

Retroperitoneal Ganglioneuroblastoma In An Adult With Intratumoral Bleed

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INTRODUCTION

Ganglioneuroblastoma (GNB) is a tumor arising from nerve tissue which is intermediate between benign ganglioneuroma (slow growing and unlikely to spread) and malignant neuroblastoma (fast growing, aggressive, and likely to spread). It is an extremely rare neoplasm of adults. It usually occurs in young children with equal frequency in either sex. Here we present a rare case of a large retroperitoneal ganglioneuroblastoma in an adult male patient with intratumoral bleed.

CASE REPORT

A 40 year old man, presented with a history of pain in epigastrium, occasional vomiting and weight loss since four months. On examination a large lobulated mass was felt in the epigastric and umbilical region, which was fixed and hard in consistency. Laboratory examination revealed mild anaemia (Hb 11.9 gm%) and raised ESR (60 mm in 1 hr).

Abdominal USG demonstrated a large lobulated mass in

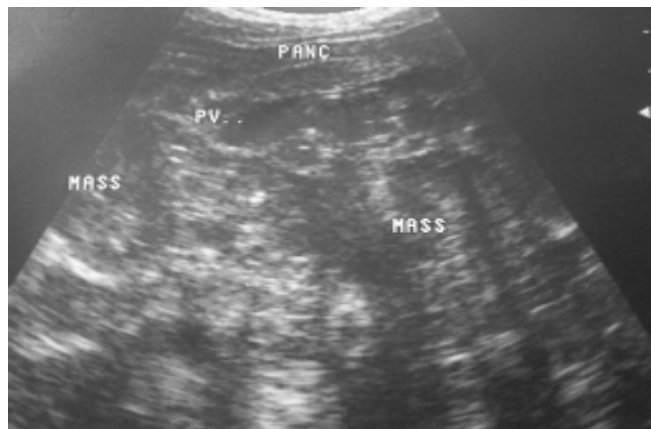


Fig.-1 : Sonogram showing mixed echoic retroperitoneal mass displacing the pancreas anteriorly and reaching up to the caudate lobe of liver.

the retroperitoneum medial to the left kidney, displacing it laterally; extending superiorly towards upper pole of spleen, displacing the pancreas anteriorly and extending across midline to reach up to the medial margin of caudate lobe of liver (Fig-1). In the midline the mass was seen to encase the aorta and the vessels arising from it (celiac axis and superior mesenteric artery) (Fig-2). The mass was composed of innumerable hypo to mixed echoic lobules with echogenic non shadowing calcific foci dispersed diffusely within it. Additional findings included small anechoic cysts in liver and sludge in gall bladder.

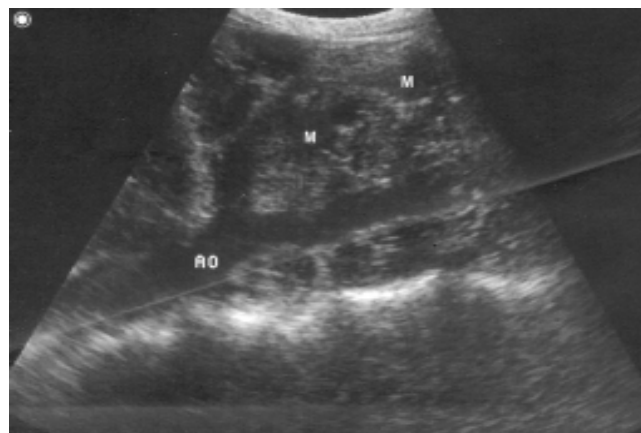


Fig.-2 : Sonogram showing the lobulated mixed echoic mass encasing the aorta and vessels arising from it.

CT examination revealed a large approximately 18.1 x 8.7 x 15 cm lobulated heterogeneously enhancing mass with smooth margins and density varying from 6.8 HU to 38.6 HU on unenhanced scans, increasing up to 58 HU in solid region of the mass after contrast injection. Enhancing septae and amorphous calcific foci (73 to 201 HU) were also seen within the mass (Fig-3). The mass was located superior to the left kidney extending further superiorly medial to spleen, displacing the hilar vessels anteriorly; inferiorly extending between the aorta and the left kidney up to the level of L3 vertebra; anteromedially extending

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across the spine encasing the midline vessels (aorta, celiac axis, superior mesenteric artery and renal arteries) without compromising their lumen and reaching upto the medial margin of right kidney and the caudate lobe of liver (Fig-4). Pancreas, stomach, jejunal loops, inferior vena cava, portal vein, splenic and hepatic artery were displaced anteriorly by the mass. Normal right adrenal could be identified separately in its usual position. Left adrenal gland could not be identified separately from the mass. An interesting feature, a fluid blood level was seen in one of the lobules of the mass signifying high vascularity and intratumoral bleed within it (Fig-5). Enlarged retrocaval lymph nodes showing faint enhancement were identified (Fig-6). Both the kidneys showed normal contrast excretion. The proximal left ureter was displaced laterally by the mass. Other findings included nonenhancing cysts in the liver and sludge in the gall bladder. There was no evidence of any free fluid or parenchymal metastatic deposits. Vertebral body and posterior ends of ribs did not show any feature of erosion



Fig.-3 : CT with Oral contrast only showing the lobulated retroperitoneal mass with amorphous calcific deposits causing lateral displacement of left kidney.

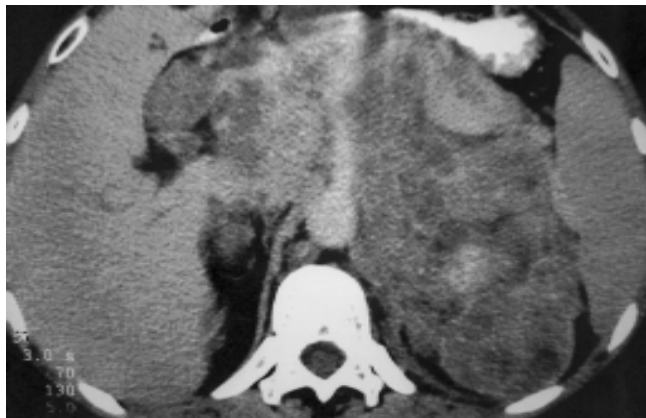


Fig.-4 : CT with both Oral and IV contrast showing encasement of the celiac trunk by the mass and anterior displacement of hepatic and splenic artery. Enhancing septae can also be seen within the mass.

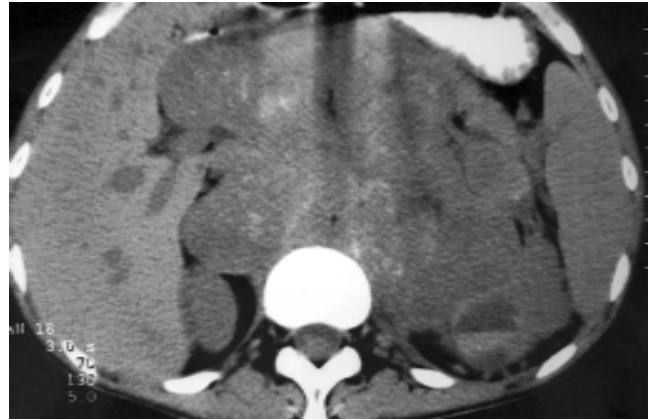


Fig.-5 : CT with Oral contrast showing lobulated mass with a fluid blood level in one of the lobules in left lumbar region.

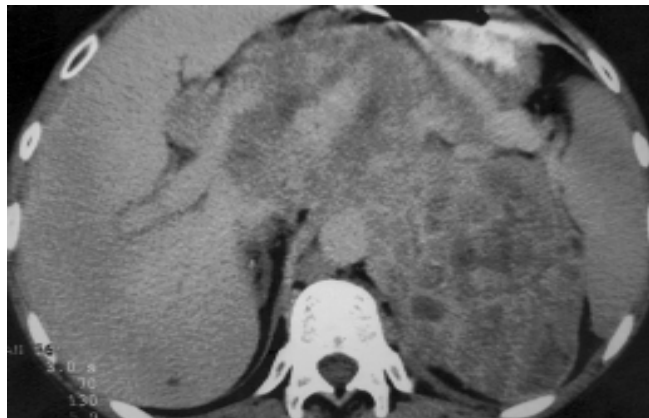


Fig.-6 : CT with both Oral and IV contrast showing normal right adrenal, heterogeneously enhancing retroperitoneal mass and enlarged retrocaval lymph nodes.

On the basis of location of the mass, lobulated heterogenous nature, vessel encasement, amorphous calcific foci, nonvisualisation of the left adrenal, retrocaval lymphadenopathy; the differential diagnosis of malignant neurogenic tumor or a conglomerate nodal mass was made.

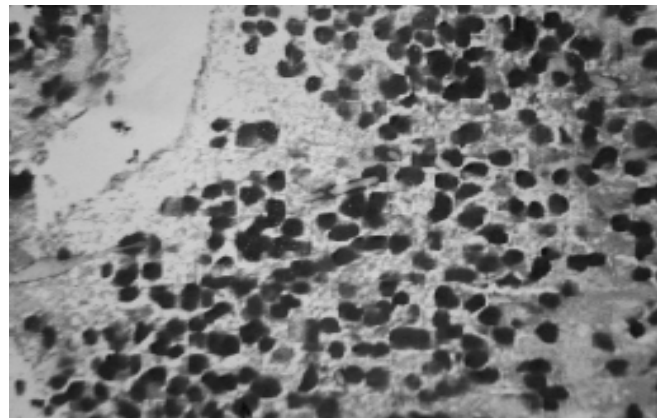


Fig.-7 : Histopathological slide with H& E stain (40X magnification) showing ganglion cells mixed with small round cells in a fibrillary background.

USG guided trucut biopsy revealed collagenous fibrous tissue with a tiny fragment of lesion. The fragment of lesion showed ganglion cells mixed with small round cells in a fibrillary background. There was no evidence of necrosis (Fig 7). Histopathology examination was consistent with ganglioneuroblastoma.

The patient was referred to higher center for treatment where on the basis of CT findings the lesion was deemed to be inoperable. Prognosis was explained to the patient and his relatives and the patient was discharged on symptomatic treatment. After four months of regular follow up, the patient is still alive and has lost upto 24 kg of weight.

DISCUSSION

Tumors of ganglion cell origin include ganglioneuroma (GN, benign); neuroblastoma (NB, malignant); and ganglioneuroblastoma (GNB, intermediate). Neuroblastoma and ganglioneuroblastoma most often occur in infants and children, whereas ganglioneuroma tends to occur in adolescents and young adults [1-3]. The term 'ganglioneuroblastoma' was first applied by Robertson in 1915 to neurogenic tumor with combined histologic features of ganglioneuroma and neuroblastoma[4].

It is a rare tumor that has a yearly occurrence of less than 5 per million children, most often seen in patients of 2-4 years of age and is exceedingly rare after the age of 10 years [4]. According to Yamanaka et al [5] only 33 cases of retroperitoneal ganglioneuroblastoma in adults have been reported in the English medical literature and Mehta et al [6] have reported a case of bilateral intra-abdominal ganglioneuroblastoma in a patient of 20 years of age from India.

The most common tumor site is the abdomen (adrenal and sympathetic chain), followed by the mediastinum, neck, and lower extremity. It is usually less aggressive than a neuroblastoma with significantly more favorable prognosis and response to therapy than that of neuroblastoma [7].

At gross examination, ganglioneuroblastomas may be partially or totally encapsulated and frequently contain granular calcification [8]. Histopathologically, they are malignant tumors containing primitive neuroblasts and

mature ganglion cells. Their appearance varies depending on the number of ganglion cells, their degree of differentiation, and their relationship to immature elements. Therefore, the reported CT findings in ganglioneuroblastomas also vary, ranging from a predominantly solid mass to a predominantly cystic mass with a few thin strands of solid tissue.

In our case the tumor arises from the left sympathetic chain and has metastasized to the regional lymph nodes. It is predominantly solid in nature with few cystic components in it. There is no evidence of any paraneoplastic syndrome. These findings are in harmony with Kilton et al [] who described that local invasion and regional lymph node metastasis were noted frequently and that only a small percent of adults with ganglioneuroblastoma seem to produce biologically active hormones. The radiological finding of fluid blood level signifying intratumoral bleed within a GNB has not been reported so far. Although vascular encasement has been described in GN & NB, such extensive encasement and displacement of vessels by a GNB has not been reported earlier.

Although ultrasound could identify most of the features of the tumor, but the nature of the mass (solid, cystic, bleeding and calcification) and its extent were better delineated by CT. CT also had an edge over ultrasound in depicting enlarged retrocaval lymph nodes and relationship with other intra abdominal structures.

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