Retroperitoneal Paraganglioma: A Case Report

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Abstract:
Background: *Retroperitoneal paraganglioma is a rare tumor that develops from chromaffin cells that secrete catecholamines in the sympathetic ganglia. Universally known as pheochromocytoma, tumors located outside the adrenal gland have been designated as extraadrenal paragangliomas.*

Aim of the work*: The authors report the case of a retroperitoneal paraganglioma with analysis of clinical and imaging features and review of the relevant literature.*

Case report: *We report the case of 84-year-old woman with medical history of hypertension and who was complaining of vomiting and pain in the epigastrum and upper right quadrant. Physical examination and laboratory tests revealed no abnormalities. Ultrasound sonography and abdominal CT-scan showed a round well limited retroperitoneal lesion of c. 7 cm composed of cysts and plain tissue adjacent to the tail of the pancreas, the posterior surface of the stomach and the left renal vein. The Magnetic resonance Imaging (MRI) showed a lesion that was hypointense on T1 and heterogeneous hyperintense on T2. The preoperative diagnosis was tumor of the tail of the pancreas or the gastrointestinal stromal tumor (GIST) or the extra digestive stromal tumor (E-GIST). The patient was operated on via a midline incision. We had found a lesion - that was deeply embedded in the left transverse mesocolon near the duodeno-jejunal flexure. It was well encapsulated, surrounded by several peri-capsular veins having connection with the inferior mesenteric vein and the left renal vein. A complete tumor resection was performed.. The histopathological diagnosis was retroperitoneal paraganglioma without any signs of malignancy.*

Conclusion: *When confronted to a posterior abdominal mass, the first step is to be sure of its retroperitoneal origin. Then, the second step is to try to guess its nature and characterize it. When symptoms and signs are not clear, it is essential to know their most frequent location, in order to think of the diagnosis of paraganglioma. Surgery is the main treatment because of their malignant potential. The removal must be complete to be curative.*

Key-words*: retroperitoneum, pancreas, retroperitoneal paraganglioma.*

Introduction:

Retroperitoneal paraganglioma is a rare tumor that develops from chromaffin cells that secrete catecholamines in the sympathetic ganglia. The paraganglioma of the adrenal medulla, which is universally known as pheochromocytoma. Paragangliomas located outside the adrenal gland have been designated as extraadrenal paragangliomas.

The authors propose to describe the clinical and morphological characteristics of a retroperitoneal paraganglioma discovered in an 83 year old patient.

Case report:

A 84-year-old woman was admitted to our department of surgery with a diagnosis of retroperitoneal mass. Her medical history was significant for hypertension. She was complaining of pain in the epigastrum and upper right quadrant and vomiting. Physical examination and laboratory tests revealed no abnormalities. Ultrasound sonography showed a tumor of c. 7 cm near to the left side of the pancreas without any specific characteristics. Abdominal CT-scan showed a round well limited retroperitoneal lesion; composed of cysts and plain tissue adjacent to the tail of the pancreas, the posterior surface of the stomach and the left renal vein. The left adrenal gland seemed to be intact (Fig1a+b). The Magnetic resonance Imaging (MRI) showed a lesion that was hypointense on T1 and heterogeneous hyperintense on T2 (Fig2). The preoperative diagnosis was tumor of the tail of the pancreas or the gastrointestinal stromal tumor (GIST) or the extra digestive stromal tumor (E-GIST). The patient was operated on via a midline incision. We had found a

lesion - that was deeply embedded in the left transverse mesocolon near the duodeno-jejunal flexure. It was well encapsulated, surrounded by several peri-capsular veins having connection with the inferior mesenteric vein and the left renal vein (Fig. 3a+b). A complete tumor resection was performed without capsular opening (fig4a+b). The mobilization of the tumor caused hypertensive peaks that were monitored with the use of nicardipine during and after the surgery. The aftermath of the surgery was simple with normalization of blood pressure. Pathology concluded that it is a retroperitoneal paraganglioma without any signs of malignancy.

Discussion:

Paraganglia make up a dispersed neuroendocrine system near or in the autonomic nervous system, which has a roughly symmetric distribution with extension from the skull base down to the pelvic floor [1].The most conspicuous member of this system is known as the adrenal medulla. Extraadrenal paraganglia, defined as paraganglia located outside the adrenal gland, can be divided in two broad groups: paraganglia associated with the parasympathetic system and those related to the sympathetic system [2].
Extraadrenal paragangliomas of the abdomen arise predominantly from paraganglia located in the retroperitoneum. They are known to play a role in the production and secretion of catecholamines causing rapid physiologic changes, and they probably represent lesser homologues of the adrenal medulla [1]. Of the paraganglionic tissues adjacent to the abdominal aorta, the most prominent collection is seen like in our patient; near the origin of the inferior mesenteric artery, which is known as the organs of Zuckerkandl [3, 4]. Men are affected more frequently than women, and most patients are between the ages of 30 and 45 years [5, 6]. Our patient is a 84-year-old woman.

Retroperitoneal paragangliomas are functional in up to 60% of patients [2, 6- 10]. Patients commonly present with symptoms related to excess secretion of catecholamine, such as palpitations, headache, sweating, and hypertension [11].For our patient, she was known to be hypertensive and the mobilization of the tumor caused hypertensive peaks that were monitored with the use of nicardipine during and after the surgery.

For those patients with nonfunctional extraadrenal retroperitoneal paragangliomas, diagnosis usually depends on nonspecific factors related to the growth of a retroperitoneal mass [9, 12,13]. In addition, approximately 10% of paragangliomas are clinically silent and detected incidentally at imaging study during evaluation of patients with unrelated symptoms [3, 14].

Most paragangliomas are solitary and arise sporadically; but they can be multicentric in approximatively 10 % of cases; and familial occurrence is well recognized [15, 16]. Thus, a careful family history is required. In general, the mode of transmission is known as an autosomal-dominant pattern with incomplete penetrance [17]. These familial tumors are commonly associated with these conditions: multiple endocrine neoplasia (MEN IIA and IIB) and neuroectodermal syndromes (tuberous sclerosis, neurofibromatosis, and von Hippel-Lindau disease). Paragangliomas may also occur as part of Carney's triad, which consists of gastric stromal tumors, pulmonary chondroma, and extraadrenal paraganglioma [18].

On contrast-enhanced CT scans, these tumors appear as paraaortic soft-tissue masses with either homogeneous enhancement or central areas of low attenuation. Smaller tumors are more likely to be homogeneous in attenuation and sharply marginated as compared with larger ones [19]. Punctate calcification or focal areas of high attenuation caused by acute hemorrhage may be seen in some tumors [19].

These tumors are usually hypointense or isointense compared with the liver parenchyma on T1-weighted MRI and are markedly hyperintense on T2-weighted MRI [20, 21]. Because of superior tissue characterization and absence of radiation hazard, MRI is recommended as the technique of first choice in evaluating patients with suspectedparagangliomas [20].

MIBG scintigraphy provides useful functional information and can be used for the detection of multiple primary tumors, tumors outside the usual locations, or metastases [20, 21]. MIBG scanning offers very good specificity (95–100%) but suffers from relatively imperfect sensitivity (85%) [22].

Radical surgery is the mainstay of treatment with radical resection possible in 75% of cases [23]. Choosing between conventional or laparoscopic surgery remains controversial due to the undeniable side effects of laparoscopy. Microscopically, all paraganglia have a similar morphologic appearance characterized by well-defined cell nests ("Zell-ballen"). The criteria for malignant paraganglioma on the basis of histopathology are not well defined, except the presence of documented metastases. A well-developed vascular network is present in most paragangliomas, and in some tumors vascular ectasia or an arborizing pattern may be found like in our case. Because of their hypervascular nature, biopsy of paragangliomas can result in significant bleeding [7]. The incidence of malignant paragangliomas, based on extensive local invasion or metastases, varies according to the series, from a low of 2% to a high of 36% [8, 9]. The frequent sites of metastases are regional lymph node, bone, liver, and lung.

We might resort to complementary therapies such as chemotherapy or radiation therapy in metastatic forms with a positive response in c. 50% of the cases but without any significant influence on prognosis. Surgery is the only mean that allows a significant improvement of prognosis with a five-year and 10 –year survival rate without relapse of 75% and 45% respectively. The median survival duration is c. 3 years in metastatic forms and 4 years in the case of incomplete resection [23].

Conclusion:

When confronted to a posterior abdominal mass, the first step is to be sure of its retroperitoneal origin. Then, the second step is to try to guess its nature and characterize it. When symptoms and signs are not clear, it is essential to know their most frequent location, in order to think of the diagnosis of paraganglioma. Surgery is the main treatment because of their malignant potential. The removal must be complete to be curative.

References:

1. Lack EE. Paraganglioma. In: Sternberg SS, ed. Diagnostic surgical pathology, 2nd ed. New York, NY: Raven Press, 1994:599–621

2. Rosai J. Ackerman's surgical pathology, 8th ed. New York, Mosby, 1996:1015–1058

3. Saurborn DP, Kruskal JB, Stillman IE, Parangi S. Best cases from the AFIP: paraganglioma of the organs of Zuckerkandl. Radio Graphics 2003; 23:1279–1286

4. Glenn F, Gray GF. Functional tumors of the organ of Zuckerkandl. Ann Surg 1976; 183:578–586

5. Mikhail RA, Moore JB, Reed DN Jr, Abbott AR. Malignant retroperitoneal paragangliomas. J Surg Oncol 1986; 32: 32-36.

6. Enzinger FM, Weiss SW. Paraganglioma. In: Enzinger FM, Weiss SW. eds. Soft tissue tumors. St. Louis: Mosby, 1988:836-860.

7. Liew SH, Leong AS, Tang HM. Tracheal paraganglioma: a case report with review of the literature. Cancer 1981; 47:1387–1393

8. Nora JD, Hallett JW Jr, O'Brien PC, Naessens JM, Cherry KJ Jr, Pairolero PC. Surgical resection of carotid body tumors: long-term survival, recurrence, and metastasis. Mayo Clin Proc 1988; 63:348–352

9. O'Riordain DS, Young WF Jr, Grant CS, Carney JA, van Heerden JA. Clinical spectrum and outcome of functional extraadrenal paraganglioma. World J Surg 1996; 20:916–921

10. Lack EE, Cubilla AL, Woodruff JM, Lieberman PH. Extraadrenal paragangliomas of the retroperitoneum: a clinicopathologic study of 12 tumors. Am J Surg Pathol 1980;4 :109-120

11. Erickson D, Kudva YC, Ebersold MJ, et al. Benign paragangliomas: clinical presentation and treatment outcomes in 236 patients. J Clin Endocrinol Metab 2001; 86:5210–5216

12. Law NW, Alfano L. Non-functioning retroperitoneal paraganglioma. J R Soc Med 1987; 80:246-247

13. Kryger-Baggesen N, Kjaergaard J, Sehested M. Nonchromaffin paraganglioma of the retroperitoneum. J Urol 1985; 1

14. Dunnick NR, Korobkin M. Imaging of adrenal incidentalomas: current status. AJR 2002; 179:559–568

15. Magliulo G, Zardo F, Varacalli S, D'Amico R. Multiple paragangliomas of the head and neck. An Otorrinolaringol Ibero Am 2003; 30:31–38

16. Grufferman S, Gillman MW, Pasternak LR, Peterson CL, Young WG Jr. Familial carotid body tumors: case report and epidemiologic review. Can- cer 1980; 46:2116–2122

17. Young AL, Baysal BE, Deb A, Young WF Jr. Familial malignant catecholamine-secreting paraganglioma with prolonged survival associated with mutation in the succinate dehydrogenase B gene. J Clin Endocrinol Metab 2002;

18. Carney JA, Sheps SG, Go VL, Gordon H. The triad of gastric leiomyosarcoma, functioning extra-adrenal paraganglioma and pulmonary chondroma. N Engl J Med 1977; 296:1517–151887:4101–4105

19. Hayes WS, Davidson AJ, Grimley PM, Hartman DS. Extraadrenal retroperitoneal paraganglioma: clinical, pathologic, and CT findings. AJR 1990; 155:1247–125034:536-538

20. van Gils AP, Falke TH, van Erkel AR, et al. MR imaging and MIBG scintigraphy of pheochromocytomas and extraadrenal functioning paragangliomas RadioGraphics 1991; 11:37–57

21. Quint LE, Glazer GM, Francis IR, Shapiro B, Chenevert TL. Pheochromocytoma and paraganglioma: comparison of MR imaging with CT and I-131 MIBG scintigraphy. Radiology 1987; 165:89–93

22. Shapiro B, Sisson JC, Shulkin BL, Gross MD, Zempel S. The current status of meta-iodobenzylguanidine and related agents for the diagnosis of neuroendocrine tumors. Q J Nucl Med 1995; 39 [suppl 1]:3–8

23. Farthouat P, Platal JP, Meusnier F, Pourrire M, Thouard H. Paragangliome rétropéritonéal sécrétant: à propos d'un cas. J Chir.1997; 134 (5-6): 248-251. PubMed | Google Scholar.

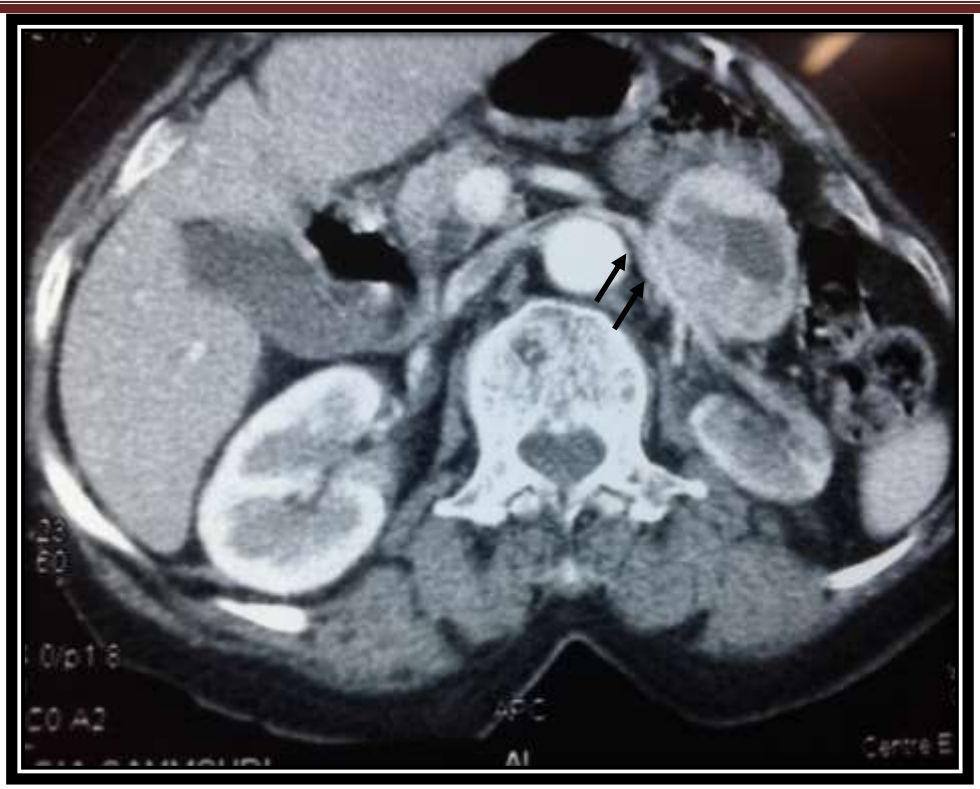

Fig.1a: Contrast enhanced tomography: heterogeneous lesion with low attenuation in central) areas (black arrow. We note the adherence to the renal vein.

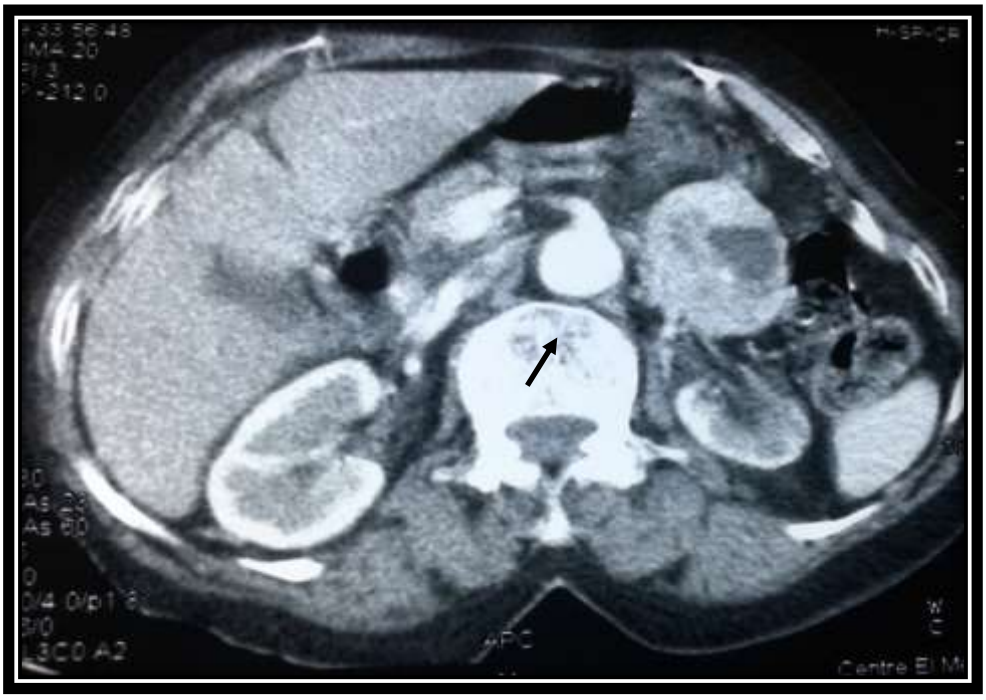

Fig.1 b: Contrast-enhanced tomography: the adrenal gland seemed intact from tumor.

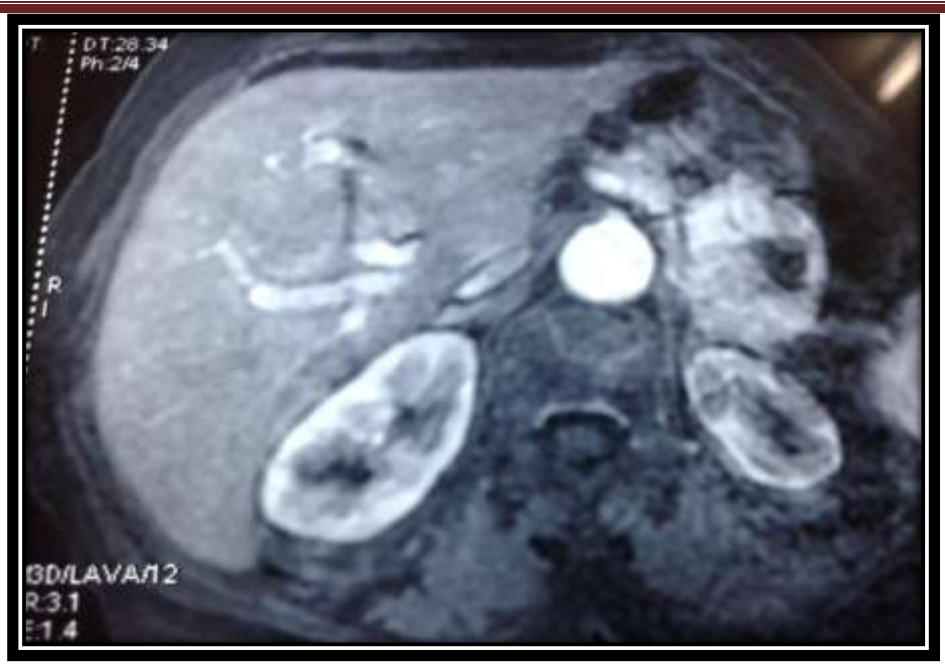

Fig2.T1 weighted MRI with gadolinium injection: well-defined hyperinse lesion with Low attenuation in central areas proximal to the renal vein.

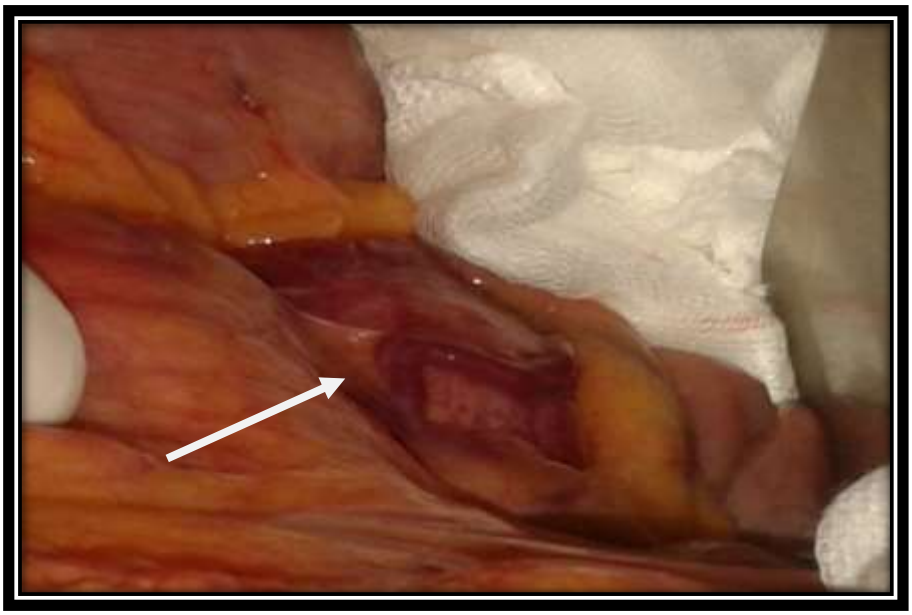

Fig. 3a: Operative view of the tumor deeply embedded in the left meso colon.

Fig3b*. Operative view. Ligation *of the drainage vein after resection of the tumor.

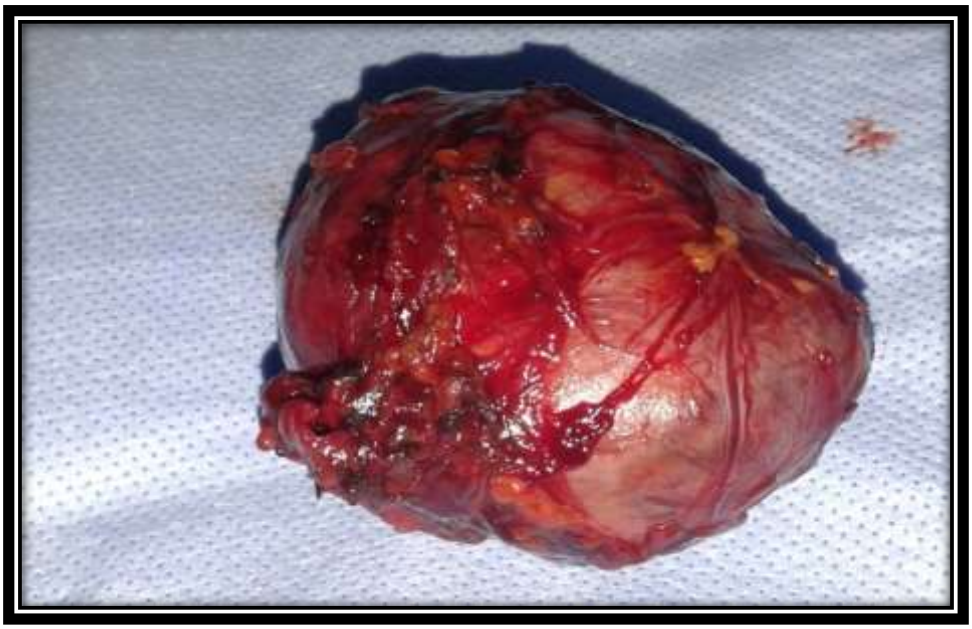

Fig.4a

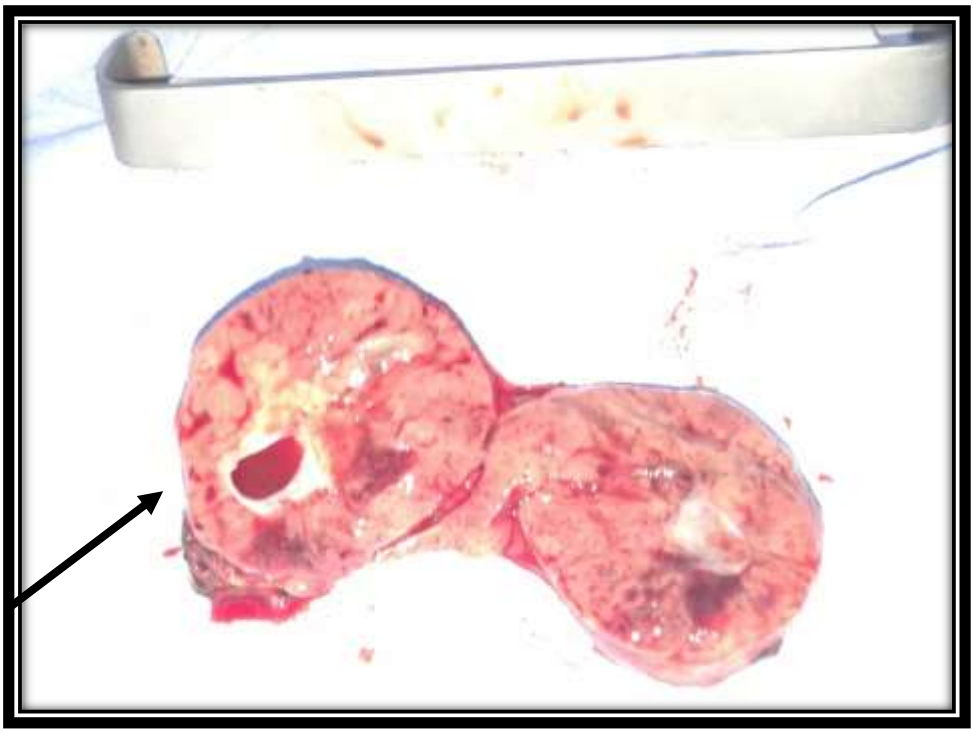

Fig. 4a+b. Macroscopic view of the operative specimen. After section; we note the mixed aspect with central cystic component.