CASE REPORT

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Atypically Located Paraganglioma Excisable by Partial Sternotomy

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ABSTRACT Paragangliomas are rare neoplasms that originate from chemoreceptors and can be found in different locations. Their growth rate is slow, and they are usually benign tumors. The surgical approach may vary according to the location of the tumor. In this study, we report the case of a 56-year-old patient who underwent paraganglioma excision. The tumor was in close proximity to the aortic arch and main vascular origins and also extended into the retrosternal space. Despite the incision matching the carotid bifurcation tracing, the tumor could not be completely reached. Therefore, the tumor could be excised by performing a partial sternotomy. We wanted to share the case and surgery due to the rarity of a paraganglioma of this size, its atypical location, and the fact that it requires an extra anatomical incision.

Keywords: Carotid body tumor; paraganglioma; neuroendocrine tumors

Carotid body tumors are rare neoplasms that originate from chemoreceptors and can be found in different locations. They can be named according to their location (glomus tumor, carotid paraganglioma, jugular paraganglioma, vagal paraganglioma).

Paragangliomas account for approximately 0.03% of all malignancies and are more common in 3-6 decades and in women.[1,2] Approximately half of all paragangliomas are carotid body tumors.[3]

They are slow-growing and usually benign tumors. The choice of treatment depends on the patient's age, symptoms, tumor size, growth rate, and general condition (surgical, medical, radiotherapy). Surgical excision is considered the standard approach for the treating carotid body tumors.[4]

Shamblin et al. classified carotid body tumors into three groups according to the degree of involvement of the internal carotid artery.[5] The most advanced stage according to the rate of carotid artery invasion is Type 3, and the tumor at this stage invades arterial structures. Complications during surgery are frequently observed in Shamblin Type 2 and Type 3. Their advanced vascular network and proximity to important neurovascular structures may lead to undesirable situations during surgery.

In this study, we report a 56-year-old patient who underwent paraganglioma excision. The tumor was in close proximity to the aortic arch and main vascular origins and also extended into the retrosternal space. Despite the incision matching the carotid bifurcation tracing, the tumor could not be completely reached. Therefore, the tumor could be excised by performing a partial sternotomy. We wanted to share the case and surgery due to the rarity of a paraganglioma of this size, its atypical location, and the fact that it requires an extra anatomical incision.

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56-year-old female patient. She had a history of congenital hemangioma in the bilateral upper extremities, hypertension, 10 pack years of cigarette smoking and cholecystectomy. She had complained of dizziness, pain, and weakness for two months. Her family history was unremarkable. On physical examination; general condition was good, oriented, coherent, bilateral upper and lower extremity pulses were palpable. There was a painless mass approximately 1×2 cm in size with unclear borders on palpation in the distal 1/3 of the left sternocleidomastoid muscle. While investigating the etiology of anemia in the internal medicine clinic, a mass in the upper mediastinum was detected on non-contrast thorax computed tomography (CT). The CT report showed a soft tissue density of approximately 45×38 mm in the left paratracheal area, extending to the neighborhood of the aorta, with a density similar to that of the thyroid, whose relationship with the thyroid could not be clearly evaluated (Figure 1). Contrast-enhanced magnetic resonance angiography of the neck was performed for further examination; there was a mass lesion in the left infrahyoid carotid speys in the clavicular region and retrosternal area, which was encapsulated in the left infrahyoid carotid speys, with a thin hypointense rim around it, containing sagittal visual areas and intensely stained close to homogeneous. The lesion is approximately 4.5×4×3.5 cm in size and is in close proximity to the aortic arch and main vascular origins. It pushed the left common carotid artery medially and the subclavian artery laterally. It is also closely adjacent to the innominant vein. In the center of the lesion, vascular structures with a huge caliber are remarkable (Figure 2).

A preoperative embolization procedure was performed. In left subclavian artery injection; tumoral feeding was observed from the left thyrocervical branch with dense vascular supply. The tumor feeder was entered with a microcatheter and embolized (Bead Block is a preformed, biocompatible polyvinyl alcohol hydrogel. It is produced by Boston Scientific, USA).

The patient underwent surgery 48 h after embolization. During the operation; under general anesthesia, an appropriate incision was made in the left carotid trunk. The carotid artery was explored. Sufficient surgical space could not be provided for tumor excision. Partial sternotomy was then performed. Arcus aorta, left subclavian artery, and left carotid artery were explored and the surrounding tissues were freed. The tumor was excised subadventitial with its capsule. The patient was discharged without complications on the 4th postoperative day.

In the pathology report; macroscopic findings were as follows; 5×3×3 cm mass, tumor cells immunohistochemical chromogranin (+), s-100 in sustantacular cells (+), vimentin (+), sma (-),

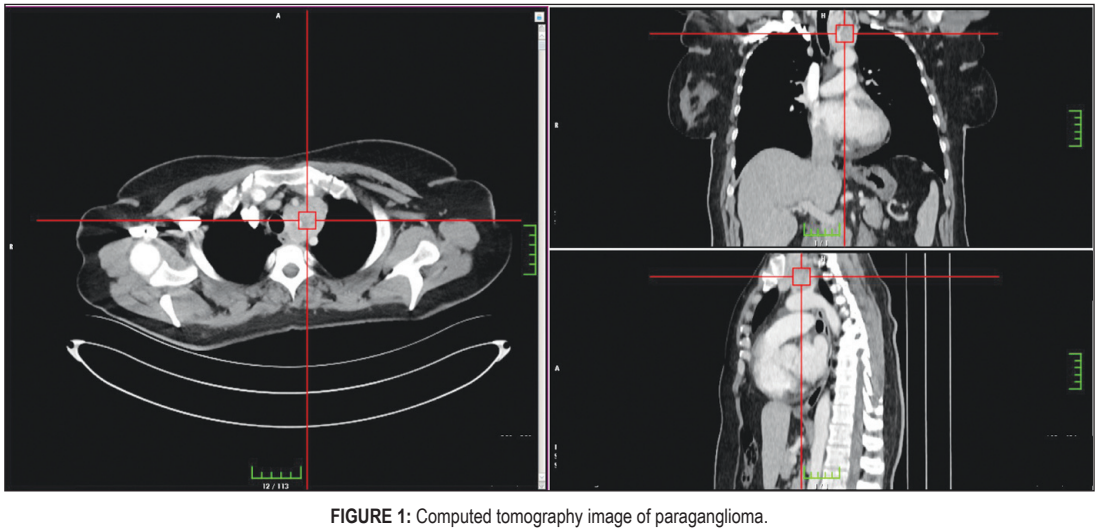

FIGURE 1: Computed tomography image of paraganglioma.

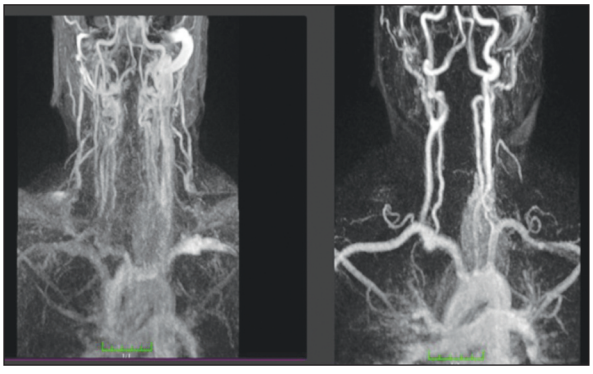

FIGURE 2: Contrast-enhanced neck magnetic resonance angiography image of paraganglioma.

pancytokeratin (-), ki67 index less than 1% (Figure 3).

Angiography of the aortic arch, and its branches performed at the first month after discharge showed normal diameters and free flow in the ascending aorta, aortic arch and its branches. Truncus brachiocephalicus, left common carotid artery, left subclavian artery and aortic arch outlets were patent.

Consent was obtained from the patient and their relatives for the article publication.

DISCUSSION

Intrathoracic paragangliomas may be confused with ectopic goiter during diagnosis. Ectopic goiter is the first diagnosis that comes to mind due to its frequency, and angiography may be required for differentiation.

Treatment of carotid body tumors includes medical follow-up, radiotherapy and surgical excision. The patient's age, tumor size and Shamblin classification should be considered when planning treatment (In this case, the use of the classical Shamblin classification is inadequate due to the extra anatomical extension of the tumor). Treatments other than surgery should be considered in patients with unfavorable medical conditions, recurrences, and large tumors where resection would cause severe morbidity due to extensive neurovascular involvement.[4,6] Even if there is a mediastinal location, the classical treatment recommended is surgical excision.[7]

Familial predisposition has been reported in 7-9% of cases and malignant transformation in 3-12.5%.[8] In this case, the pathology result of the excised mass was benign and the patient had a nega-

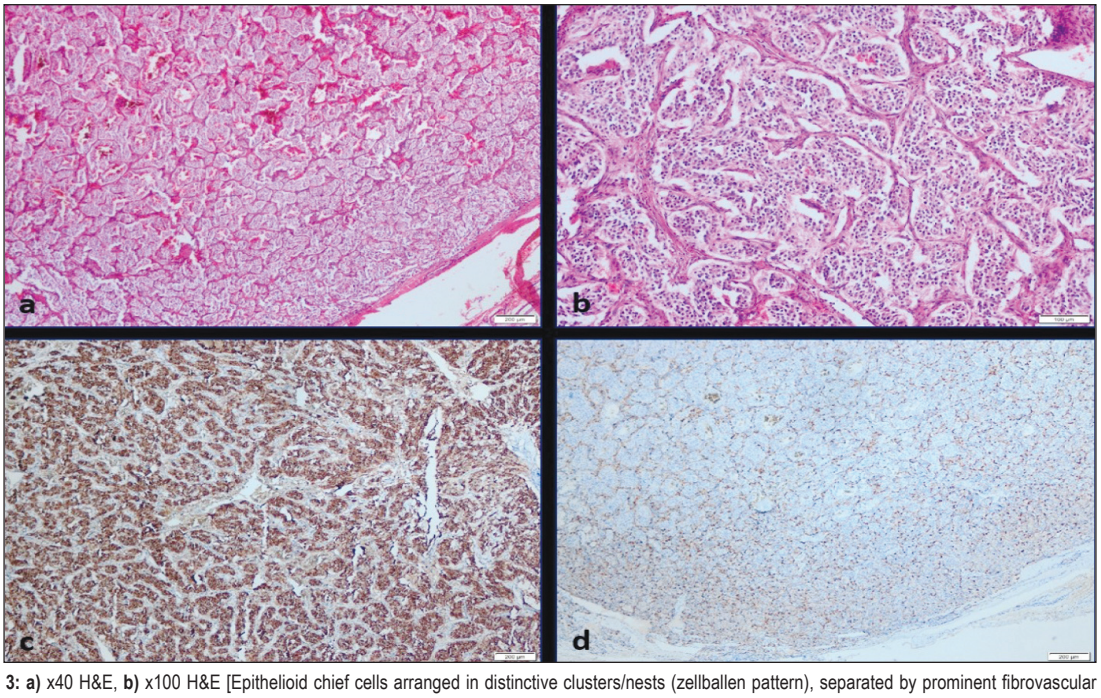

FIGURE 3: a) x40 H&E, **b)** x100 H&E [Epithelioid chief cells arranged in distinctive clusters/nests (zellballen pattern), separated by prominent fibrovascular stroma], **c)** Immunohistochemical chromogranin positivity x100, **d)** Immunohistochemical S-100 positivityin sustentacular cells x100.

tive family history. He had no symptoms other than dizziness, pain, and weakness for two months. Additionally, it was investigated for pheochromatoma and Von Hippel-Lindau disease, which may be associated with paragangliomas, and negative results were obtained.

Paragangliomas can sometimes cause serious difficulties during surgical resection because of their dense capillary network. Unparticular, complications related to the invaded carotid arteries and adjacent nerves can be observed. The incidence of neurologic complications is reported to be 14%-20% and is commonly related to cranial nerve damage.[9]

Preoperative embolization contributes to favorable outcomes in cases where complications may occur.[9] However, there are also opinions to the contrary.[10,11] In the preoperative evaluation of this case, the tumor was very large and invaded the carotid arteries. Therefore, embolization was performed 48 hours before surgery.

The general surgical technique for carotid body tumors is a transcervical approach (over the carotid bifurcation). It involves excluding the sternocleidomastoid muscle, ensuring control of the neurovascular structures, and then complete resection of the tumor subadventitially. There is no standardized surgical technique because of the rarity of these cases. In our case, we first attempted to reach the tumor with a classic incision. However, due to the large size of the mass and its extension to the left subclavian artery, the tumor and adjacent vascular structures could not be completely reached. Partial sternotomy was performed to control the aorta and its branches invaded by the tumor. Thanks to the new incision, surgical field control was achieved and the mass was easily excised.

As a result, thoracic paragangliomas can be confused with ectopic goiter. Before surgical excision of paragangliomas, the location and boundaries of the tumor should be well determined. Because the tumor is rich in collateral networks, serious bleeding may occur. The incision location is important as control over the surgical field may be lost in case of bleeding. It should be kept in mind that in cases with atypical locations, the incision site may change or an extra incision may be required.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

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