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
with specialists from the General Surgery Clinic resected the tumor. During the operation, we could see a tumor of $7 \times 7$ 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.” The paraganglioma is located in the extra-adrenal region and secretes catecholamine; it stems from chromaffin cells of the sympathetic nerve system, 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. Eighty-five percent of tumors are located beneath the thoracic diaphragm; they do not usually invade between the abdominal aorta and inferior vena cava.

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, particularly in men in the 4th and 5th decades of their lives; however, the tumor can be detected in people of other ages as well. 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. 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.

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.

**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.
References