

Case Report

Inter-aorta-caval Located Tumor: A Case Report

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A paraganglioma is a rare tumor that develops out of extra-adrenal chromaffin cells and pheochromocytomas originating from the adrenal medulla. Early diagnosis and surgical planning are crucial, since the tumor secretes catecholamine and is adjacent to large vessels in the abdomen. Furthermore, since complete resection improves the prognosis, we recommend a meticulous surgical technique. Here, we present a case of paraganglioma in a 32-year-old male patient who initially presented with a stomachache. After conducting the required tests, we resected the tumor that was pressing against the vena cava in the interaortocaval region.

Key words: pheochromocytoma, aortography, surgery

Introduction

A Pheochromocytoma is a rare tumor that stems from chromaffin cells of the sympathetic nervous system. The tumors secrete catecholamine and cause hypertensive episodes. In 90% of patients, the cells stem from the adrenal medulla, and in 10%, from the extra-adrenal medulla, and are called paraganglioma.¹⁾ Hypertension, headache, sweating, weight loss and palpitation are common symptoms of these aggressive and malignant tumors. Furthermore, since they are adjacent to major vessels such as aorta and vena cava inferior, they must be treated aggressively. It is possible to diagnose these tumors with magnetic resonance imaging (MRI), scintigraphy and computerized tomography (CT).^{2,3)}

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Case Report

A 32-year-old male patient who had been diagnosed with a tumor in his abdomen was subsequently referred to the Department of Cardiovascular Surgery, Ondokuz Mayıs University Medical School, Turkey. The general examination of the patient indicated no serious health problems. His blood pressure was 130/75 mm Hg with a pulse of 86 beats/min. We detected sensitivity on his right quadrant with palpation. Other systemic examinations produced normal results. Upper abdominal CT examination showed a tumor on the anterior part of the anteraortocaval region. The tumor occupied the area from the inferior part of the liver to the iliac bifurcation and pressed against the inferior vena cava. The tumor also contained a necrotic part in its center, with dimensions of 9 × 6 × 8 cm (**Figs. 1** and **2**). We suspected paraganglioma due to the location of the tumor and the level of contrast material, revealed by angiography (**Fig. 2**). Metaiodobenzylguanidine (I-131 MIBG) concentrated in the right part of the mid line of the inferior liver in all body scanning images, and the catecholamine level in 24-h urine examinations was high, which led to the diagnosis of paraganglioma. The serum counting blood cells (CBC) and biochemical examinations were normal.

The patient exhibited no hypertensive episode or fever during the clinical observations. Considering the diagnosis of paraganglioma, the cardiovascular surgical team

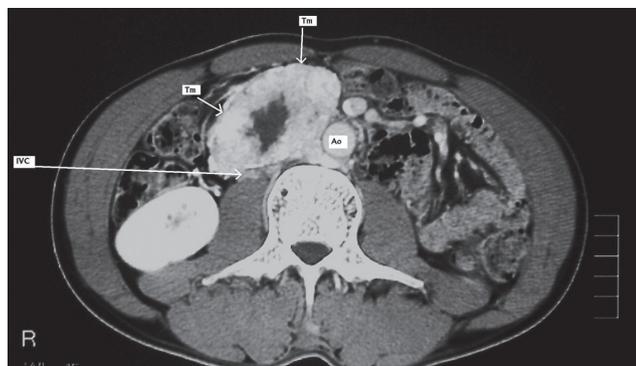


Fig. 1 Abdominal-CT scan of tumor mass.
Ao, aorta; Tm, tumor; IVC, inferior vena cava

with specialists from the General Surgery Clinic resected the tumor. During the operation, we could see a tumor of 7×7 cm that was adherent to the aorta and vena cava. The tumor was retroperitoneally located on the inferior part of the duodenum. We carefully excised the tumor from the edges of the aorta and vena cava, so as not to damage the adjacent vessels. The patient had a complication-free post-operative period and was discharged on the sixth day following the operation. The pathological examination of the surgical specimen indicated pheochromocytoma and confirmed paraganglioma, as well.

Discussion

Pheochromocytoma stemming from extra-adrenal chromaffin cells is called “extra-adrenal pheochromocytoma” or “paraganglioma.”⁴⁾ The paraganglioma is located in the extra-adrenal region and secretes catecholamine; it stems from chromaffin cells of the sympathetic nerve system,⁵⁾ proliferating on the carotid mass, jugular foramen, mediasten, Zuckerkandl organ (located around the aortic bifurcation) and the peri-aortic area. However, paraganglioma also proliferates in areas near sympathetic ganglions.⁶⁾ Eighty-five percent of tumors are located beneath the thoracic diaphragm; they do not usually invade between the abdominal aorta and inferior vena cava.²⁻⁸⁾

In the case described here, the tumor was located on the inferior part of the liver and anterior part of the interaortacaval region. We carefully dissected the tumor from the vessel walls and the surrounding tissues. In comparison with adrenal pheochromocytoma, the metastasis ratio for paraganglioma is higher at the time of diagnosis,⁸⁾ particularly in men in the 4th and 5th decades

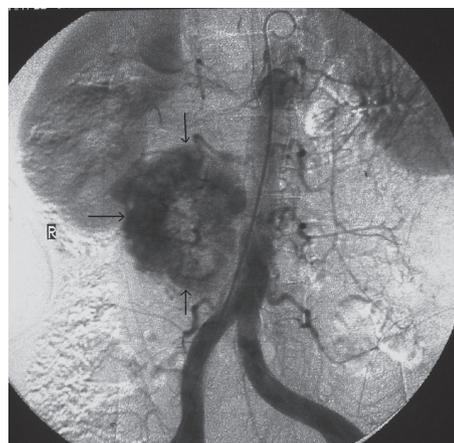


Fig. 2 Abdominal DSA angiogram of hypervascular mass, located to the right of the aorta (black arrows).

of their lives; however, the tumor can be detected in people of other ages as well.^{6, 9)} Most of the retroperitoneal paragangliomas are functional, leading to symptoms that are similar to those of adrenal pheochromocytoma, such as paroxysmal hypertension, sweating, headache, palpitation springing from the increase in serum catecholamine.¹⁰⁾ Paraganglioma is generally observed sporadically; however, approximately 10% of the tumors are associated with genetic diseases such as neurofibromatosis type 1, von Hippel-Lindau disease, carney triad syndrome and multiple endocrine neoplasia (MEN) type 2.¹⁻⁴⁾ The serum and 24-hour urine catecholamine levels are often high in such cases; however, our patient had normal catecholamine levels and no history of a genetic disease.

Several methods are used in detecting the localization of the tumor. Although CT and MRI have high sensitivities, their specificities are low. For I-131 MIBG, its sensitivity is 77% for paraganglioma, and its specificity is 88%–100% for adrenal pheochromocytoma. Furthermore, MIBG is valuable because it can reveal areas of tumor metastases; thus, it is an indispensable tool in the arsenal of post-operative follow-up.⁷⁻¹⁰⁾

Conclusion

A Paraganglioma, a rare tumor, can pose a serious risk in the patient since it usually presses against major vessels in the abdomen. Thus, early diagnosis and immediate surgical intervention are essential. Surgery is especially preferred for large tumors, which require long-term follow-up since they are malignant and can metastasize.

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