# Case report

## Intraoperative diagnosis of a paraganglioma

*Kamal E Elsiddeg [1], Abdrahim A, Dabora [1] Waleed E Abdrahim [1], Abduraheem F Abduraheem [2], Mohammed elkhalifa [2]

[1] *Department of Surgery, Faculty of Medicine, University of Khartoum*

[2] *Department of Internal Medicine, Faculty of Medicine, University of Khartoum*

### Introduction:

Paragangliomas are extra-adrenal chromaffin tumors that arise from neuroectodermal cells of the autonomous nervous system [(1).] They are rare tumors, and may be located in the skull base, neck, chest and abdomen. Also they can be functioning secreting catecholamine and producing symptoms or non-functioning producing no symptoms (silent). When found within the abdomen, a silent paraganglioma may be mistaken for other retroperitoneal tumors, such as a lymphoma or a tumor of the pancreas.

### Case presentation:

A 19-year-old female originally presented in September 2015 with complaints of upper abdominal pain accompanied with pain radiating from the waist to the left lower limb, headaches and two attacks of what had been thought of as a seizure. She had been evaluated and diagnosed as epilepsy and started on treatment. But her abdominal pain and the headaches did not settle. A contrast-enhanced CT of the abdomen demonstrated a 3X4cm left adrenal mass with predominantly peripheral enhancement and central areas of low attenuation, presumed to represent necrosis fig 1.

The patient underwent surgical exploration. Unexpectedly, the patient became hypertensive with a systolic blood pressure reaching 200 mmHg and supraventricular tachycardia (SVT) during the initial manipulation of the tumor and were managed by phenoxybenzamine 40mg Iv and labetalol infusion 1mg/min by the anaethetist. Paraganglioma was suspected to be the cause of this circulatory problem. The blood pressure and SVT were rapidly controlled and the tumor was completely resected.

Pathological examination of the specimen revealed well-encapsulated tumor forming tubules , trabeculae and solid sheets . The tumor was composed of polygonal cells displaying vesicular nuclei and acidophilic cytoplasm. Pleomorphic and multinuclei, as well as foci of necroses, were noted and confirmed the diagnosis of paraganglioma(fig 2). The patient's postoperative course was uneventful; her blood pressure returned to normal and she was discharged 5 days after the operation.

***Corrosponding author :***Department of surgery, Faculty of Medicine, University of Khartoum,*
*e-mail:elzakikamal@gmail.com*

### Discussion

Extra-adrenal phaeochromocytomas (EAPs) are known as paragangliomas. The majority of these occur intra-abdominally (85% below the diaphragm), along the sympathetic chain or from the organ of Zuckerkandl [(2)].

Paraganglioma is probably the most fascinating of all the tumors as it can present with a wide range of clinical manifestations [(3)] .The clinical symptoms vary according to the amount of catecholamines released. Observable clinical effects are only present if the tumor secretes a sufficient quantity of catecholamines. However, circulating

catecholamine levels do not have a strong correlation with the degree of hypertension in paraganglioma. It is considered that 30% of functional paraganglioma patients have normal blood pressure [4]. In this case, the patient had normal blood pressure and did not receive a metanephrine examination, with a diagnosis of retroperitoneal tumor. However, the blood pressure rose intraoperatively upon touch and mobilization of the tumor. Once the tumor was removed, the patient's blood pressure returned to normal.

In symptomatic patients, initial diagnostic investigations include demonstration of catecholamines and/or its metabolites in plasma or urine. Imaging then subsequently plays an important role in localizing the adrenal and EAPs. CT scan is usually the initial modality of choice for detecting these tumours. If the mass is not localized on CT scan, and the disease is clinically or biochemically suspected, MRI is then indicated. Functional imaging can also be performed to confirm the presence of EAPs. The most commonly studied functional imaging is $^{131}$I–labeled metaiodobenzylguanidine ($^{131}$I-MIBG) which shows high sensitivity and specificity in detecting EAPs. It can also be used to assess multifocality, metastasis and recurrence of the tumor.

As far as the treatment of paragangliomas is concerned, the best choice is complete surgical resection since these tumors are potentially malignant. However, it is important to note that for those with functional paragangliomas, the tumor's ability to produce catecholamines may cause abrupt changes in the blood pressure, which may cause an abnormal cardiac rhythms and ,even, asystole. Thus, surgery may induce life-threatening complications as mentioned above. However, pre-medication of symptomatic patients with positive biological tests has been recommended.The treatment strategy remains unclear when the patient is asymptomatic and has low catecholamine levels. In this study, the patient did develop a hypertensive reaction during surgery although she was asymptomatic preoperatively.

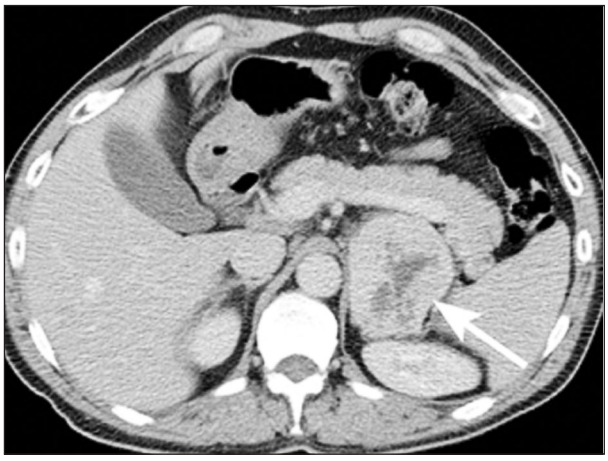

**Fig. 1:**19-year-old female with pheochromocytoma. Axial, contrast-enhanced CT demonstrated a 3X4cm left adrenal mass (arrow) with predominantly peripheral enhancement and central necrosis (arrow).

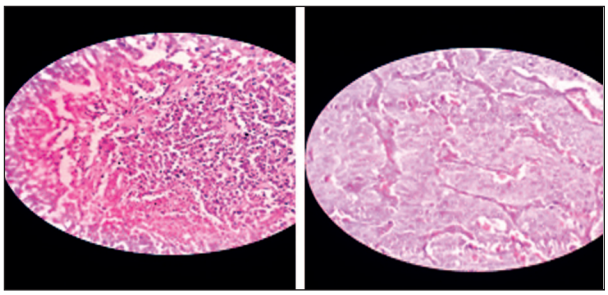

**fig.2:** show well-encapsulated tumor forming tubules , trabeculae and solid sheets . The tumor is composed of polygonal cells displaying vesicular nuclei and acidophilic cytoplasm. Pleomorphic and multinuclei as well as foci of necroses are noted .

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