limits. Computed tomography of the abdomen demonstrated a left retroperitoneal tumor of 52*56 mm, extrarenal, located in contact with the left renal vein, laterally of aortal axis, with dense collateral circulation, no lymphnodes, no metastases (figure2, 3).

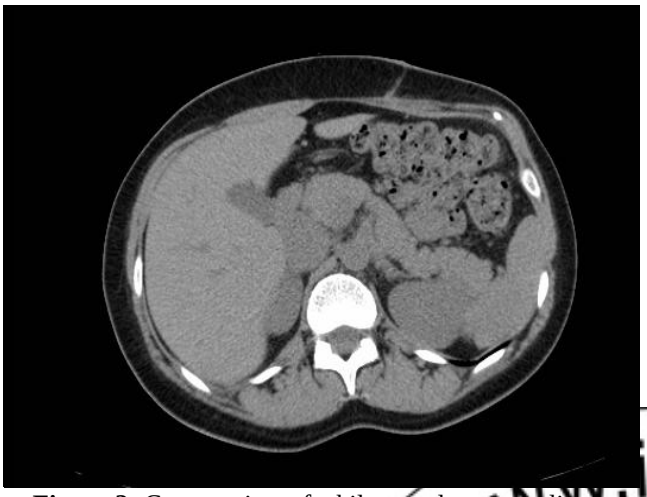

**Figure 2:** Ct-scan view of a hilar renal paraganglioma

Surgery was arranged 2 weeks after hypertension was medically treated by the endocrinologist. During surgery exploration, a 60*40 mm retroperitoneal tumor was found, encapsulated, which appeared soft and richly vascularised. It was localized between aorta and left kidney, laterally a inferiorly of cauda pancreatis, behind and inferiorly spleen. The left adrenal gland was intact.

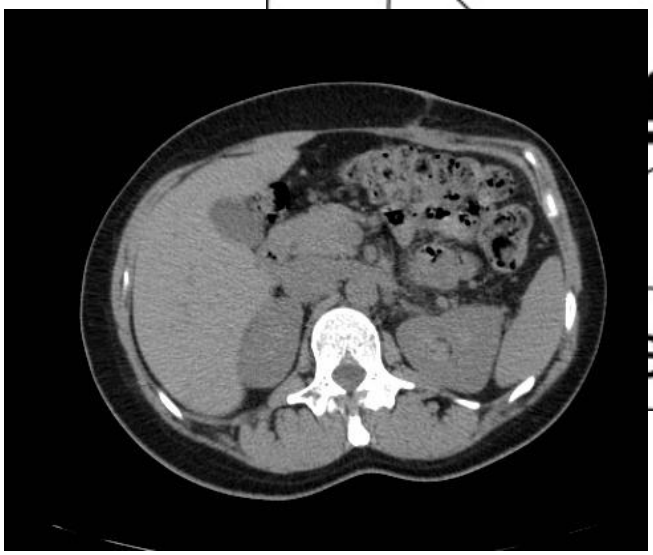

**Figure 3:** Ct-scan view of a hilar renal paraganglioma

Removal of the tumor was done and it was sent for histopathological examination (figure 4, 5).

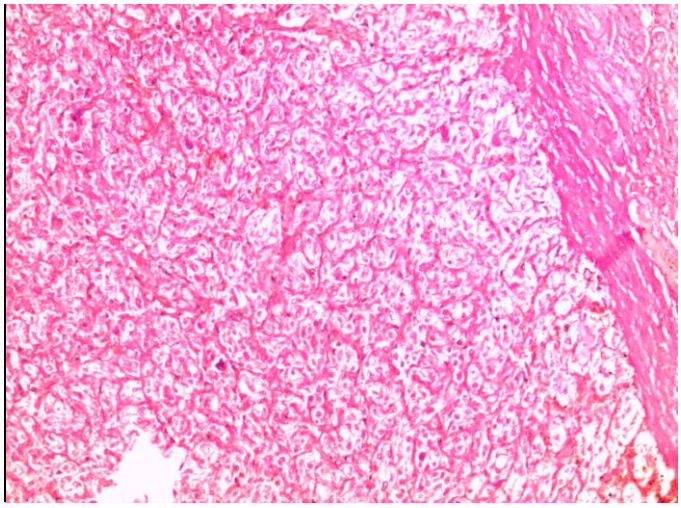

**Figure 4:** Histopathology view of the paraganglioma

The biopsy revealed a paraganglioma tumor. The patient had complete resolution, safely discharged from hospital. The 5 years follow up process revealed the patient is healthy.

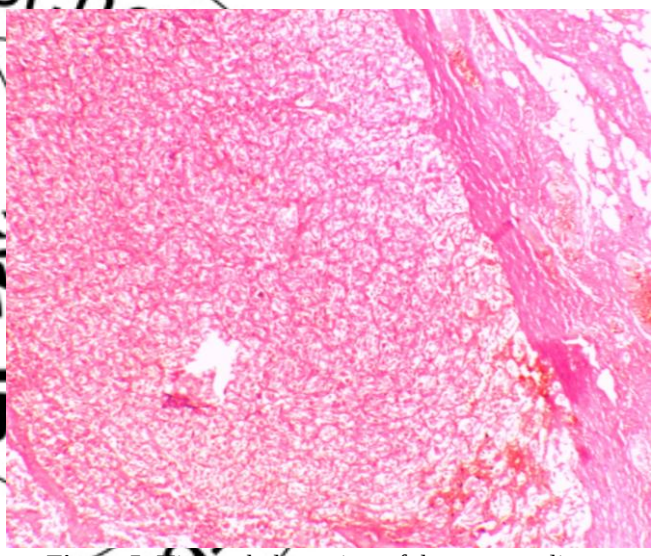

**Figure 5:** Histopathology view of the paraganglioma

## 3. Discussion

According to the World Health Organization classification of tumors, in 2004, paragangliomas are a type of neuroendocrine tumors derived from embryonic neural crest. They are mostly found in the adrenal glands (pheochromocytoma) and in the extra-adrenal paraganglia of the autonomic nervous system (paragangliomas)[1]-[4].They usually occur sporadically, but they can be a feature of two disorders with an autosomal dominant pattern of inheritance: multiple endocrine neoplasia type 2 and 3 (MEN 2 and MEN 3) and von Hippel-Lindau desease. They may be part of Carney's triad associated with gastric leiomyosarcoma and pulmonary chondroma[12]- [13]. These tumors can be associated with clinical evidence of epinephrine secretion which makes them functioning tumors in 50-60% of cases [2]-[3], [7].In fact the surgical manipulation of the neoplastic mass may produce a risky hypertensive crisis [2]-[3].There are no histological criteria to distinguish between functioning and non functioning tumors[11].But usually head and neck

paragangliomas, arising from parasympathetic paraganglions are nonsecretant. Histology and imunohistochemistry remain the gold standard for making the definitive diagnosis. Approximately 10%of them are malignant, nevertheless this cannot be determined on a biochemical or histological basis. It is the presence of local invasion, on gross or microscopic exam and metastasis presence that define malignancy [15]-[16]. Plasma free metanefrine and normetanefrine or urine metanefrine are the first tests to make the diagnosis of the desease[12].Imaging studies such as CT-Scan and IRM are essentials before surgery for localization, size and evidence of metastasis. According to the NCCN guidelines the treatment of a paraganglioma is total resection when possible. If not, cytoreductive resection must be followed by radiation or systemic chemotherapy, depending on the extent of the metastases [17]. Paragangliomas are more likely to be malignant then pheochromocytomas(40% *vs* 10%)[2]-[15].However evidence of metastasis is the only definitive criterion to label the tumor as malignant[2], [18]- [19].

## 4. Conclusion

In summary paragangliomas must be considered a possible cause of arterial hypertension in young people. Upon diagnosis, surgery is the only curative treatment. Adequate preoperative preparation and treatment, a multidsciplinary team (surgeon, endocrinologist, and anesthesiologist) can reduce morbidity and mortality.

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