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Abstract- Extraadrenal paraganglioma constitute only 10%. Among them paraganglioma outside usual distribution is less than 1%. Most of the cases are solitary and sporadic. Familial cases are also seen. They are associated with several syndromes. Extra-adrenal paragangliomas are rarely diagnosed preoperatively unless the lesion is functional. Paraganglioma of mesentery of small intestine are very rare. Till date 10 cases are reported and this is the eleventh case. This is the second case where the paraganglioma is in the anterior aspect of the mesentery of small intestine and 1st reported case involving the wall of jejunum. Our case is the youngest reported till date. We hereby report a case of 23 years male patient who presented to us with discomfort and mass per abdomen. Investigations revealed a mass in the mesentery of small bowel. Provisional diagnosis was GIST of jejunum. Laparotomy was performed mass was resected, histopathology and IHC revealed the diagnosis. The patient is followed up till date and there is no evidence of recurrence. Most of them are clinically benign, but prediction of behaviour is difficult. Surgical resection is the treatment for both benign and malignant paragangliomas. Adjunctive therapies like Radiotherapy can be considered palliative in malignant cases and unresectable cases. This condition should be considered as differentials of any solid tumours at this site to prevent disasters peroperatively in case of catecholamine producing tumours.

Keywords: mesenteric tumours, adrenal and extraadrenal paragangliomas, carney's triad, zellballan pattern, chromogranin, synaptophysin CD 56, S 100, mib-1 labelling index.

I. INTRODUCTION

Solid tumours arising from the mesentery of the small bowel are rare and are usually metastatic^{1,2,3}. Only a few are primary tumours. Paragangliomas are considered in the differential diagnosis of the solid primary tumours in the mesentery of small bowel^{1,3}. Paragangliomas of mesentery are rare neuroendocrine tumours⁴. Still rare are their location in the anterior mesentery adjacent to the small bowel wall⁵.

II. CASE REPORT

A 23 years old male patient who was moderately built and nourished reported to our hospital with discomfort in the upper abdomen and a vague mass in the epigastric and left hypochondriac region. On examination blood pressure and pulse were within normal range. A mass of around 10cm x10cm was noted in the left hypochondrium. Its borders were ill defined the upper border could not be felt. Finger insinuation was possible. The mass was not moving with respiration. There was no hepatomegaly or ascitis. There was no lymphadenopathy. Other systemic examination was normal.

Ultrasound abdomen revealed a mass at the tail of pancreas, no mesenteric lymph nodes and no ascitis.

CT scan of abdomen revealed enhancing mesenteric soft tissue lesion with Central necrosis (Pic1).

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Picture : 1

A GIST arising from the wall of small bowel was suspected. Patient was planned for Diagnostic Laparoscopy. A mass was noted in left hypochondriac region in the mesentery of jejunum with dilated vessels on its surface and a loop of jejunum stretched out on its surface. Mass lying anterior to tail of pancreas. Conversion to open surgery was done. With left subcostal incision abdomen was opened (Pic 2). A globular mass

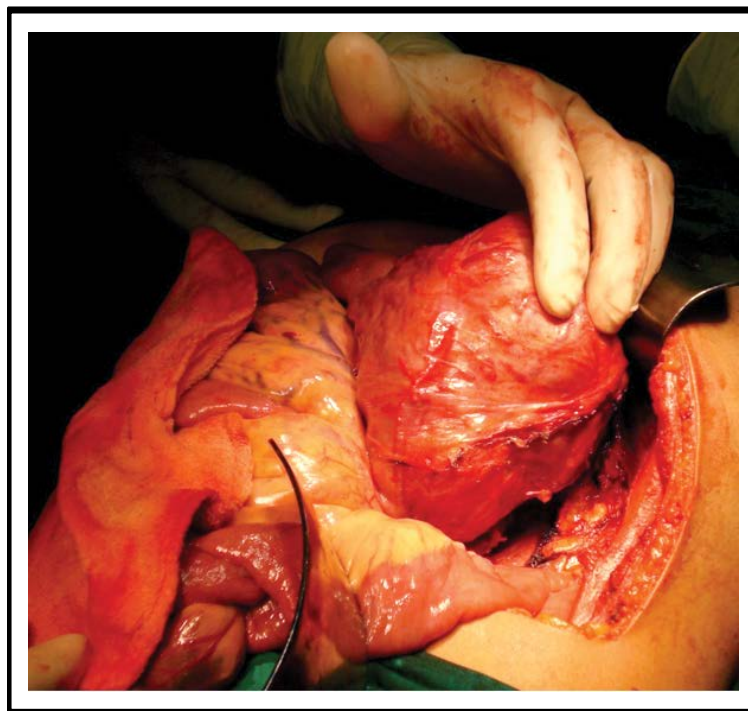
around 10cm x10cm x9cm was noted in the mesentery of jejunum involving the wall (Pic 3, 4 & 5). Resection of the segment of jejunum was done along with the mesentery and the mass. Jejunojunctional anastomosis was done (Pic 6). Patient had uneventful post operative recovery and he was discharged on 9th postoperative day.



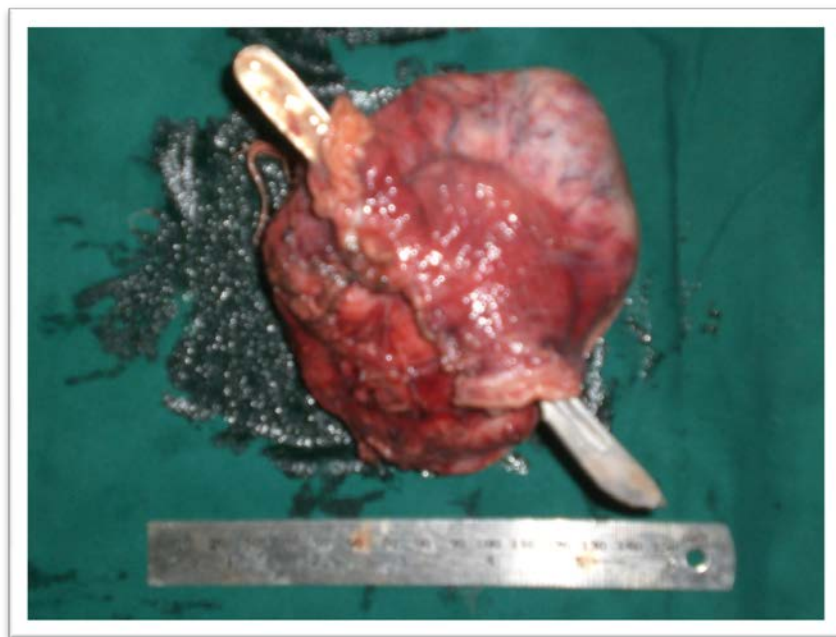
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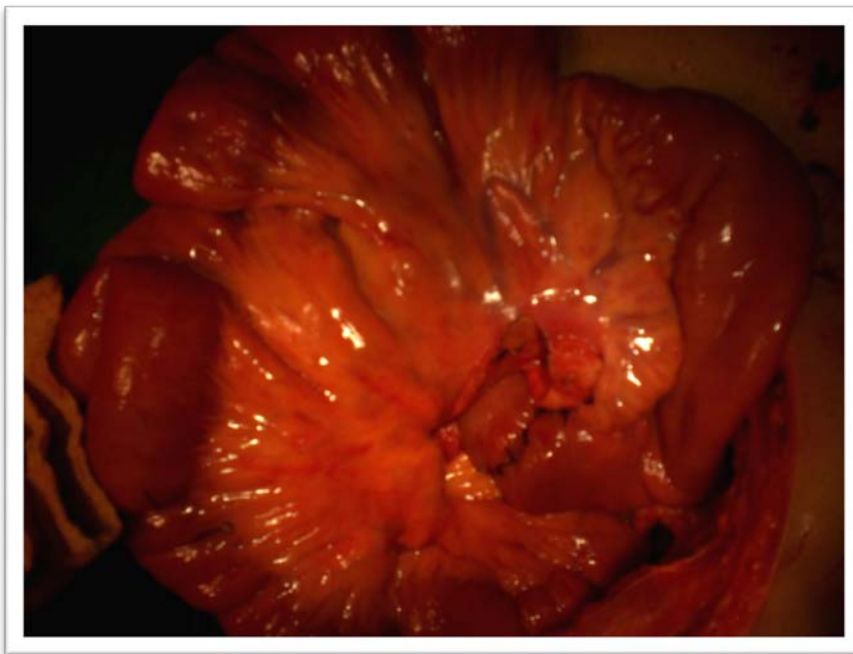
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Picture : 5



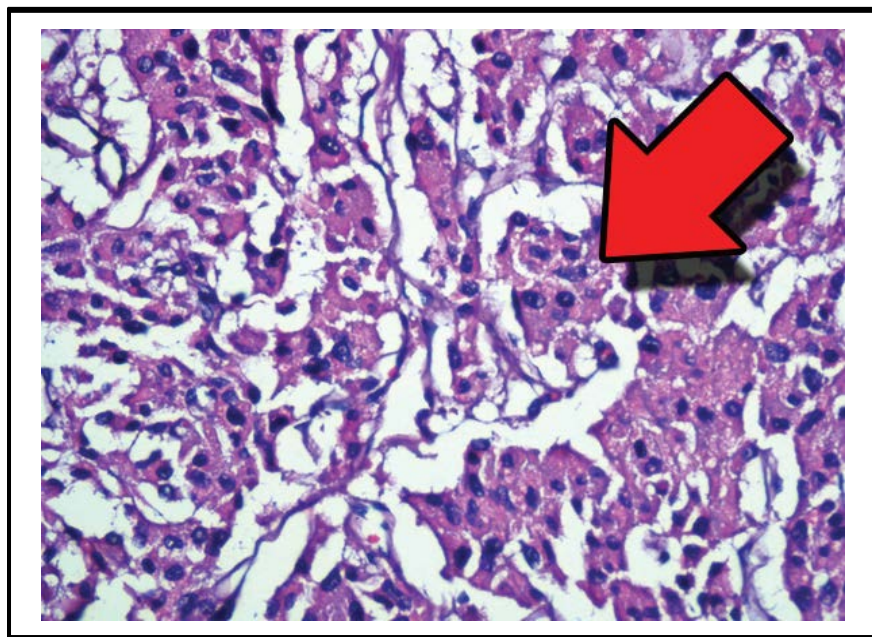
Picture : 6

Morphologically the tumour was well encapsulated and was highly vascular (Pic 4). It was attached to the wall of Jejunum measuring 8cm x 9cm x 10cm. Cut section showed well circumscribed grey white area with central area of haemorrhage and myxoid changes (Pic 7).



Picture : 7

Histopathology revealed nests and groups of tumour cells separated by fibrovascular connective tissue in Zellballen pattern (Pic 8).



Picture : 8

Immunohistochemistry done concurred with HPE reports stating Synaptophysin, Chromogranin and CD56 positive, Mib-1 labeling index is 0 -1%, S-100 protein staining sustentacular cells were also positive.

Since Mib-1(Ki-67) labelling index is low our patient is at low risk for malignancy. The patient is

followed up every three months till date (28 months). Patient is asymptomatic, vitals are measured to look for transformation into functional tumour and serial ultrasound evaluation of the abdomen is done to look for recurrence. There is no significant evidence for recurrence or functional transformation.

III. DISCUSSION

Paraganglioma is a rare neuroendocrine neoplasm^{1,2,3}. According to WHO classification of neuroendocrine neoplasm they arise from the chromaffin negative cells derived from embryonic negative cells⁶. They belong to Group II tumours as categorized by Wick in updated terminology for neuroendocrine neoplasm⁷.

About 75% cases are sporadic and the rest 25% are hereditary^{1,4}. In hereditary paragangliomas the lesions are multiple, aggressive and appear at early age⁴. They are usually associated with mutation of genes like Succinate Dehydrogenase (SDHB, SDHC, SDHD called Carney Stratakis Syndrome.)^{1,4}, MEN 2a & 2b⁴, VHL⁴; NF-1⁴ and syndromes like Carney's triad includes gastrointestinal stromal tumor, pulmonary chondroma, and extra-adrenal paraganglioma⁸.

In the fetal life, paraganglionic tissue is derived from pheochromoblasts, highly concentrated at a level extending from the root of the inferior mesenteric artery or the renal artery to the aortic bifurcation, known as the organ of Zuckerkandl. In the adults the neoplasms arising from paraganglia occur most frequently in the adrenal medulla, where they are called as pheochromocytoma. The remaining small amount of normal paraganglionic tissue occurs in extra-adrenal sites extending from the upper cervical region to the pelvis, paralleling the autonomic nervous system. Neoplasms derived from paraganglionic tissue in these sites are known as paragangliomas⁵. Paraganglioma can arise from carotid and aortic bodies, organ of zuckerkandl and other unnamed paraganglia in the distribution of sympathetic and parasympathetic outflow^{4,9}. Small amounts of paraganglionic tissue have been described

in sites outside the conventional distribution such as in the interatrial septum of the heart, liver hilus, gallbladder, urinary bladder, prostatic capsule, and mesenteric vessels potentially capable of giving rise to paragangliomas in these unusual sites¹⁰. Extraadrenal paragangliomas outside the distribution of the autonomic nervous system, where normal paraganglia have not been described can probably be explained by the migratory property of the neural crest cells during embryogenesis along the blood vessels^{3,4}. Under the Glenner and Grimley classification these paraganglia may qualify as the viscer-autonomic type, a group of poorly defined paraganglia that occur in association with visceral organs¹¹.

Approximately 5% to 10% of sporadic cases occur in extra adrenal sites⁴. 70% to 80% of these cases are intraabdominal most common being adjacent to aorta⁴. Paraganglioma of mesentery of small intestine is extremely rare with only 10 cases reports till date. Our case is the eleventh case with paraganglioma in anterior part of the mesentery involving the bowel wall. This is the second reported case with paraganglioma in the anterior mesentery. This is the first reported case involving the bowel wall.

The previously reported cases with paraganglioma in the mesentery of small intestine are as shown in the Table-1. Most of the cases were above the age of 60 yrs. And our case is the youngest, earliest presentation at 23 years. Most of patients were females (F:M;;7:4). Except for the case report by Jaffer et al all other cases are posterior mesenteric paragangliomas⁵. Our case is the second reported anterior mesenteric paraganglioma of small bowel. All cases were managed with resection of the bowel with involved mesentery and mass followed by anastomosis.

Table 1 : Clinical information about the previous reported cases

Sl. No.	Reference	Age	Sex	Tumour location in mesentery	Symptoms	Size of tumour (cms)	Htn	Pre Op diagnosis	Surgery	Prognosis
1	Arean et al. ¹²	32	M	Small intestine. (Posterior)	Nausea, vomiting, diarrhea.	10x7x6	-	Abdominal mass.	Resection and anastomosis	8 m. Alive No recurrence
2	Carmichael et al. ¹³	62	F	Small intestine. (Posterior)	Nausea, vomiting, back ache.	3.2	+	Abdominal mass.	Resection and anastomosis	Not documented
3	Onoue et al. ¹⁴	38	F	Small intestine. (Posterior)	None.	4.5x3.2	-	Mesenteric mass.	Resection and anastomosis	24 m. Alive No recurrence
4	Jaffer et al. ⁵	76	M	Small intestine. (Anterior)	Pain abdomen, vomiting, diarrhea.	8.5x8	+	Abdominal mass.	Resection and anastomosis	Not documented

5	Muzaffer et al. ¹⁵	76	F	Small intestine. (Posterior)	Mass in the abdomen	20x15	-	Abdominal mass.	No document.	15 m. Alive No recurrence
6	Ponsky et al. ¹⁶	35	F	Small intestine. (Posterior)	Mass in the abdomen	5.5	+	Abdominal mass.	Resection and anastomosis	24 m. Alive No recurrence
7	Kudoh et al. ¹⁷	72	F	Ileum (Posterior)	Pain abdomen and mass	10x9x9	-	Mesenteric tumour.	Resection and anastomosis	12 m. Alive No recurrence
8	Nobeyama et al. ¹⁸	53	M	Ileum (Posterior)	Pain abdomen and mass	15x10x7	-	Abdominal mass.	Resection and anastomosis	Not documented
9	Matsumoto et al. ¹⁹	77	F	Small intestine. (Posterior)	Pain abdomen and mass	7x5.5	-	Mesenteric tumour.	Resection and anastomosis	9 m. Alive No recurrence
10	Fujita T et al. ⁴	78	F	Small intestine. (Posterior)	None	3x1.5x1.5	-	Mesenteric tumour.	Resection and anastomosis	8 m. Alive No recurrence
11	Present study	23	M	Jejunum (Anterior)	Abdominal discomfort and mass	8x9x10	-	GIST of jejunum	Resection and anastomosis	28 m. Alive No recurrence

Most of the extra adrenal paragangliomas are asymptomatic or present as a mass and mass associated symptoms^{1,4}. 1% to 3% of them are functional due to catecholamines released and presents with paroxysmal episodic hypertension, palpitation, headache or profuse headache^{1,4}. If the extra adrenal paraganglioma is functional diagnosis is easy⁴ first investigation would be biochemical analysis of catecholamine in serum should be done before any imaging⁴.

Majority of cases are found incidentally in patients evaluated for other reasons⁴. CT features are nonspecific soft tissue density and these features are similar to any other neoplasm. Hence preoperative diagnosis of extra adrenal paraganglioma is usually difficult. MRI and Angiography may be useful to know the soft tissue involvement and vascularity of the tumour⁴. Though ¹³¹I Metaiodobenzyl guanidine (MIBG) scintigraphy⁴ is useful in functional tumours it can be used to rule out clinically silent cases. FDG-PET⁴ scan may be helpful to locate metastasis and is superior to MIBG scintigraphy. Mitotic figures and Ki-67 labelling index⁴ are of significance in malignant case grading.

Treatment is surgical resection^{1,4}. Most of the times significant mesentery has to be removed hence resection of the bowel is inevitable and anastomosis is required. Adjunctive therapies like Radiotherapy and chemotherapy can be considered palliative in malignant

cases and unresectable cases². Patients with malignant paraganglioma respond to multikinase inhibitor SUNITI NIB MALATE²⁰.

Histologically, paragangliomas have well-defined characteristics. The lesions are composed of cell balls (Zellballen) separated by thin fibrovascular septa¹. These cell balls are composed of two types of cells chief cells and sustentacular cells¹. Other patterns are angiomatoid, fusocellular and clear cell. Some paragangliomas show intense fibrosis, which can compress and distort the cell balls, giving rise to a pseudoinfiltrative appearance (sclerosing paraganglioma)¹.

With immunohistochemistry the chief cells are positive for neuroendocrine markers (neuron specific enolase, chromogranin A, synaptophysin, serotonin) while sustentacular cells are positive for S-100 protein¹.

Cervical lymphnode metastases are noted in retroperitoneal paragangliomas. 15%-40% of extra adrenal paragangliomas undergo malignant transformation⁴. In these cases clinical and histological distinction between benign and malignant tumours is difficult. Only presence of metastasis can prove malignancy.

Neither local nor distant metastasis are reported in paragangliomas of mesentery of small intestine till date. In retroperitoneal tumours 5yrs and 10yrs survival rates are 75% and 45% respectively⁴. But the prognosis for paraganglioma in the mesentery of small intestine is

not available. Recurrence of mesenteric paraganglioma is not reported till date. However long term follow up after surgical resection is advised⁴.

IV. CONCLUSION

Paraganglioma of mesentery of small intestine is a very rare disease. Our case is the 11th case reported, 2nd case with anterior mesenteric paraganglioma of jejunum and 1st reported case involving the wall of jejunum. This is youngest patient reported with this tumour till date. It is difficult to diagnose a case of paraganglioma in the mesentery of small intestine preoperatively. Hence it should be considered in the differentials when managing a patient with a solid tumour in mesentery. It also prevents disasters preoperatively in case of catecholamine producing tumours^{1,3}. Postoperatively patient should be followed up for very long periods to look for recurrence⁴.

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