoneum followed by deep soft tissue of thighs, head and neck region, trunk, mediastinum, and the spermatic cord (Fletcher *et al*., 2013 and Weis *et al*., 2008). Retroperitoneal DDLS are usually large and have a tendency to involve the other native organs. Around 90% of DDLS occurs denovo and 10% arise as recurrence of a previously diagnosed liposarcoma, most commonly from a Well differentiated liposarcoma/Atypical lipomatous tumors. DDLS presents as large yellow to tan yellow, multinodular masses often more than 20 cm in size. The dedifferentiated component appear as sharply demarcated grey white firm to hard nodules. On microscopy, lesion is composed of ALN/WDL areas along with a dedifferentiated

area which can be minor or the dominant component. Transition between the two components can be abrupt or there can be a gradual transition as in our cases. The dedifferentiated component varies in patterns from low to high grades of various differentiations. About 90% cases have high grade dedifferentiation towards Undifferentiated pleomorphic sarcoma (Malignant fibrous histiocytoma) or Fibrosarcoma. Though few authors consider that dedifferentiated component should have a mitotic count of atleast 5/10 HPF, this criteria is not applicable since many non lipomatous components do not fulfill it (Evans, 2007). The undifferentiated pleomorphic sarcoma can be any subtype including the more prevalent storiform-pleomorphic and myxoid types to the unusual giant cell and inflammatory forms. Very few cases in literature have quoted the existence of morphologically low grade dedifferentiated liposarcomas consisting of spindle cells arranged in a vague fascicular pattern with minimal cytological atypia and necrosis and mitosis, sometimes called as "Cellular atypical lipomatous tumor" (Evans, 2007). Heterologous differentiation towards Rhabdomyosarcomatous, leiomyosarcomatous and osteosarcomatous component has been known to occur. Most of the DDLS shows positivity for MDM 2 and CDK4 on immunohistochemistry. DDLS have demonstrated abnormalities in the ring chromosome associated with MDM2 gene amplification .Loss of retinoblastoma protein (RB1 gene product) through promoter methylation and allelic losses is also commonly seen (Binh *et al*., 2005).

Conclusion

This paper presents a case of dedifferentiated liposarcoma of the retroperitoneum showing a low grade differentiation towards smooth muscle. This case has been chosen for discussion here because of the infrequent occurrence of low grade differentiation in DDLS.

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