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Case Report

Mesenteric paraganglioma a rare mesenteric tumor

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ABSTRACT

Paragangliomas arise from chromaffin tissue, most commonly found in the Zuckerkandl body, the sympathetic plexus of the urinary bladder, the kidneys, the heart or in the sympathetic ganglia of the head or neck. Some paragangliomas have been described in the Gastrointestinal System, the majority of which were associated with the duodenum. Only a very select few were described to arise from the mesentery.

Keywords: CT Scan, Mesenteric tumor, Non-functional paraganglioma, Surgical excision

INTRODUCTION

Paraganglioma is a rare tumor known to arise from neural crest cell, that arises from sympathetic or parasympathetic neural paraganglia. The commonest location of this tumor is adrenal medulla- where it is known as Pheochromocytoma.[1,2] Though it commonly occurs in adrenal medulla a few 5-10% of these tumors can occur at extra adrenal sites, where normal paraganglia exist.[2] About 70-85% of sporadic Paraganglioma occur intraabdominally and that too adjacent to the aorta especially near the area of bifurcation of abdominal aorta.[3] The paraganglioma occurring in the mesentery of Proximal jejunum is an extreme rarity with only 13 cases reported in the literature.[4,5] Here study present a case report of such a rare disease entity. A 23-year-old male presented with an acute abdominal pain in whom an abdominal mass was discovered on ultrasonography. CT scan revealed a solid polylobulated mass in the retroperitoneum. Exploratory laparotomy revealed a Large multi-nodular tumor mass, which contained solid and cystic components. The excision of the mass as well as the surrounding intestine was done. Histopathological studies confirmed the presence of a paraganglioma.

CASE REPORT

A 23 years old healthy male presented with acute abdominal pain, on physical examination, Vague palpable mass in the upper abdomen, blood pressure was 124/82 mm. of Hg, pulse rate was 82/min. Ultrasonography revealed a lobulated mixed echogenic mass in left hypochondriac and lumber region of size 15.6 X 8.6 cms. CT scan confirmed -lobulated heterogeneous mass with solid and cystic components, in retroperitoneum and on either side of the superior mesenteric artery (SMA) and Superior mesenteric vein (SMV). CT guided FNAC tried but was inconclusive. For a definitive diagnosis, surgical resection was recommended. Before surgery, although the differential diagnosis included gastrointestinal stromal tumors, leiomyoma, we could not definitively diagnose this tumor.

Exploratory laparotomy was done and multilobulated mass of about 16 x 10 cms in the mesentery of the proximal jejunum was seen lying on either side of the SMA and SMV there was another cystic mass on the duodenal wall 2nd part and on to the pancreatic head. The mass was excised with a 30 cms. segment of jejunum,

bowel continuity restored by enteroenteric anastomosis. Postoperative period was uneventful.

Histopathology - revealed mesenteric paraganglioma with confirmation by Immunohistochemistry (IHC).

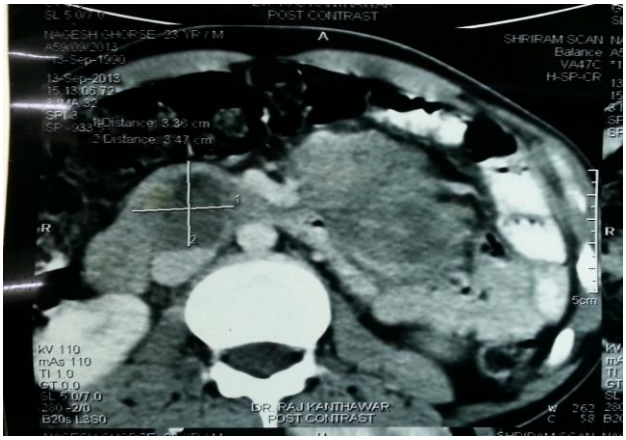

Figure 1: Huge mesenteric mass encircling the mesenteric vessels.

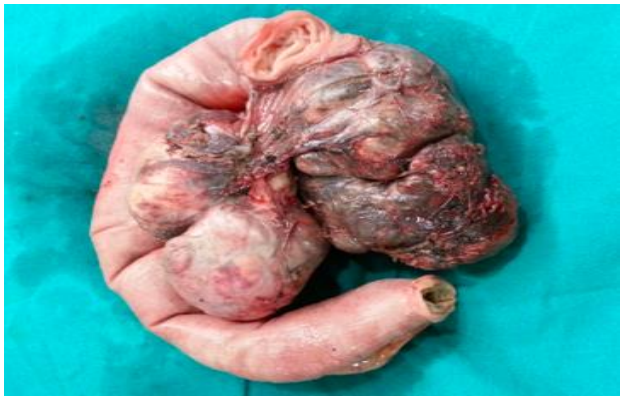

Figure 2: Macroscopic features-excised jejunal mesenteric mass of size 16cms x 10cms with loop of Jejunum.

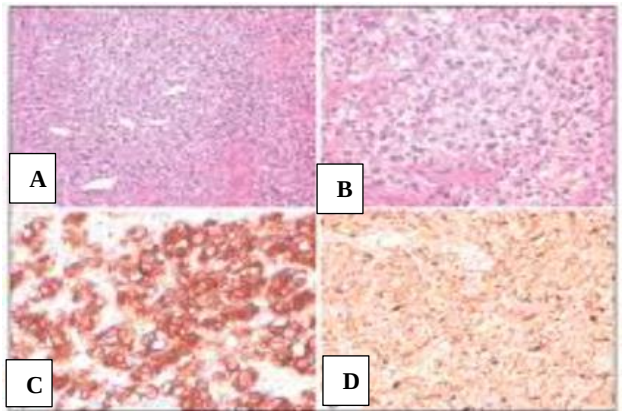

Figure 3: A) H and E Stain Normal; B) H and E stain 200 X magnification; C) Chromogranin A 400 X magnification; D) CD-100 200 X Immunohistochemistry confirmed the diagnosis.

DISCUSSION

The Diagnosis in cases of nonfunctional Paraganglioma is difficult, in contrast to the functional tumors were features like Sustained and episodic hypertension, Pyrexia of unknown origin, palpitation, headache, make investigations and diagnosis easy.[4-6] So when functional Paraganglioma is suspected, biochemical analysis of catecholamine hyper secretion should precede any form of imaging.[7,8]

Literature suggests that females are more sufferers 9:3, which contrasts with the slight male predominance (1.3:1) reported for retroperitoneal paraganglioma.[6,7] At the time of diagnosis, most patients are older (median, 57.5 years of age) than those with retroperitoneal Paraganglioma (median, 39-43 years of age).[6,7] No significant difference was noted in the size of mesenteric (average, 9.3 cm) and retroperitoneal tumors (average, 7.4-10.5 cm).[6]

The disease pathogenesis is not clearly understood, They are described as hereditary in around 10%-50% cases and out of these 20%-50% are considered multicentric but in Sporadic type, only 10% are multicentric.[2,3] Those occurring at an earlier age may be multicentric and may be considered as Hereditary, So family history is important while in elderly patients the disease has no genetic background.[4] For this reason, especially in patients diagnosed before 50 years of age and in those who present with bilateral, multifocal, and malignant Paraganglioma, genetic testing may be beneficial.[8]

In contrast to adrenal paraganglioma most of extra-adrenal paraganglioma are nonfunctional, as happened in our case it was an incidental detection, where the presentation was pain in abdomen for which ultrasound of the abdomen was done and mass was diagnosed, it was followed by CT scan of abdomen which was not suggestive of Paraganglioma even the FNAC or Guided biopsy was not very conclusive.[8,9]

In study case, preoperative imaging played an important role to determine tumor localization, vascularity, and extent of disease including any metastasis. We entertained differential diagnosis including gastrointestinal stromal tumors, leiomyoma, Lympho proliferative disorder, and other metastatic tumor, in the mesentry.[8,11] However, pitfall for misdiagnosis in our case was tumor location. Because of the tumor location away from the para-aortic area, a preoperative diagnosis of paraganglioma could not be made.

Functional paraganglioma the best investigation to confirm the preoperative diagnosis is 131I-metaiodobenzylguanidine (MIBG) scan. MIBG scan may also be helpful to rule out clinically silent cases, in some cases, FDG-PET may be indicated to investigate metastatic disease. It was recently reported that the newest technique using fluorine-18-

dihydroxyphenylalanine-PET imaging offers even higher accuracy than MIBG scintigraphy in the localization of paraganglioma.

Considering the size and the malignant potential, the incidence of malignant change reportedly ranges from 14% to 50%.[15,16] In these reports, the clinical and histological distinction between benign and malignant tumors was unclear, and the definitive diagnosis of malignancy was based solely on the presence of metastases.

The mainstay of treatment for paraganglioma is surgical resection. Most tumors were excised along with a segment of small bowel, probably because of the large tumor size and intestinal vascularity. From the viewpoint of lymph node dissection, however, recurrence in cervical lymph node was reported for retroperitoneal paraganglioma.[5] neither local nor distant lymph node metastasis was reported for mesenteric paraganglioma.

CONCLUSION

Occurrence of paraganglioma in the jejunal mesentery is extremely rare, preoperative diagnosis is seldom possible in nonfunctional tumor. The localization, extent, can be very well known by CT, Or MRI. Surgical resection is the mainstay of treatment while chemotherapy and radiotherapy have both been proven ineffective in previous cases. The diagnosis may be confirmed by Immunohistochemistry. It is difficult to predict the malignant potential of these tumors even on Histopathology. Recurrence of mesenteric Paraganglioma has not been reported, still careful long-term follow-up after surgical excision is necessary.

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