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Article · March 2013

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CASE REPORT

Retro-Peritoneal Extra-Adrenal Ganglioneuroma Encasing Major Blood Vessels

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ABSTRACT

Ganglioneuroma is a rare, benign, slow-growing asymptomatic tumor of the neural crest cells. It occurs in one per million population with female predominance. It most commonly occurs in the retro=peritoneum. Although CT scan and other imaging modalities can be used, its diagnosis is essentially based on histopathology. We report a case of 18 year old male who presented with abdominal pain and a mass in the left lumbar quadrant. CT scan abdomen showed a retroperitoneal mass encasing aorta, IVC and left common iliac artery. A diagnosis of Ganglioneuroma was established on ultrasound guided Trucut biopsy. He underwent laparotomy for total excision of mass. Per-operatively the mass was separated from the vital structures and the major blood vessels. His post-operative course was uneventful and he was discharged on 7th post-operative day. His final histopathology confirmed the initial diagnosis of ganglioneuroma. Nonetheless, a possibility of ganglioneuroma should be kept under consideration in cases of all retroperitoneal extra-adrenal masses.

KEY WORDS: *Ganglioneuroma, Retroperitoneal Mass, Extra-Adrenal Mass*.

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INTRODUCTION

Ganglioneuroma (GN) is rare benign neuroblastic tumor which arises from ganglion cells mainly from the sympathetic chain. It may arise from base of skull, neck, posterior mediastinum, retroperitoneum or adrenal glands.[1] There is female predominance with peak age of incidence 10 to 40 years and most of the cases occur before the age of 20.[2] These tumors are slow-growing and are asymptomatic; sometimes diagnosed as incidental findings. They are usually non-functioning and give rise to symptoms only due to significant pressure on surrounding structures.[1] The important differentials are neuroblastoma and ganglioneuroblastoma. Nonetheless, biopsy is the only definitive tool of diagnosis.

CASE REPORT

An 18 year old boy presented to our out-patient clinic with complains of left upper abdominal pain since the age of six. His parents had first noticed a swelling in left side of the abdomen when the patient was 3 year old. This was initially painless but became symptomatic at the age of 6 yrs. The pain was intermittent and was very mild in intensity. The mass gradually increased in size over the years and caused a gradual increase in intensity of pain. There was no associated weight loss, urinary symptoms or hypertension. On examination a mildly tender mass was palpable in the left Hypochondrium to lumbar quadrant. It was smooth, firm and irregular in shape.

A CT scan abdomen was done which showed a heterogeneous retroperitoneal mass 20.8 x 7.1 x 4.2 cm^3 abutting aorta, Inferior Vena Cava (IVC) and left common iliac artery. There was no involvement of adrenal gland or left ureter (Fig 1). There was an irregular calcification within the mass which measured 4.3 x 1.2 cm^2. On the basis of these findings a diagnosis of benign non-secreting retroperitoneal mass was made and the patient underwent ultrasound guided Trucut biopsy of the lesion. The histopathological findings of biopsy were typical of Ganglioneuroma and hence surgical excision of the mass was planned.

The patient underwent a midline laparotomy. During the procedure vital structures including left ureter, aorta, IVC and common iliac vessels were identified. Meticulous blunt dissection of the tumor was done from the vessels followed by separation of tumor from its posterior attachment. Total excision of mass was done using dissection in the cleavage plain (Fig 2). All surrounding structures were preserved as far as possible. The posterior part of parietal peritoneum was not closed and a gravity drain was placed in-situ before closing the wound. Post-operatively the patient made an uneventful recovery and the patient was discharged in 7th post-op day.

The histopathology of the patient showed cores composed of spindle shaped cells arranged in short fascicles with elongated wavy nuclei, indistinct nucleoli and moderate eosinophilic cytoplasm. Intermingled large polygonal cells displaying abundant amphophilic cytoplasm, large nuclei and distinct nuclei were noted. Foci of dystrophic calcification and collection of small lymphocytes were also seen. Spindle cells were stained positive for S-100. These features were consistent with extra-adrenal Ganglioneuroma

DISCUSSION

Ganglioneuroma belongs to the family of neuroblastic tumors which arise from the embryonic undifferentiated cells of the neural crest. It comprises of Schwann cells, neuritis, ganglion cells and fibrous tissue.[3] The other tumors in this class are neuroblastoma and ganglioneuroblastoma.[4] GN have familial predisposition and are occasionally associated with Multiple Endocrine Neoplasia - II (MEN – II) and Turner's syndrome.[5] It most commonly occurs in the paraspinal retroperitoneum from the sympathetic ganglia. Other common sites include posterior mediastinum (39%), Adrenal medulla (20-30%), neck or pelvis (9%).[4,7] It can rarely be found in spermatic cord, heart, bone, intestine.[7]

The reported incidence is about one per million population.[5] There is female preponderance with peak incidence between second and fourth decade. It is rarely diagnosed after the sixth decade of life and approximately two-third of the lesions are diagnosed before the age of 20 years.[6]

The clinical features of GN are non-specific including vague abdominal pain and neurological symptoms due the mass effect on the surrounding structures.[6] Labile hypertension, diarrhea and flushing can be seen in patients with hormone secreting tumor.[6] Secreted hormones include vasoactive intestinal peptide (VIP), androgenic hormones, vinylmandalic acid (VMA) and homovanilic

acid (HVA). About 30% of the patients have elevated plasma and urinary catecholamine level.[3] Catecholamine secreting GN may cause hypertensive crisis during surgery.[5]

GN remains asymptomatic for years before being diagnosed incidentally or during work-up of non-specific symptoms. On Ultrasound it appears as circumscribed homogenous or hypo-echoic lesion.[6] CT scan shows density of lesion less than or equal to that of liver; with or without patchy irregular calcification. It may grow around major vessels without invasion.[6] On MRI it appears as hypointense lesion in T1-weighted with is hyperintense signals on T2W, which is specific for diagnosis of GN resulting from combination of ganglionic cells and myxoid material.[6]

Iodine-tagged metaiobenzylguanidine (MIBG) uptake can be used but is non-specific since it is increased in all catecholamine producing tumor.[4] Increased MIBG uptake is found in 57% of GN and 92% of neuroblastoma.[3] High level of MIBG uptake along with increased urinary catecholamine levels can differentiate GN from neuroblastoma.[1] Nonetheless CT scan is the most frequent radiological investigation performed because it identifies the organ of origin, extent of tumor, vascular encasement, regional invasion, adenopathy and calcification if any.[5]

Histological evidence is required to establish the diagnosis of GN and pre-operative diagnosis is usually difficult to establish. Tissue can be taken using ultrasound or CT guided needle aspiration or open biopsy.[4,6] Histologically GN should be differentiated from ganglioneuroblastoma on the basis of ganglioneumatous component which is minimal in the former and more than 5% in the latter.

The treatment of GN essentially remains surgical. Total excision with clear margins should be achieved wherever possible. Surgical approach is dictated by the tumor size and differential diagnosis based on radiology. Complete excision becomes difficult when it is encasing the vital structures like aorta, IVC, Ureter or nerves. It causes minimal or no compromise of the lumen.[5] Therefore there is usually a cleavage plain available for clear dissection. Laparoscopic excision of retroperitoneal GN has been reported[6]; However, tumor size and encasement of surrounding structures should be kept under consideration.

In our patient the tumor was surrounding the aorta, IVC, left common iliac artery. Therefore a retroperitoneal approach was not taken. An anterior midline laparotomy enabled us to clearly identify and preserve the major vessels. Following which the tumor was taken out completely. During separation of tumor from the vessels, venous bleeding is disastrous. It is usually far more dangerous than arterial bleeding owing to minimal musculature in the vessel wall.

Role of chemotherapy or radiotherapy is limited to ganglioneuroblastoma only and is not indicated in GN.[5,7] Post-surgical prognosis of GN is excellent; however, there is evidence of delayed recurrence and malignant transformation with metastasis which justifies periodic radiological follow-up.[5]

In conclusion, retroperitoneal GN is essentially benign neural crest tumor. They may remain asymptomatic for years and grow into very large mass. It surrounds major vessels but does not invade them. Complete surgical excision usually cures it but regular follow-up is recommended.

Figure 1: CT scan abdomen with contrast showing retroperitoneal mass encasing the aorta, and left iliac artery. It is also abutting inferior vena cava and right common iliac artery. There are no signs of invasion of blood vessels.

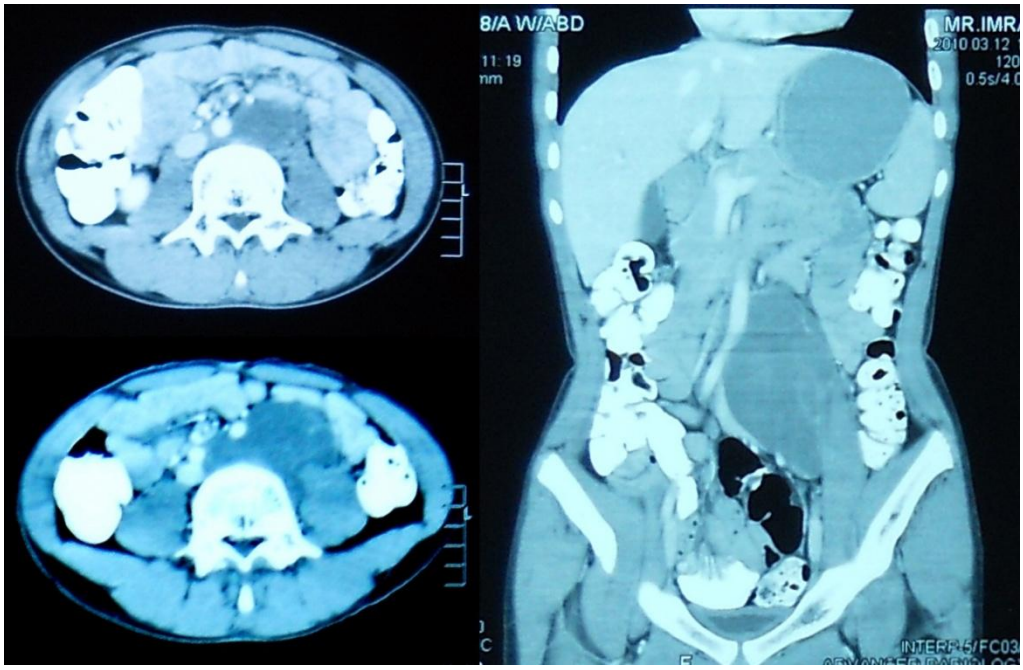
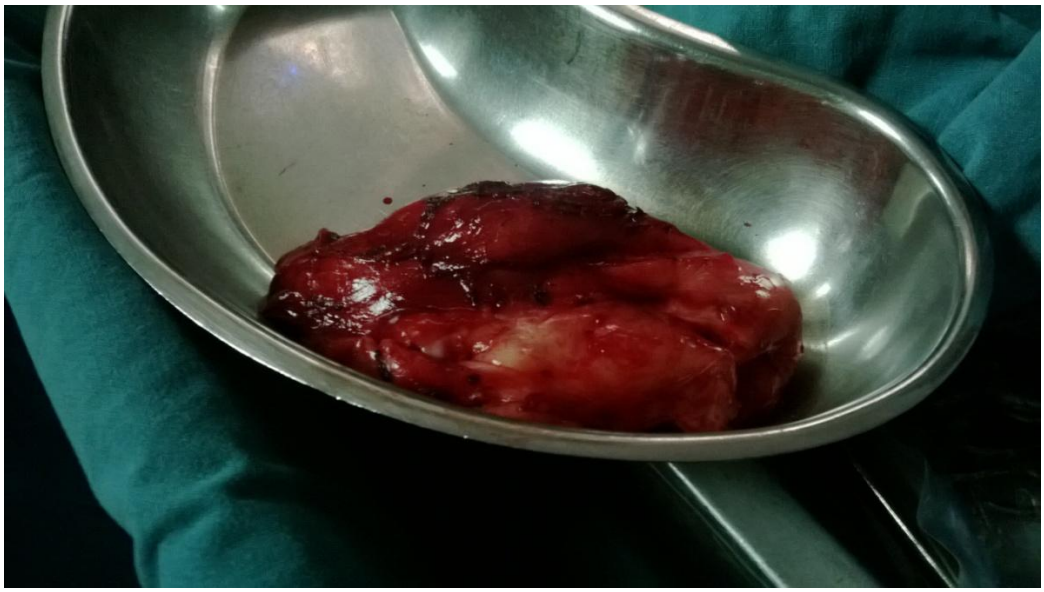


Figure 2: Resected tumor mass.



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