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Lipomatous Ganglioneuroma of the Retroperitoneum

Dr. Garima Pandey[1], Dr.Ashwani Tandon[2], Dr.Rekha Singh[3]
[1] *Assistant Professor , Pathology, SunderlalPatwa Government Medical College Mandsaur(M.P)*
[2] *Professor, Pathology, All India Institute of Medical Sciences ,Bhopal(M.P)*
[3] *Additional Professor , Endocrinology, All India Institute of Medical Sciences ,Bhopal(M.P)*

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ABSTRACT
Lipomatousganglioneuroma (LG) is a rare ,differentiated ,benign,slow growing tumor that originates from primordial neural crest cells and is composed of mature schwann cells, ganglion cells and nerve fibres[1] .Ganglioneuroma is most frequently diagnosed in patients between ages of 10 years and 29 years and is most commonly located in posterior mediastinum, followed by the retroperitoneum[2] .We report a peculiar ganglioneuroma with the unusual prominent lipomatous component, namely lipomatousganglioneuroma , occurring in adrenal.
Keywords: Ganglioneuroma, Lipomatous component, Retroperitoneal mass

I. INTRODUCTION

Ganglioneuromas are fully differentiated tumors that contain no immature elements. They are rare when compared with other benign neural tumors, such as, schwannoma and neurofibroma. They are usually seen in children above 10 years of age and are most often located in the posterior mediastinum, followed by the retroperitoneum[3]. Microscopically ganglioneuroma resemble neurofibroma, with a dominant schwannian stroma, along with the presence of fully mature ganglion cells. Fatty replacement of the ganglioneuroma is very rare. The presence of fat alters the tissue composition, hence, it alters their density when examined radiologically by imaging techniques like CT/magnetic resonance imaging (MRI), and serves as a useful diagnostic sign[4]. Histopathology still remains the gold standard in their diagnosis. This rare change also helps to narrow down the list of clinical differential diagnosis for retroperitoneal masses.

II. CASE REPORT

A 53-year-old male presented with hypertension,pain in abdomen,palpitations since 3 months associated nausea ,vomiting and weight loss.

Magnetic resonance imaging revealed a well defined homogenous soft tissue lesion epicentered in left suprarenal location of size 5.6cm X 3.5cm X 5.6cm,arising from medial limb of left adrenal gland.Lesion is homogenously hypointense on T1 and mixed iso-mildly hyperintense on T2.No loss of signal seen on fat saturated sequences.

Grossly, the smooth encapsulated and glistening neoplasm had a homogenous greywhite cut surface and rubbery consistency.

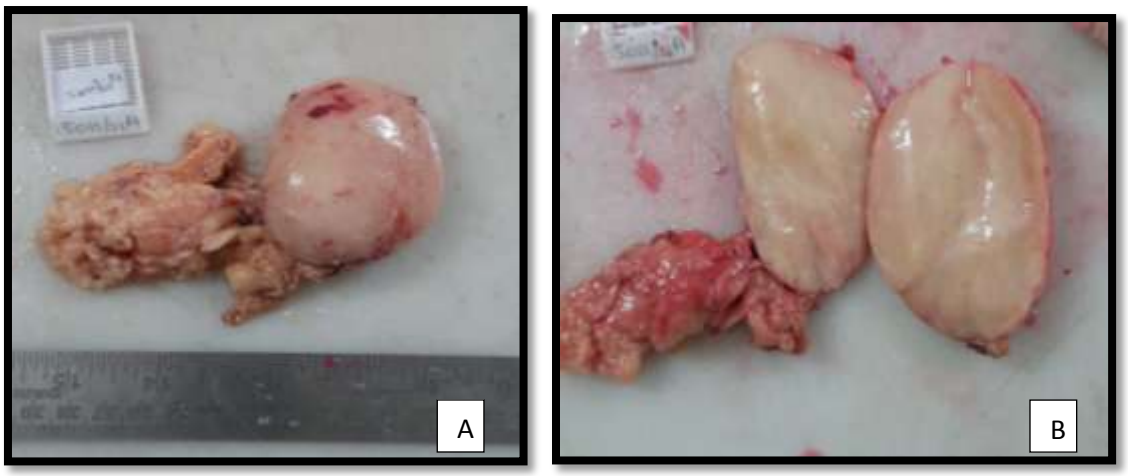

Fig.1:-On gross examination encapsulated tumour with smooth glistening outer surface with homogenous grey white cut surface

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Microscopic evaluation revealed an encapsulated lesion that consisted of areas of ganglioneuroma admixed with areas of mature fat. Ganglioneuroma regions showed fascicular growth patterns of mature schwann cells with collagenous stroma. Characteristically,mature adipocytes were seen all over the lesion in varying areas of tumor,such as in lobules and the foci of fat cells.The spindled schwann cells were ill defined and had scanty ,weakly eosinophillic cytoplasm with serpentine nuclei. Nuclei atypia and mitoses were absent and no cellular atypia, necrosis or vascular invasion was observed. Mature ganglion cells are scattered in between the schwann cells . The large polygonal ganglion cells had abundant granular cytoplasm and large round nuclei. No neuroblasts were identified. By immunohistochemistry, the ganglion cells were positive for chromogranin and synaptophysin, whereas the Schwann cells were positive for vimentin, S-100 protein, and glial fibrillary acidic protein (GFAP).

On the basis of these findings a diagnosis of LipomatousGanglioneuroma was made.

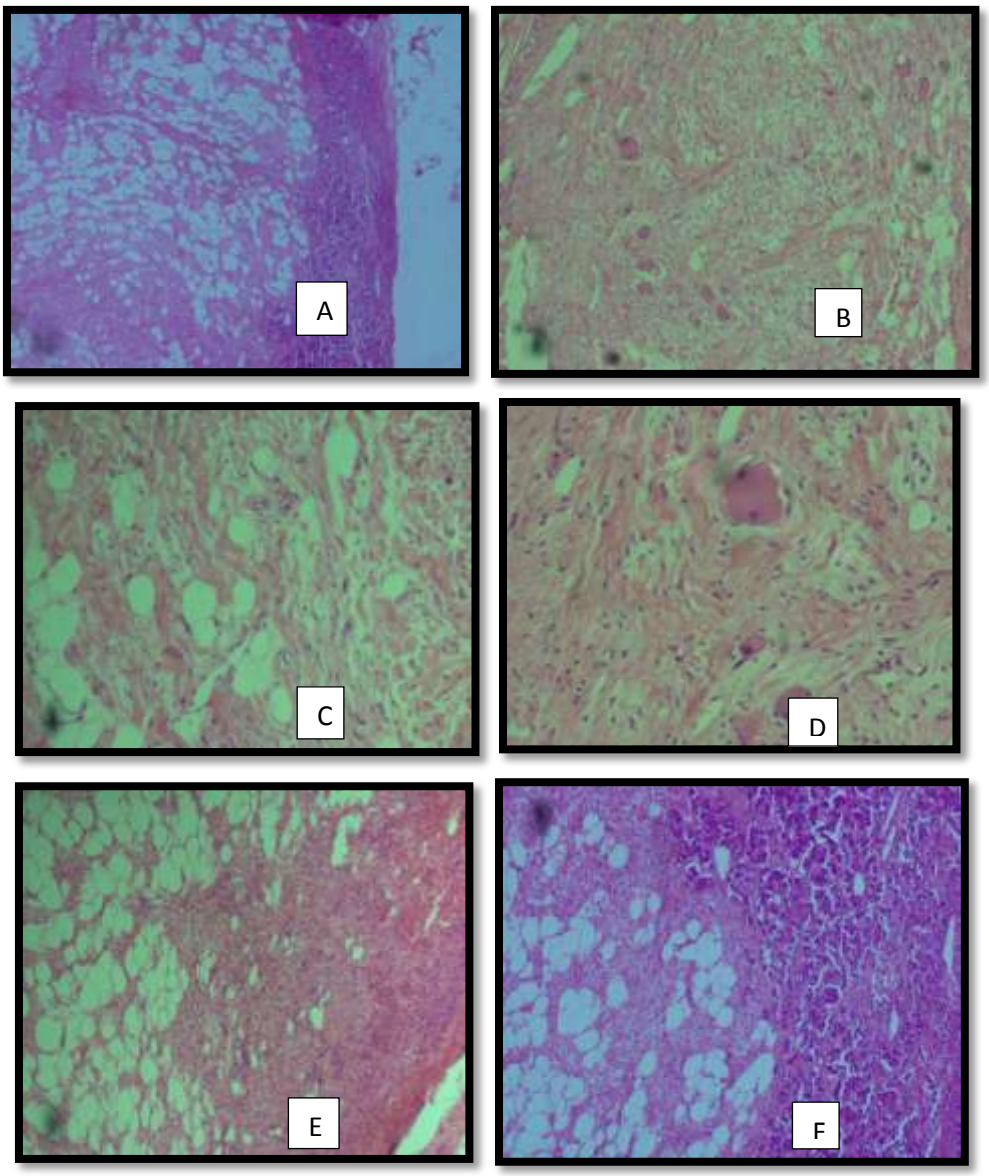

Fig.2Hematoxylin and eosin (40X)stained sections from adrenal gland shows an encapsulated lesion that consisted of areas of ganglioneuroma admixed with areas of mature fat.

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III. DISCUSSION

Lipomatous Ganglioneuroma (LG),previously called "ganglioneuroma with fatty replacement" is extremely rare variant of ganglioneuroma first reported by hara et al [5].It is characterized by a mature adipocytic component admixed with a conventional ganglioneuroma. Most lesions presented clinically as asymptomatic tumors and were discovered fortuitously. As documented in table LG seems to occur in middle aged adult patients(27-73 years).The sex distribution for LG cases was 3 females and 2 males. The size of lesions ranged from 2cm to 11cm.The posterior mediastinum and retroperitoneum were the only affected sites reported till date.

LG is grossly sharply marginated with or without encapsulation. Cut section show whitish and yellowish tissue. The hallmark microscopic feature is a varying combination of ganglioneuroma like and lipomatous elements. The histogenesis of fat element is controversial.Hara et al [5] reported presence of adipose tissue in tumor as spontaneous degeneration with fatty replacement. Adachi et al [7]reported adipocytes in the tumor derive from tumor cells themselves.As all reported tumors were well circumscribed, infiltration or entrapment of surrounding adipose tissue could not have generated this. Tumor cells originating in neural crest may have the capacity for lipomatous differentiation.

Table 1.**Published Cases Of Ganglioneuroma At Different Locations With Fatty Replacement**

REFERENCE	SEX/AGE(Yr)	LOCATION	SIZE(cm)
Hara et al [5]	F/54	Mediastinum	11
Duffy et al [4]	F/27	Mediastinum	NA
Ko et al [6]	F/53	Mediastinum	9
Adachi et al [7]	M/73	Retroperitoneum	2
Qing da meng et al[1]	M/44	Retroperitoneum	9.5
Present case	M/53	Retroperitoneum	5.6

The differential diagnosis of fat containing retroperitoneal masses includes lipoma,liposarcoma,spindle cell lipoma and angiomyolipoma. In our study, absence of atypical lipoblasts and presence of characteristic schwannian background exclude well differentiated liposarcoma.Differentiating an angiomyolipoma,especially with a prominent lipomatouscomponent,from an lipomatousganglioneuroma is based on former's distinctive arrangement of spindle cells around thick walled vessels .

IV. CONCLUSION

In conclusions,we report a very rare variant of ganglioneuroma that is lipomatousganglioneuroma in the adrenal gland. The presence of Schwannian stroma with mature ganglion cells in the entire tumor area formed the basis for the diagnosis of ganglioneuroma in our case. Although ganglioneuromas are not rare, the significant fatty replacement of the tumor stroma is a rare finding and this change serves as a useful radiological and pathological tool to narrow down the list of clinical differential diagnosis for the retroperitoneal masses.Neuroblastic tumor group includes ganglioneuroma, ganlioneuroblastoma, and neuroblastoma.In this spectrum, the ganglioneuromas are benign, whereas, neuroblastomas are aggressive in nature and ganglioneuroblastomas are intermediate in biological behavior. Therefore, more cases of Lipomatous Ganglioneuroma must be studied to draw definitive conclusions about its distinct behavior and management.

Conflict of interest: None

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