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Dedifferentiated Liposarcoma with Meningothelial Like whorls And Metaplastic Bone: A Case Report.

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Abstract: Dedifferetiated liposarcoma is a malignant adipocytic neoplasm containing a non- lipogenic sarcoma of variable histologic grade along with well differentiated liposarcoma . Liposarcoma is the most frequent soft tissue sarcoma in adults but may also occur in adolescents and children. We report a case of dedifferentiated liposarcoma of the retro-peritoneum in a 47 years old male. Tumor showed dedifferention into meningothelial like whorls and metaplastic bone. The excised specimen was large, multi -nodular , firm, yellowish to tan gray on cut surface . The purpose of this report is to emphasize the histological features in dedifferentiated liposarcoma.

Keywords:- Dedifferentiated, Liposarcoma , Meningothelial, Retroperitoneal and whorl .

I. Introduction

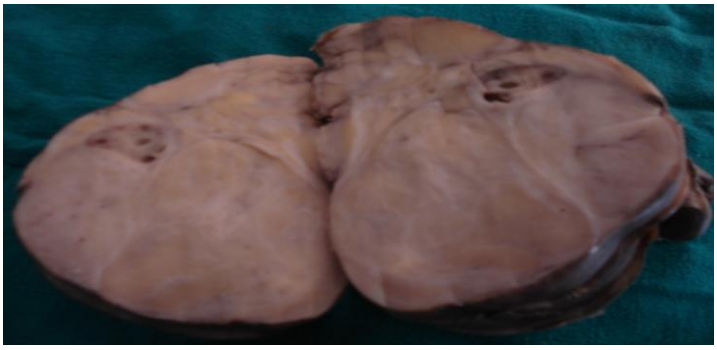
Sarcomas arising from retroperitoneum are rare tumors accounting for 10-15 % of all soft tissue sarcoma . Dedifferentiated liposarcoma was first described by Evans [1] in 1979 as a tumor composed of areas of non-lipogenic sarcoma associated with an atypical lipomatous tumor (ALT) /well differentiated liposarcoma (WDL). Liposarcoma are the most common sarcoma in adult , with commonest location in the retro-peritoneum . Distinctive, concentric whorl like formations occur commonly in Meningiomas and perineurial cell tumors , but occasionally similar formation can be seen in unrelated tumors. We report a case of liposarcoma with meningothelial like whorl formation and metaplastic bone along with areas of well differentiated liposarcoma.

II. Case Report

A 47 years old male presented in Surgery OPD of RIMS, Ranchi with a history of abdominal mass since 8 months and pain for 3 months. There was no bladder or bowel dysfunction. His past medical and family history were non- contributory to the present complaint. General physical examination was within normal limits . Per abdomen examination revealed a firm, lump extending from the left lumbar region to left iliac fossa , measuring about 17 x10 cm. Routine investigations like haemogram , Blood sugar, Blood urea, serum creatinine and serum electrolyte were within normal range. **CECT abdomen** showed a well defined fat attenuating mass lesion in left lumbar region extending to iliac fossa , measuring 15.7 x 9.7 cm with areas of solid and septal enhancement along with foci of calcification. Left colon was displaced downward and anteriorly. Retroperitoneal mass was removed surgically and sent for histopathological examination.

Gross pathology

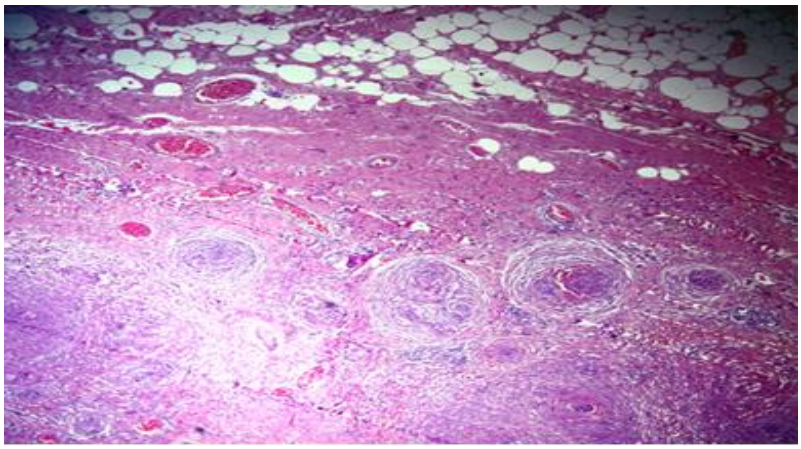
The excised specimen consisted of a large tumor mass, measuring 17 x10 x 7 cm in size multi - nodular, well- circumscribed, yellowish white in colour with smooth external surface , It was firm to cut with few areas of osseous consistency . Cut surface was variegated , showing lobules of varying size , yellowish to tan –gray in colour . Few cystic space were also seen.



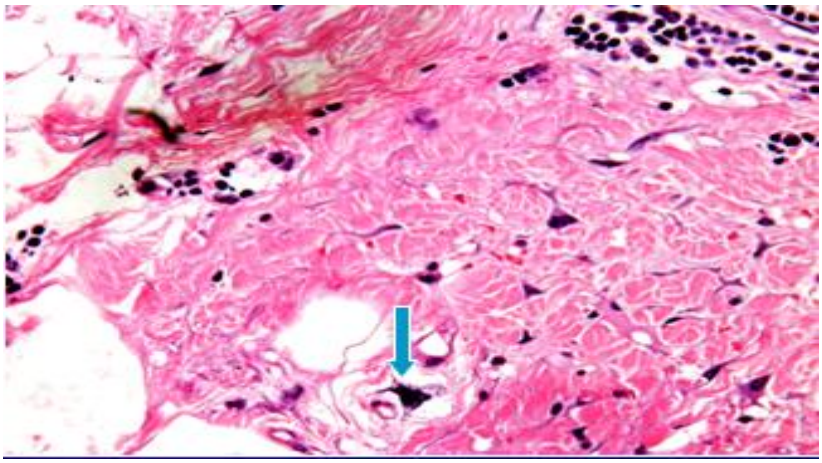
Lobules of varying size and few cystic spaces are seen (Figure 1)

Microscopy

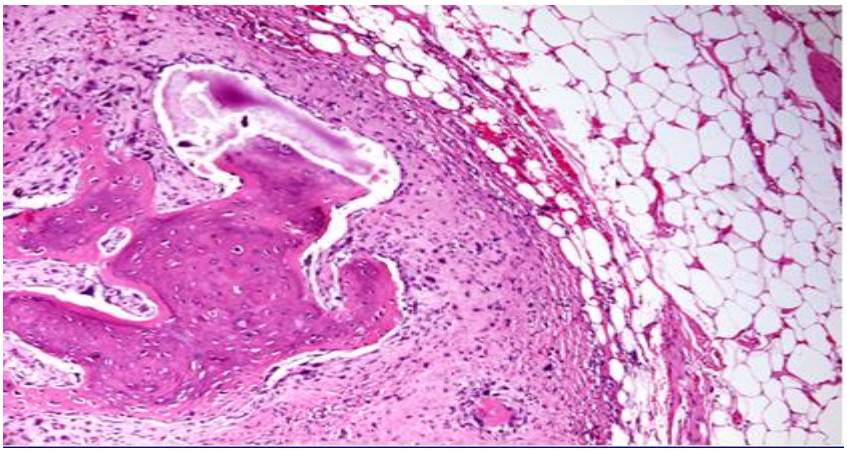
Scanner view showed sharply distinct lipomatous and cellular non- lipomatous areas with whorled structures. Lipomatous area showed atypical and multinucleated cells, lipoblasts and lymphoid infiltrate . Non-lipomatous areas showed scattered whorled structures in sclerotic stroma and foci of metaplastic bone . The whorls were composed of concentric lamellae of spindle or epithelioid cells embedded in a sclerotic stroma . The cells were plump with vesicular nuclei , well –defined nuclear borders, small or indistinct nucleoli and pale cytoplasm with indistinct cytoplasmic border . Numerous small capillaries were present in the whorls - predominantly in the centre . Psammoma bodies were not seen (Figure 2 , 3 and 4).



Scanner view showing lipomatous area & cellular nonlipogenic component with whorled structures sharp distinction b/w two components . (Figure 2)



(40x) - lipoblast, atypical cells & lymphoid infiltrate (Figure 3).



Metaplastic bone formation (Figure 4)

Panel of immunohistochemistory profile was set up to discern the nature of whorls. It showed positivity for Vimentin and α-smA . CD31 and CD34 was positive in blood vessels. Nuclear reactivity for p53 was seen in spindle cells forming whorls and was negative in lipomatous areas . S-100, EMA, and Desmin were negative .

III. Discussion

Our case showed dedifferentiated variant of Liposarcoma comprising of lipomatous and non lipomatous areas with meningothelial like whorls and metaplastic bone . Dahlin first coined the term Dedifferentiation in the context of chondrosarcoma in 1971[1] .He described it as a morphologically bimorphic neoplasm showing areas of well-differentiated low grade tumors juxtaposed with high grade non –chondroblastic tumors without obvious areas of gradual transition. In 1979, Harry Evans [2] used the term Dedifferentiated to describe a Liposarcoma, which was characterized by a combination of well – differentiated liposarcoma and a non –lipogenic dedifferentiated sarcoma like component. The dedifferentiation may occur as a de novo phenomenon also known as primary dedifferentiation or dedifferentiation in the recurrence of a previous entirely well differentiated liposarcoma [3].

Liposarcoma encompasses [4] a broad morphological range of histological pattern. It includes three groups (a) Atypical lipomatous tumor or well dedifferentiated liposarcoma (b) Myxoid and round cells and / or cellular myxoid spectrum and (c) Pleomorphic liposarcoma. Dedifferentiated liposarcoma is a biphasic neoplasm , one component is well diffrentiated liposarcoma tumor and other component is cellular non-lipogenic sarcoma. 90% arise denovo and 10% are recurrent tumors[4]. 5-15 % well differentiated liposarcoma undergo dedifferentiation. Retroperitoneum is the most common location . 90 % of dedifferentiated tumors have appearance of high grade fibrosarcoma , malignant fibrous histiocytoma, rhabdomyosarcoma or osteosarcoma . 10% appear as low grade dedifferentiated liposarcoma[5]. The close association of whorls with metaplastic bone in a background of liposarcoma , differentiates these whorls from those seen in meningeal or neural tumors [7] . In our case , whorls comprising of spindle cells in a sclerotic stroma were seen in close association with metaplastic bone. The dedifferentiated areas were sharply distinct from areas of Well differentiated liposarcoma . Whorls of liopsarcoma have different cell composition as demonstrated by immunohistochemistry . Spindle cells in the present case were negative for EMA and S-100 but positive for alpha smooth muscle actin there by showing pericytic or myofibroblastic phenotype . Liposarcoma with meningothelial like whorls is a morphological variant of liposarcoma . These whorls lack the immunoprofile of meningothelial and perineural cells but show mesenchymal cell markers, which may undergo myofibroblastic and osteoblastic differentiation. Spindle cells of whorls in our case exhibited p53 positivity suggestive of disease progression . Whorl formation represents an early sign of dedifferentiation in liposarcoma [6]. Clinical behavior of these tumours is variable . 41% have local recurrence and 15-20% develop metastasis. There is 28-30% mortality at 5year follow up [4].

IV. Conclusion

Present case may be categorized as Low grade de-differentiated liposarcoma , with presence of whorls, metaplastic bone and component of well differentiated liposarcoma . Dedifferentiated liposarcoma represents aggressive variant of liposarcomas . Overall biological behavior is determined by most aggressive element which is present in non-lipomatous Portion of tumor. Meningothelial whorls represent mesenchymal proliferation and may undergo differentiation towards Pericytic , myofibroblastic or ostoblastic components in liposarcoma . Meningothelial whorls represent an early sign of dedifferentiation of liposarcoma.

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