

Letter to the Editor

Cause of unexpected abdominal pain in a young patient: extra-adrenal retroperitoneal retro-pancreatic giant paraganglioma

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Paraganglioma is a very rarely seen neuroectodermal tumor. Paraganglioma is defined as a tumor developing from neuroendocrine cells associated with the sympathetic or parasympathetic nervous system. A tumor rooted in the adrenal medulla is referred to as a pheochromocytoma [1, 2]. Approximately 10% of cases are rooted extra-adrenally, and are seen most frequently in the abdomen, thorax, and neck region [3]. Paraganglioma rooted in the retroperitoneum is generally functional. Others rarely show any function [4]. Surgical resection and radiotherapy (RT) are the preferred methods of treatment for these tumors. The case presently described is a rarely seen retroperitoneal paraganglioma.

A 26-year-old female patient presented to the polyclinic with abdominal pain ongoing for a month. The patient history did not reveal anything significant. Arterial blood pressure was 110/70 mm Hg and pulse was 88/bpm on physical examination. Mild sensitivity and fullness were present in the epigastric region on abdominal examination. While the hemoglobin level was 9.8 g/dl and the hematocrit test value was 31.7%, other laboratory findings were normal. The abdominal ultrasonography image revealed a solid retroperitoneal mass lesion, 8 × 6 cm in size, with well-defined margins. A hypervascular, well-margined, solid mass lesion, approximately 8.5 × 6 × 5 cm in size, was observed posteriorly, adjacent to the pancreas on abdominopelvic computed tomography (Figures 1 A, B). It was seen that the mass was in close contact with the superior mesenteric artery (SMA), celiac truncus, left renal artery, left renal vein, and splenic vein. Based on these findings, the decision was made to pursue surgical treatment. The mass, pressing on the SMA posterior to the pancreas in the retroperitoneal region, splenic vein, left renal artery and vein, was totally excised with laparotomy (Figure 2). There were no intraoperative complications. The patient was discharged uneventfully. Pathological examination revealed an encapsulated, lobular, solid mass lesion, 10 × 9.5 × 5 cm in size, with yellow-white color. Capsule invasion, vascular and perineural invasion, and atypical mitosis and necrosis were present. Nuclear pleomorphism was not detected. Tests for chromogranin and CD56 were positive, and synaptophysin was weakly positive. The Ki-67 index was high (16%). According to these findings, the case was diagnosed pathologically as paraganglioma. The patient's initial follow-up of the first 6 months after surgery did not reveal any features.

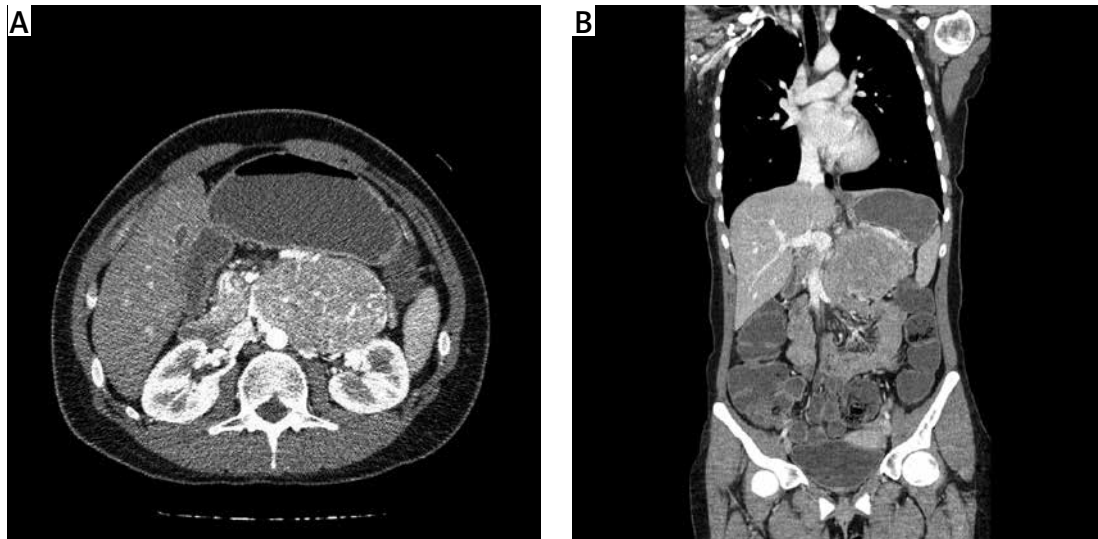


Figure 1 A, B. Axial and coronal abdominopelvic computed tomography

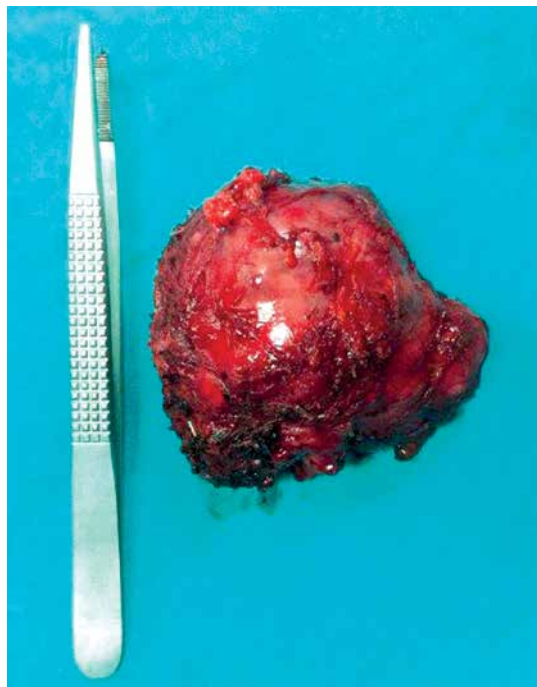


Figure 2. Surgery material

Extra-adrenal paraganglioma can be seen in the abdomen, retroperitoneum, thorax, mediastinum, and neck region. Retroperitoneal tumors manifest at an early age, are generally functional, are malignant at a rate of 40% to 50%, and show slow progression [4]. Extra-adrenal paragangliomas generally do not lead to clinical symptoms based on hormone secretion and catecholamine production, unlike those that are adrenally rooted. Non-functional paragangliomas are malignant. The only reliable criterion to confirm malignancy is presence of distal metastasis. Late-period metastases are seen frequent-

ly. Tumor extension occurs hematogenously or lymphatically. Metastases to the lymph nodes, bone, lungs, and liver may be seen. Pathologically, local tissue invasion or nuclear pleomorphism and mitosis are not signs of definite malignancy [5]. Some features related to malignant tumors include extra-adrenal localization, incomplete surgical resection, capsule invasion, vascular and perineural invasion, extensive necrosis, patient of young age, tumor size, and high Ki-67 index [6, 7]. Primary treatment of paraganglioma is surgical excision of the lesion [8, 9]. Despite complete resection, recurrence at a rate of 8% to 20% and distal metastasis at a rate of 28% to 50% can occur [10]. Surgical resection and RT are effective in local treatment of these tumors. In the present case, the tumor was excised as a retroperitoneal mass. Tumors located in the retroperitoneal region can reach an enormous size. Consequently, they may cause pressure symptoms. Therefore, even if total excision of the mass is not possible, a close-to-total excision should be performed.

In conclusion, paraganglioma should be kept in mind in the differential diagnosis of retroperitoneal masses. Relieving the patient of pressure and pain symptoms should be achieved by ensuring local control of the tumor. Moreover, close post-operative follow-up is important due to the risk of recurrence and metastasis.

References

1. Haber S. Retroperitoneal and mediastinal chemodectoma: report of a case and review of the literature. *AJR* 1964; 92: 1029-41.
2. Cunningham SC, Suh HS, Winter JM, et al. Retroperitoneal paraganglioma: single-institution experience and review of the literature. *J Gastrointest Surg* 2006; 10: 1156-63.

3. Somasundar P, Krouse R, Hostetter R, Vaughan R, Covey T. Paragangliomas: a decade of clinical experience. *J Surg Oncol* 2000; 74: 286-90.
4. Grant CS. Pheochromocytoma. In: *Textbook of Endocrine Surgery*. Clark OH (eds). 1st ed. W.B. Saunders, Philadelphia 1997: 513-28.
5. Chatterjee P, Singh A, Ayyappan A, Nair A. Extra-adrenal retroperitoneal paraganglioma associated with duplication of inferior vena cava. *Radiology Case* 2008; 2: 25-7.
6. Lack EE, Cubila AL, Woodruff JM, Farr HW. Paragangliomas of the head and neck region: a clinical study of 69 patients. *Cancer* 1977; 39: 397-409.
7. Linnoila RI, Keiser HR, Steinberg SM, Lack EE. Histopathology of benign versus malignant sympathoadrenal paragangliomas: clinicopathologic study of 120 cases including unusual histologic features. *Hum Pathol* 1990; 21: 1168-80.
8. Disick GI, Palese MA. Extra-adrenal pheochromocytoma: diagnosis and management. *Curr Urol Rep* 2007; 8: 83-8.
9. Gündeş E, Aksoy F, Taşcı Hİ, Yılmaz O. Retroperitoneal extra-adrenal paraganglioma: case report. *Med J Bakirkoy* 2016; 12: 51-3.
10. Nap RR, Meinardi JR, van den Berg G, et al. Long-term follow-up is indicated after surgery for a pheochromocytoma. *Ned Tijdschr Geneesk* 2006; 150: 1045-9.