

Preoperative embolization in the management of a non-functioning mediastinal paraganglioma

Çalışmayan mediastinal paragangliyomun tedavisinde ameliyat öncesi embolizasyon

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Paragangliomas are rare tumors of the mediastinum. Complete surgical resection is the only effective treatment modality, although it is challenging due to the location of the tumor around great vessels and hypervascularity characteristics of tumor. In this article, we report a 37-year-old female case with paraganglioma in the middle mediastinum detected incidentally by radiological imaging. The tumor was inactive; however, severe bleeding occurred in the initial diagnostic thoracotomy. The tumor was resected successfully after selective embolization. Following complete resection, the patient followed for five years with no recurrence.

Key words: Mediastinum; paraganglioma; therapeutic embolization.

Paragangliomas are tumors which develop from the chromaffin cells of the sympathetic nervous system anywhere from the pelvis to the base of the skull.^[1] Paragangliomas of the mediastinum are rare and originate from the aortosympathetic paraganglia.^[2] About 2% of all paragangliomas are located in the chest,^[3,4] and most are functional and diagnosed during the investigation of hypertension secondary to catecholamine excess.^[2,3,5] Nonfunctional mediastinal paragangliomas are diagnosed incidentally by radiological imaging performed due to symptoms related to the compression or invasion of the local structures in the mediastinum.^[3,4,6] Surgery is the

Paragangliyomlar mediasteninin nadir tümörleridir. Tümörün büyük damarlar çevresinde olması ve hiper-vasküler özellikleri nedeni ile zor olmasına rağmen, tam cerrahi rezeksiyon tek etkin tedavi yöntemidir. Bu yazıda, radyolojik görüntüleme ile tesadüfen saptanan orta mediastende paragangliyomu olan 37 yaşında bir kadın olgu sunuldu. Tümör aktif değil idi; ancak ilk tanışal torakotomide ciddi kanama meydana geldi. Tümör selektif embolizasyondan sonra başarılı bir şekilde rezeksiyon edildi. Tam rezeksiyondan sonra hasta nüksüz olarak beş yıl takip edildi.

Anahtar sözcükler: Mediasten; paragangliyom; terapötik embolizasyon.

mainstay therapy for mediastinal paragangliomas since these tumors are resistant to radiotherapy or chemotherapy.^[4] They are also hypervascular in nature. Many of these tumors invade adjacent organs such as the great vessels, heart, trachea or spine; therefore, complete resection can be difficult.^[2-7] Since mediastinal paragangliomas are rarely seen, treatment protocols have not yet been established.^[2,6] Herein, we report the case of patient with a non-functional paraganglioma located between the pulmonary artery and superior pulmonary vein extending to the mediastinum with splaying of the superior pulmonary vein branches. The tumor caused massive bleeding during an



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initial diagnostic thoracotomy, and it was embolized preoperatively using selective catheterization to reduce vascularity. This was followed by complete excision via a thoracotomy as a redo operation.

CASE REPORT

A 37-year-old woman presented with dyspnea, a cough, and chest pain that had been ongoing for two months, but she had no other symptoms such as palpitation, sweating, or headaches. Her past medical history was insignificant. A physical examination and routine biochemical tests were normal. However, a chest X-ray revealed a homogenous opacity on the right hilar area, and chest computed tomography (CT) revealed a mass measuring 51x41x35 cm in diameter that extended from the right pulmonary artery to the mediastinum. The tumor was compressing the right middle lobe bronchi via the splaying of the superior pulmonary vein branches (Figure 1). The patient underwent a non-diagnostic fiberoptic bronchoscopy that detected only the compression of the middle lobe bronchus, and a non-diagnostic transbronchial needle aspiration (TBNA) biopsy was also performed. The patient then underwent a CT-guided transthoracic fine needle aspiration biopsy (FNAB) to make a histopathological diagnosis, and it showed monomorphic epithelial cells but was also non-diagnostic. Because of the nondiagnostic transthoracic FNAB, a right posterolateral thoracotomy was performed through the fifth intercostal space along with a partial excision of the hypervascular tumor that extended to the mediastinum. There was excessive



Figure 1. A single slice of contrast-enhanced computed tomography at the level of the mass.

bleeding during the hilar dissection and wedge biopsy of the tumor, and the perioperative blood loss was estimated at 2.5 liters. When examining the frozen section, the biopsy specimen was characteristic of a paraganglioma. Due to the hypervascularity of the tumor and massive bleeding, complete excision could not be carried out. During surgery, no remarkable blood pressure changes occurred. In addition, the fractionated catecholamines and metanephrines [urinary levels of 5-hydroxyindoleacetic acid (5-HIAA), metanephrine, noradrenalin, and adrenalin] were measured, and all of the results were normal. One week after the diagnostic surgery, due to the hypervascular nature of the tumor, a consultation was held with the interventional radiology department to assess the possibility of embolization before definitive surgical resection was performed. The patient underwent CT angiography of the thorax and selective angiography of the bronchial arteries, which were originating from the ascending aorta and supplying the tumor. These were detected and embolized with polyvinyl alcohol (PVA) particles, hence, the blood supply via these arteries was thereby occluded. The embolization of all of these branches resulted in an almost complete disappearance of the tumor blush (Figures 2a and b), and the patient was asymptomatic during the procedure. She returned to the operating room on her sixth post-embolization day for a rethoracotomy and excision of the paraganglioma. Furthermore, intraoperative dissection of the mass from the superior pulmonary vein, pulmonary artery with its middle lobe branches, middle lobe bronchus, and pericardium was carried out with the utmost care. A complete resection was possible with negative margins. Hemostasis was not problematic, and the patients' blood loss was estimated at 500 mL. The postoperative course was uneventful, and she was discharged on postoperative day five.

A histological examination of the specimen showed that the tumor cells were arranged in small nests, and mitotic activity was detected. However, no vascular or capsule invasions were found. In an immunohistochemical analysis, the stained tumor cells were positive for S-100, synaptophysin, and chromogranin A. After a 60-month follow-up, the patient is doing well with no evidence of disease.

DISCUSSION

Paragangliomas are rare neoplasms of the extra-adrenal chromaffin tissue that are associated with the autonomic nervous system.^[2] These tumors are mainly asymptomatic and manifest in patients over 40 years of age at equal ratios for both genders.^[2]



Figure 2. (a) Angiogram showing a right bronchial artery and the hypervascular nature of the tumor before embolization. (b) Angiogram after the embolization of the right bronchial artery that was supplying the tumor.

The anterosuperior compartment is the most commonly involved site followed by the middle and posterior compartments.^[5] In our case, the patient presented with a middle mediastinal mass which was growing and pressing into the right hilar structures.

Functional mediastinal paragangliomas are often diagnosed during the surveillance of hypertension with symptoms related to catecholamine secretion (i.e., palpitation, flushing, sweating, and tremors).^[2-4,6] Non-functioning mediastinal tumors are often asymptomatic and are usually found incidentally.^[2,3] A chest radiography and CT are still the most frequently tools used for initial diagnostic examinations,^[3] with symptoms such as a cough, chest pain, and dyspnea due to the compression of the mediastinal organs commonly seen. The diagnosis of a non-functioning paraganglioma can be confirmed histologically.^[3,5] In our case, the patient had symptoms related to the compression of the hilar structures, but there were no symptoms associated with catecholamine discharge. The tumor was detected via CT that was performed because of the presentation of a progressive cough and dyspnea.

There is no standard histological criteria for determining malignancy. The presence of distant metastasis is currently the only absolute criterion.^[2] In our case, there was no capsule invasion or mitotic activity

to indicate malignancy. Immunohistochemically, the stained tumor cells were positive for S-100, synaptophysin, and chromogranin A, which are specific for paraganglioma.^[5,6]

Mediastinal paragangliomas are aggressive tumors with a distant metastasis rate ranging from 19-26.6%.^[2] Because of their resistance to both chemotherapy and radiotherapy, complete surgical resection may be curative, but the tumors are often located around the great vessels. They then invade the mediastinum, which precludes this option.^[2-4] Andrade et al.^[6] reviewed the literature to evaluate the prognosis of this type of tumor and found a survival rate of 84.6% in 39 complete resected patients versus 50.0% for the 40 patients who underwent a biopsy or a partial excision and adjuvant treatment.^[2] Following the complete excision of the paraganglioma, our patient is currently doing well with no evidence of disease after 60 months of follow-up.

Due to the hypervascularity of the paraganglioma, complete resection is difficult. If the diagnosis is confirmed preoperatively, angiography should be conducted to evaluate vascular supply, and embolization of the blood supply is recommended.^[1,5,6,7] In fact, there are examples of embolization being performed between the first and seventh preoperative days without serious

complications.^[5,7] Furthermore, in the literature, massive bleeding has been reported during surgery or a biopsy.^[5] Rather than video-assisted thoracic surgery (VATS), a thoracotomy is preferred to reduce the risk of bleeding. If excessive bleeding occurs during the operation, the tumor excision should be postponed.^[5] In our case, complete excision was not possible because of the hypervascularity of the tumor during the first operation; therefore, it was done after the embolization of the blood supplies of the paraganglioma one week later. This was compatible with other reports in the literature.^[5,7]

Conclusion

Non-functional paragangliomas should be considered in the differential diagnosis if the surgeon encounters an unexpected hypervascular tumor of the mediastinum. Thus, if excessive bleeding is anticipated or if it occurs during the surgery, a two-stage operation should always be an option, with the second one being performed between one and seven days after the embolization.

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