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Paraganglioma of Mesentery of Jejunum-A Case Report and Review of Literature

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Abstract

Extraadrenal paraganglioma constitute only 10

Index terms— mesenteric tumours, adrenal and extraadrenal paragangliomas, carney?s triad, zalbellan pattern, chromogrannin, synaptophysin CD 56, S 100, mib-1 lab

1 Introduction

olid tumours arising from the mesentery of the small bowell 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 tumours 4 . Still rare are their location in the anterior mesentery adjacent to the small bowel wall 5 .

2 II.

3 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 leision with Central necrosis (Pic1). Immunohistochemistry done concurred with HPE reports stating Synaptophysin, Chromogranin and CD56 positive, Mib-1 labeling index is 0 -1%, S-100 protein staining sustentecular 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.

4 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 cells 6 . They belong to Group II tumours as categorized by Wick in updated terminology for neuroendocrine neoplasm 7 .

About 75% cases are sporadic and the rest 25% are hereditary 1,4 . In hereditary paragangliomas the leisions are multiple, aggressive and appear at early age 4 . They are usually associated with mutation of genes like Succinate Dehydrogenase (SDHB, SDHC, SDHD called Carney Stratakis Symdrome.) 1,4 , MEN 2a & 2b 4 ,VHL 4 ; NF-1 4 and syndromes like Carney's triad includes gastrointestinal stromal tumor, pulmonary chondroma, and extra-adrenal para gang-lioma 8 .

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 paragangliomas 5 . Paraganglioma can arise from carotid and aortic bodies, organ of zuckerkandle 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 sites 10 . 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 **??**,4. Under the Glenner and Grimley classification these paraganglia may qualify as the viscero-autonomic type, a group of poorly defined paraganglia that occur in association with visceral organs 11 .

Approximately 5% to 10% of sporadic cases occur in extra adrenal sites 4 . 70% to 80% of these cases are intraabdominal most common being adjacent to aorta 4 . 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 paragonglioma 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 paragangliomas 5 . 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 be anastamosis. 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 functio nal 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 easy 4 first investigation would be biochemical analysis of catecholamine in serum should be done before any imaging 4 .

Majority of cases are found incidentally in patients evaluated for other reasons 4 . 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 tumour 4 . Though I 131 Metaiodobenzyl guanidine (MIBG) schintigraphy 4 is useful in functional tumours it can be used to rule out clinically silent cases. FDG-PET 4 scan may be helpful to locate metastasis and is superior to MIBG schintigraphy. Mitotic figures and Ki-67 labelling index 4 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 anastamosis is required. Adjunctive therapies like Radiotherapy and chemotherapy can be considered palliative in malignant cases and unresectable cases 2 . Patients with malignant paraganglioma respong to multikinase inhibitor SUNITI NIB MALATE 20 .

Histologically, paragangliomas have welldefined characteristics. The lesions are composed of cell balls (Zellballen) separated by thin fibrovascular septa 1 . These cell balls are composed of two types of cells chief cells and sustentacular cells 1 . 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 paragangliom as) 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 protein 1 .

Cervical lymphnode metastases are noted in retroperitoneal paragangliomas. 15%-40% of extra adrenal paragangliomas undergo malignant transforma tion 4 . 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% respectively 4 . But the prognosis for paraganglioma in the mesentry of small intestine is is not reported till date. However long term follow up after surgical resection is advised 4 .

5 IV.

6 Conclusion

Paraganglioma of mesentery of small intestine is a very rare disease. Our case is the 11th case reported, 2nd case with anterior mesenteric paraganglio ma 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 peroperatively in case of catecholamine producing tumours 1,3 . Postoperatively patient should be followed up for very long periods to look for recurrence 4 . [1] [2]

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Figure 1: 1 A

[1] © 2014 Global Journals Inc. (US)
[2] © 2014 Global Journals Inc. (US) not available. Recurrence of mesenteric paraganglioma

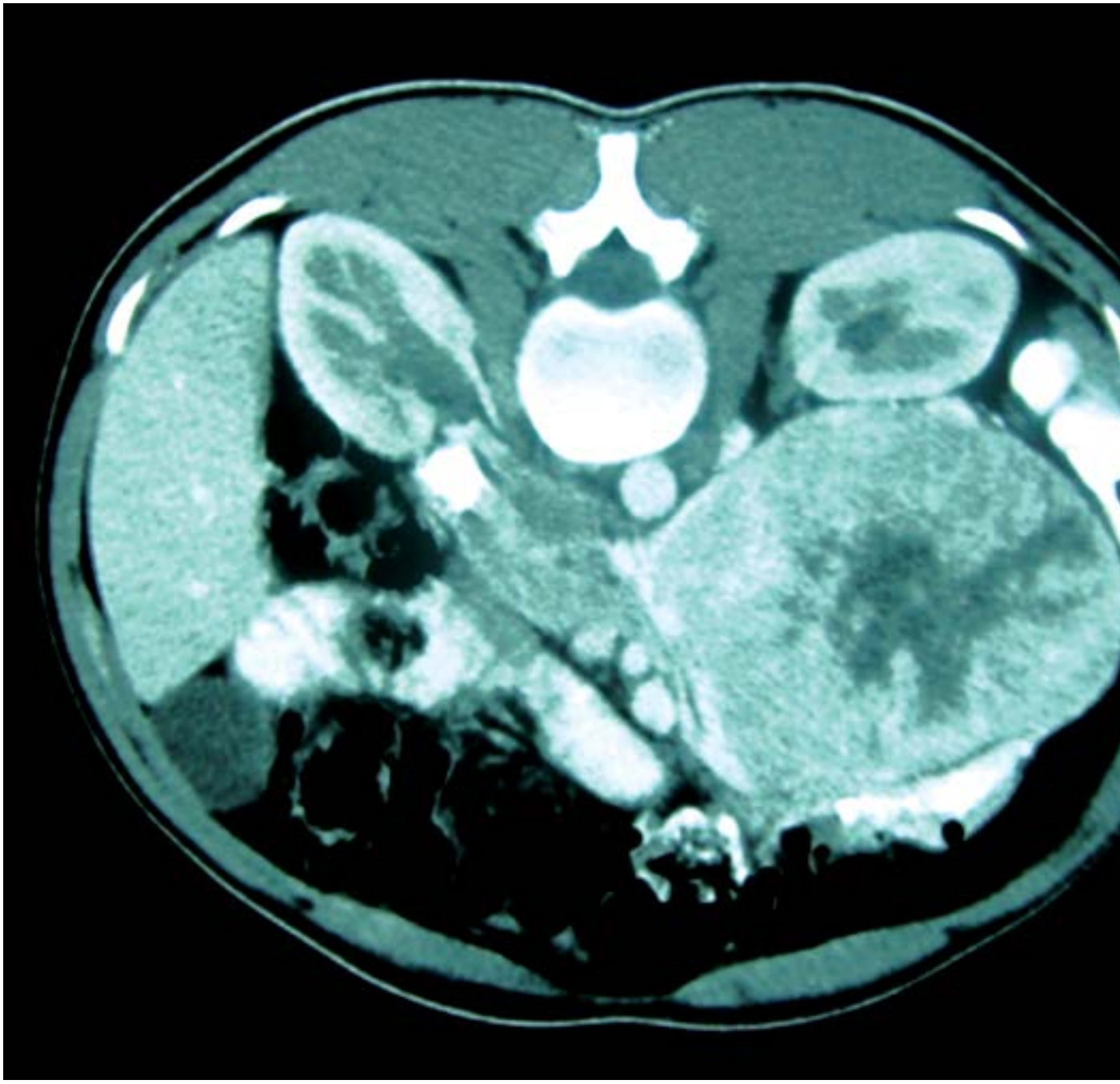


Figure 2:

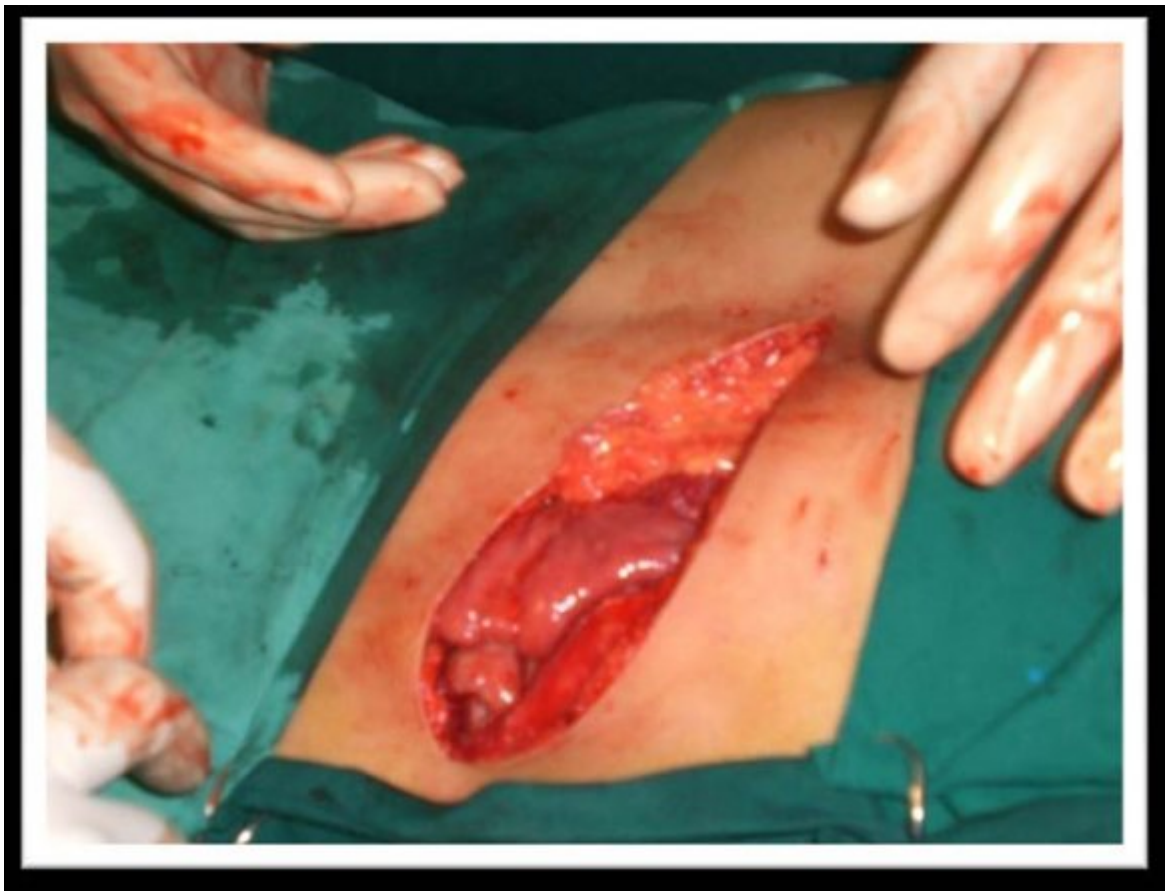


Figure 3:

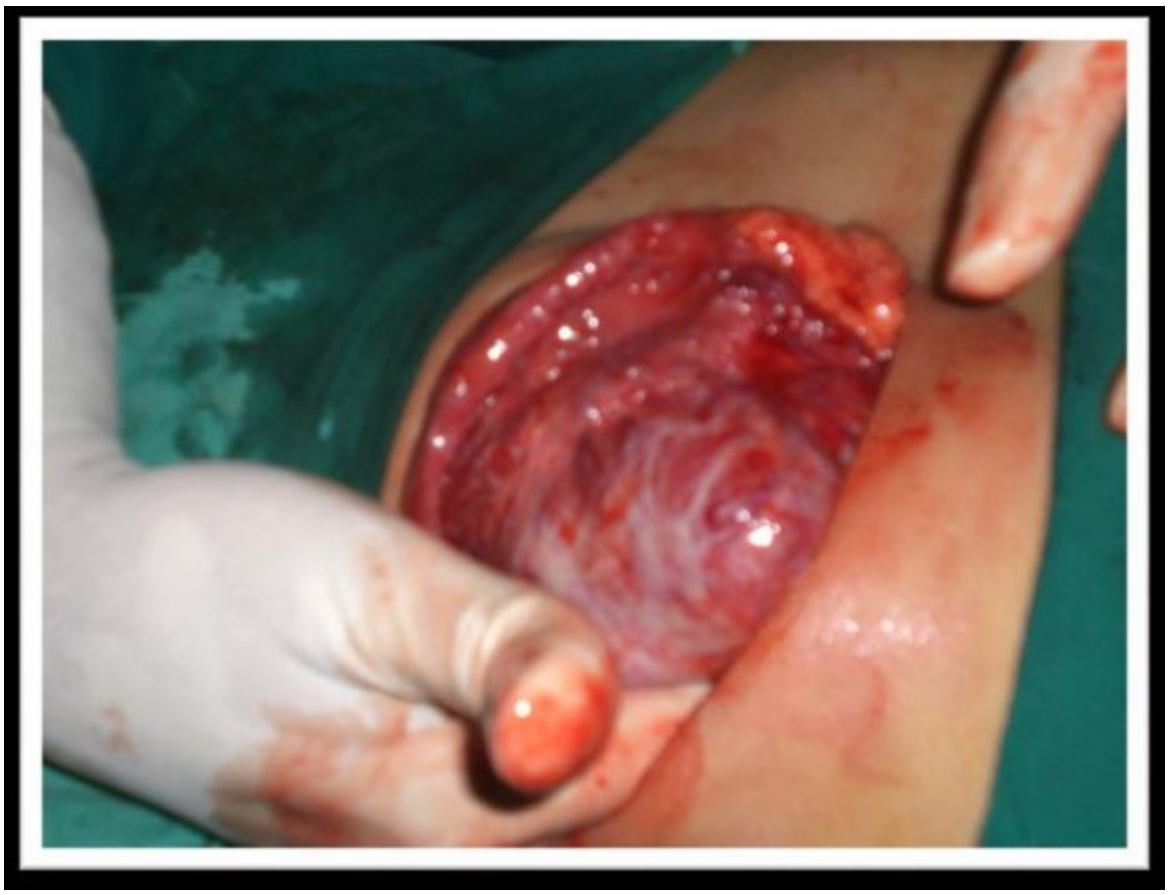
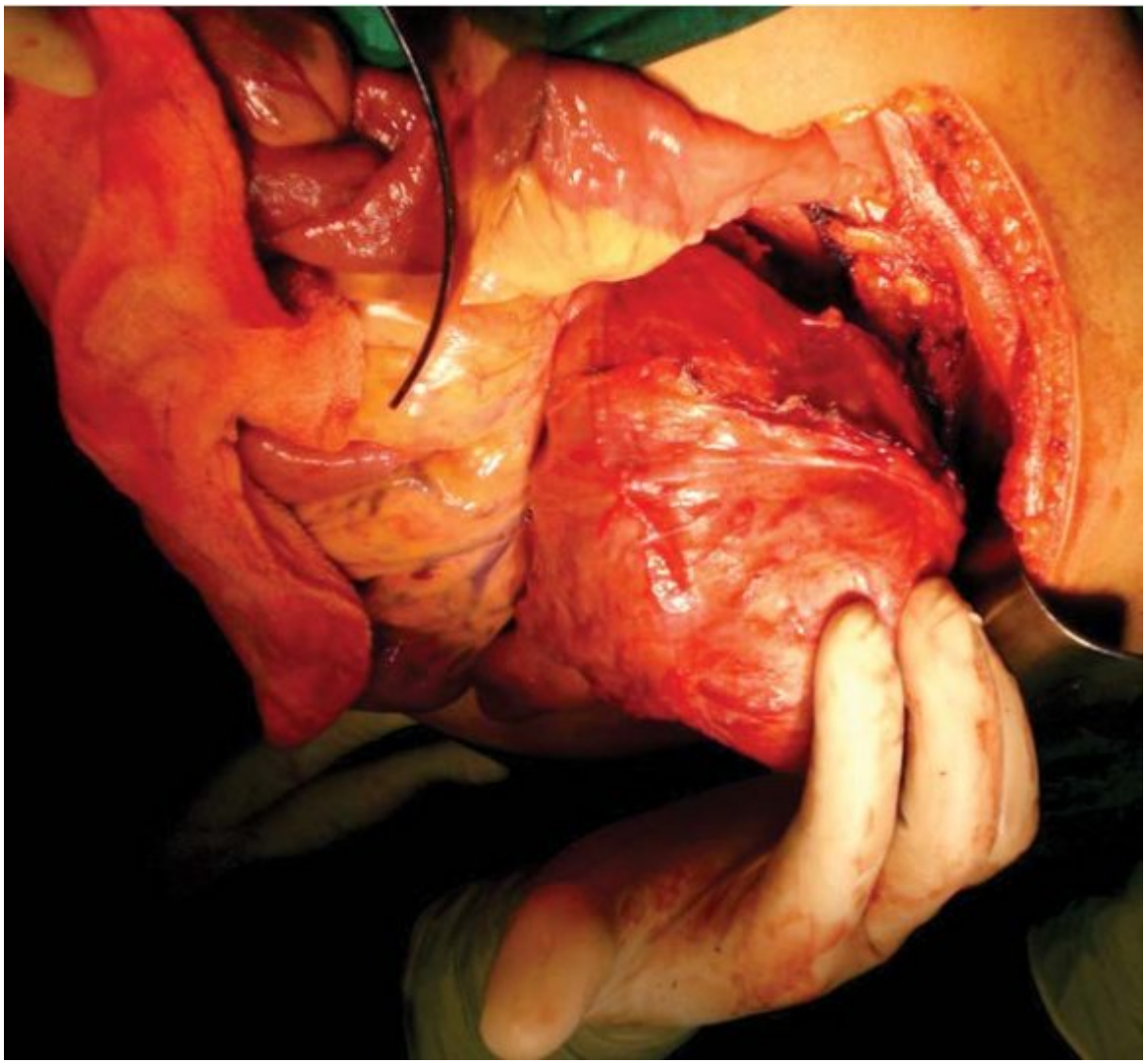


Figure 4:



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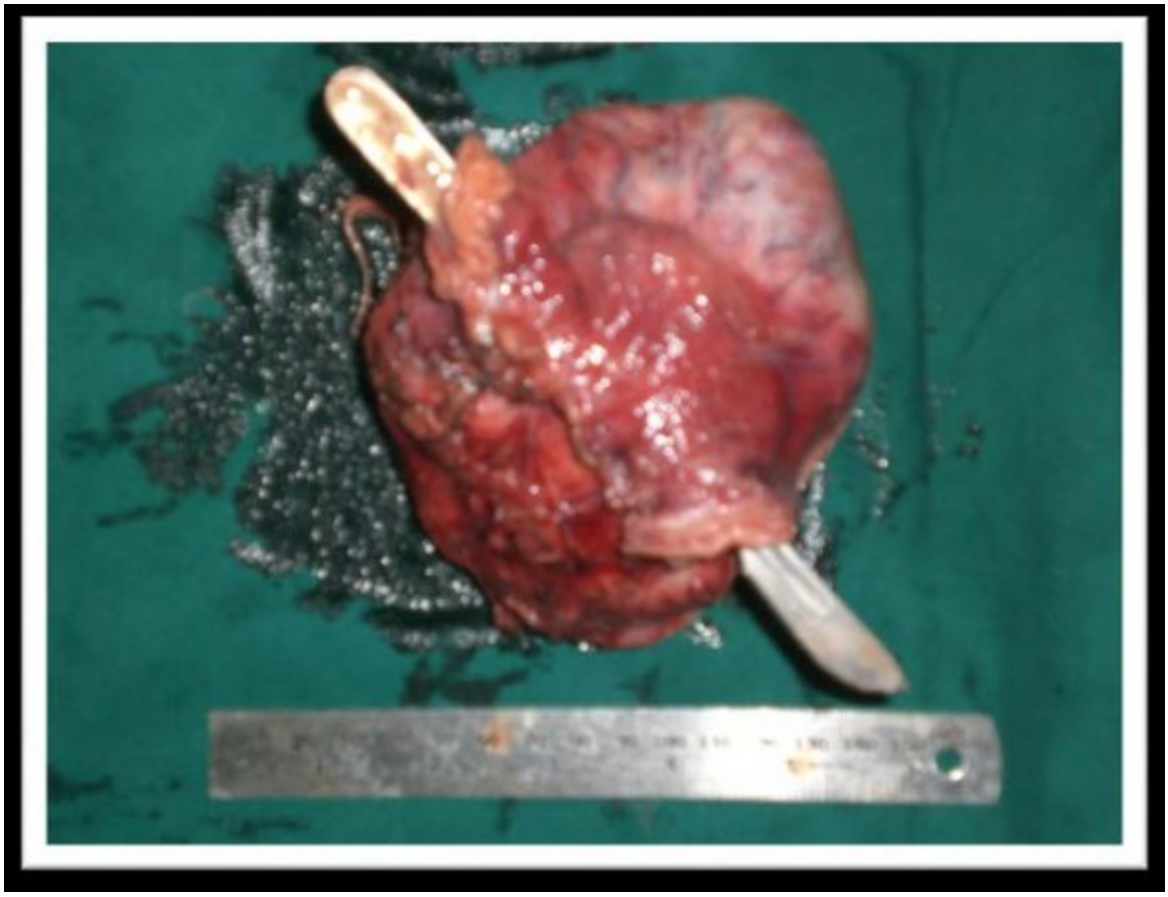

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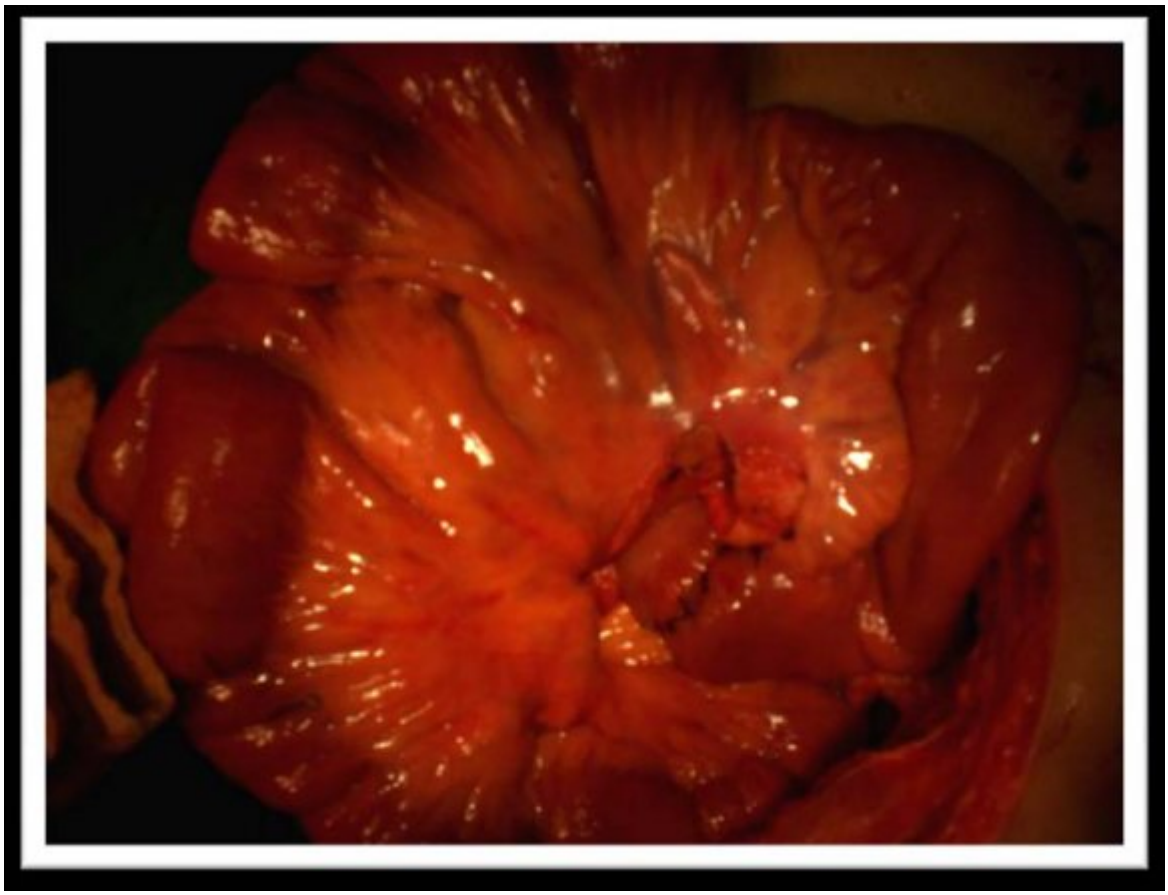

Figure 7:

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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 anastamosis	8 m. Alive No recurrence
2	Carmichael et al. 13	62	F	Small intestine. (Posterior)	Nausea, vomiting, bach ache.	3.2	+	Abdominal mass.	Resection and anastamosis	Not documented
3	Onoue et al. 14	38	F	Small intestine. (Posterior)	None.	4.5x3.2-		Mesenteric mass.	Resection and anastamosis	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 anastamosis	Not documented

Figure 8: Table 1 :

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