Case study

Paraganglioma of Carotid Body: A Case Report

Abstract

Paraganglioma occur from the paraganglionic stems of the autonomic nervous system. They develop from the carotid body and are known as carotid body tumours. These are sporadic, rare vascular lesions showing genetic transfer. Although they are generally benign and have a slow course, because of invasion to adjacent neurovascular tissues or pressure, early diagnosis and treatment is of importance. Diagnosis is made from a detailed history and physical examination and is confirmed with angiography. The treatment method selected in the majority of cases is surgery. In this paper, the case is presented of a 73-year old female who underwent surgery in our clinic for a diagnosis of carotid body paraganglioma.

Key words: Carotid body, paraganglioma, surgery.

İntroduction

Carotid body paraganglioma is a rarely seen tumour originating from neuro-ectodermal tissue, which is located in the carotid bifurcation (1). It is generally a slow-growing benign tumour (2). As the size of the mass increases, so the potential for malignancy increases, pressure is applied on the adjacent neurovascular structures, the surgical technique is made more difficult and the risk of complications inceases(3-6). Therefore, diagnosis must be made and surgical treatment planned in the early stage (1).

Case Report

A 73-year old female presented at the polyclinic with the complaint of swelling on the right side of the neck, which had been ongoing for 1 month. In the physical examination, an immobile mass was determined, 3.5 x3 cm in size with regular borders, located on the right side of the neck immediately behind the mandibular angle and extending as far as the earlobe. There were no findings of cranial nerve involvement. On the ultrasonographic (US) examination, a heterogenous, densely vascularised, solid mass was observed with dimensions of 35 x 30 x 20mm, which had regular lobular contours and was displacing vascular

structures in the right carotid bifurcation. On contrast computed tomography (CT) of the neck, a mass was observed approximately 32mm in diameter, adjacent and anterior to the right jugular vein. On the bilateral selective carotid angiographic examination, a mass was seen in the carotid bifurcation on the right side of approximately 3.5cm diameter. The mass was seen to be nourished from the parapharyngeal collateral vascular structures with approximately 30% of the superior-lateral part originating from the posterior auricular-occipital artery (Figures 1a, b).

The patient was admitted for surgery under general anaesthesia. The neck region was explored with a double parallel skin incision into the sternocleidomastoid muscle in the right side of the neck. Cranial nerves were carefully exposed. The tumour, 3.5 x 3cm in size, located between the carotid interna and externa was completely excised protecting the arteries and nerves.

The pathological examinatioon reported the mass as paraganglioma (Figure 2a, b). No neurological complication was encountered in the postoperative follow-up. Throughout the 27-month follow-up period, the patient was problem-free and no recurrence was observed.

Discussion

Carotid body paraganglioma originate from the paraganglion cells in the carotid bifurcation and is the most commonly seen form of paraganglioma of the head and neck (1). They are generally sporadic (7). There is no difference between the genders and peak incidence is seen at 40-50 years of age (1). The majority are benign in character and non-functional. Growth is generally slow and until a certain size is reached they are asymptomatic (2). As seen in the case presented here, 75% of patients present with a slow-growing painless mass on the neck. When the mass continues to grow, pressure on adjacent neurovascular structures results in symptoms being seen such as difficulty in swallowing, restricted hearing and pain in the ears. In those that are functional, symptoms may emerge asociated with catecholamine expression (8).

In the preoperative differential diagnosis, causes of the mass to be considered should include bronchial cysts, saliva gland tumours, carotid artery aneurism, lateral aberrant thyroid gland, malignant lymphoma, neurofibroma, tuberculous lymphadenitis and metastatic carcinoma (2). US, CT, MRI and angiograph are useful in the diagnosis (1). Angiography is extremely important in respect of understanding the specific arterial anatomy and also providing the means of vascular control intraoperatively (7). In the case presented here, definitive diagnosis was made with angiography.

Although the mass is generally benign, malignancy may develop in 3%-12.5% of cases which have been diagnosed late (9).

Carotid body tumours were classified by Shamblin et al (9) into 3 types according to size:
Localised mass
Surrounding the carotid artery
Completely wrapped around and adhering to the carotid artery.

The ideal choice of treatment is surgery. Removal of the tumour with careful subadventitial dissection should be selected in Shamblin types 1 and 2. The tumour in the current case was a Shamblin type 1 and the mass was excised with good surgical borders. If the mass is not completely excised, recurrence develops at a rate of 10% (1). In tumour resection, mean rates of mortality are seen at 2%, perioperative stroke at 2-3% and cranial nerve dysfunction at 40% (10).

In conclusion, these tumours which are not often seen, must be diagnosed in the early stage because of the complex relationships with adjacent structures and they must be treated surgically(11-12). Otherwise, the potential for malignancy and pressure symptoms may lead to life-threatening complications.

References

1. Manduz Ş, Altuntaş EE, Katrancıoğlu N, Karahan O, Doğan K. Karotid body tumors. Turkiye Klinikleri J Cardiovasc Sci 2009; 21: 52-7.

2. Mataracı İ, Rabuş MB, Kırali K, Kıran B, Yanartaş M, et al. Diagnosis and surgical treatment of carotid body tumors. Turkish J Thorac Cardiovasc Surg 2008; 16: 86-90.

3. Isik A, Eryılmaz R, Okan I, Dasiran F, Firat D, Idiz O, Sahin M. The use of fibrin glue without surgery in the treatment of pilonidal sinus disease. Int J Clin Exp Med. 2014 Apr 15;7:1047-51

4. Isik A, Firat D, Peker K, Sayar I, Idiz O, Soytürk M. A case report of esophageal perforation: Complication of nasogastric tube placement. Am J Case Rep. 2014 Apr 26;15:168-71.

5. Isik A, Okan I, Firat D, Idiz O. A rare complication of colorectal surgery and its management: Chylous leakage.Cir Esp. 2015;93:118-20

6. Isik A, Alimoglu O, Okan I, Bas G, Turgut H, Sahin M. Dieulafoy lesion in the stomach. Case Rep Gastroenterol. 2008 ;2:469-73.

7. Boedeker CC, Neumann HP, Maier W, Bausch B, Schipper J, Ridder GJ. Malignant head and neck paragangliomas in SDHB mutation carriers. Otolaryngol Head Neck Surg 2007; 137: 126-9.

8. Üçüncü H, Aktan B, Erdoğan F, Eren S, Sütbeyaz Y. Laryngeal paraganglioma causing air way obstruction: a case report. Turk Arch Otolaryngol 2005; 43: 37-41.

9. Shamblin WR, ReMine WH, Sheps SG, Harrison EG Jr. Carotid body tumor (chemodectoma). Clinicopathologic analysis of ninety cases. Am J Surg 1971; 122: 732-9.

10. Leonetti JP, Donzelli JJ, Littooy FN, Farrell BP. Perioperative strategies in the management of carotid body tumors. Otolaryngol Head Neck Surg 1997; 117: 111-5.

11. Jeevan DS, Saleh M, LaBagnara M, Neil JA, Hillard VH. Malignant carotid body tumor presenting with myelopathy: case report. J Neurosurg Spine. 2016 Jan 1:1-4.

12. Dixon JL, Atkins MD, Bohannon WT, Buckley CJ, Lairmore TC. Surgical management of carotid body tumors: a 15-year single institution experience employing an interdisciplinary approach. Proc (Bayl Univ Med Cent). 2016 Jan;29(1):16-20

Figure Legend

Figure 1: Right carotid angiography at the level of the carotid bifurcation.

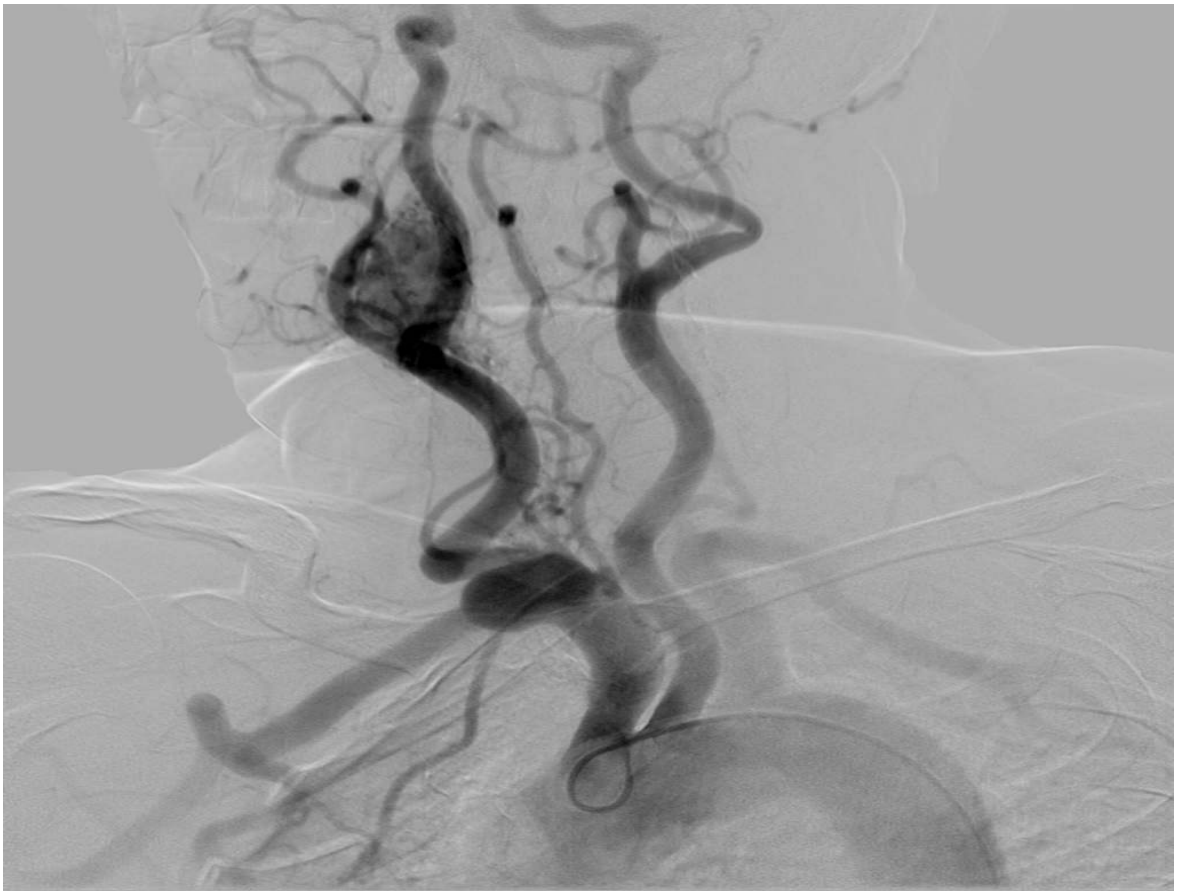
a: in the early arterial phase, a mass lesion with regular contours showing heterogenous contrast of moderate intensity.

b: in the late arterial phase, a mass lesion showing intense contrast enhancement.

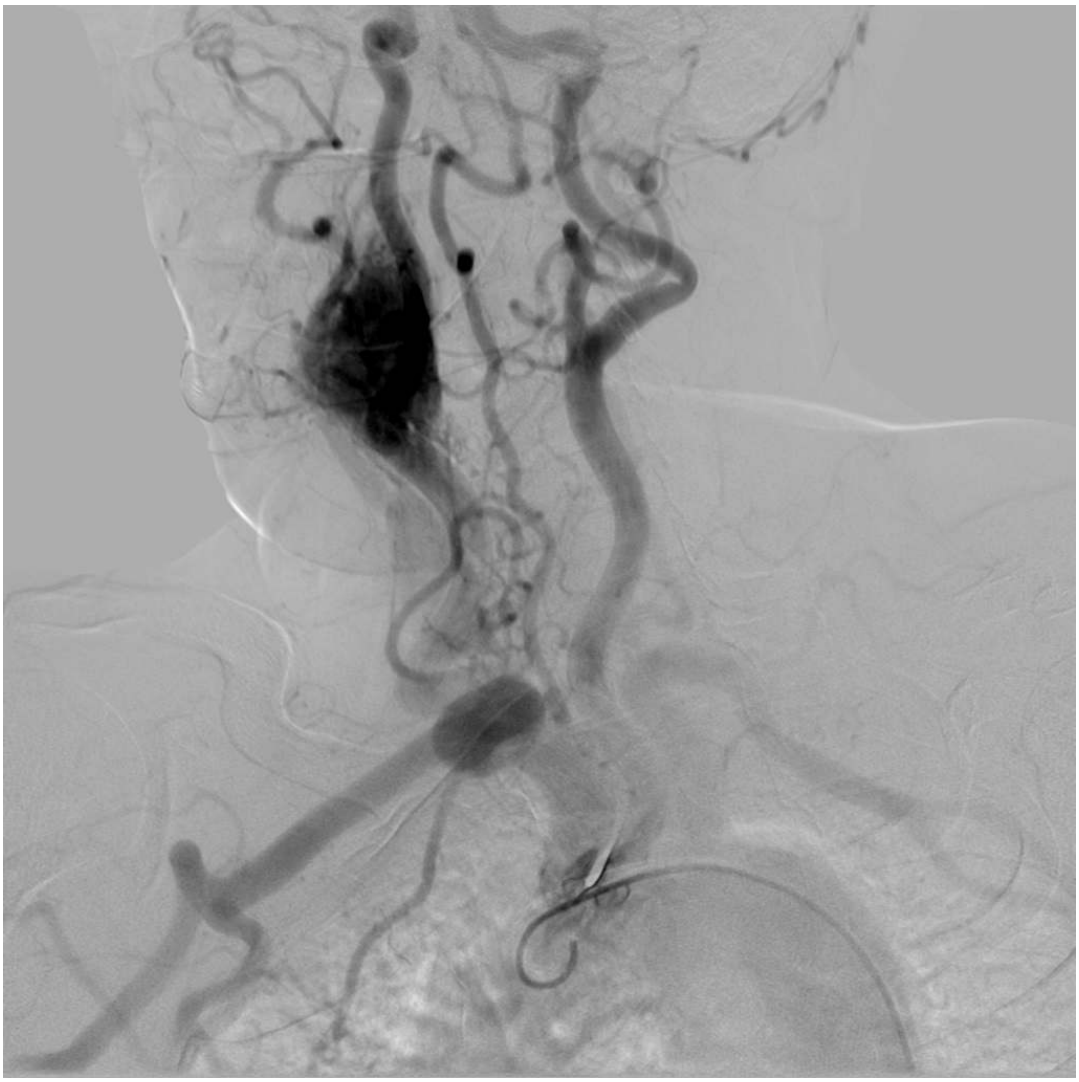
Figure 2: Histopathological image of the paraganglioma.

a: the overall picture (Hematoxylin-Eosinx40).

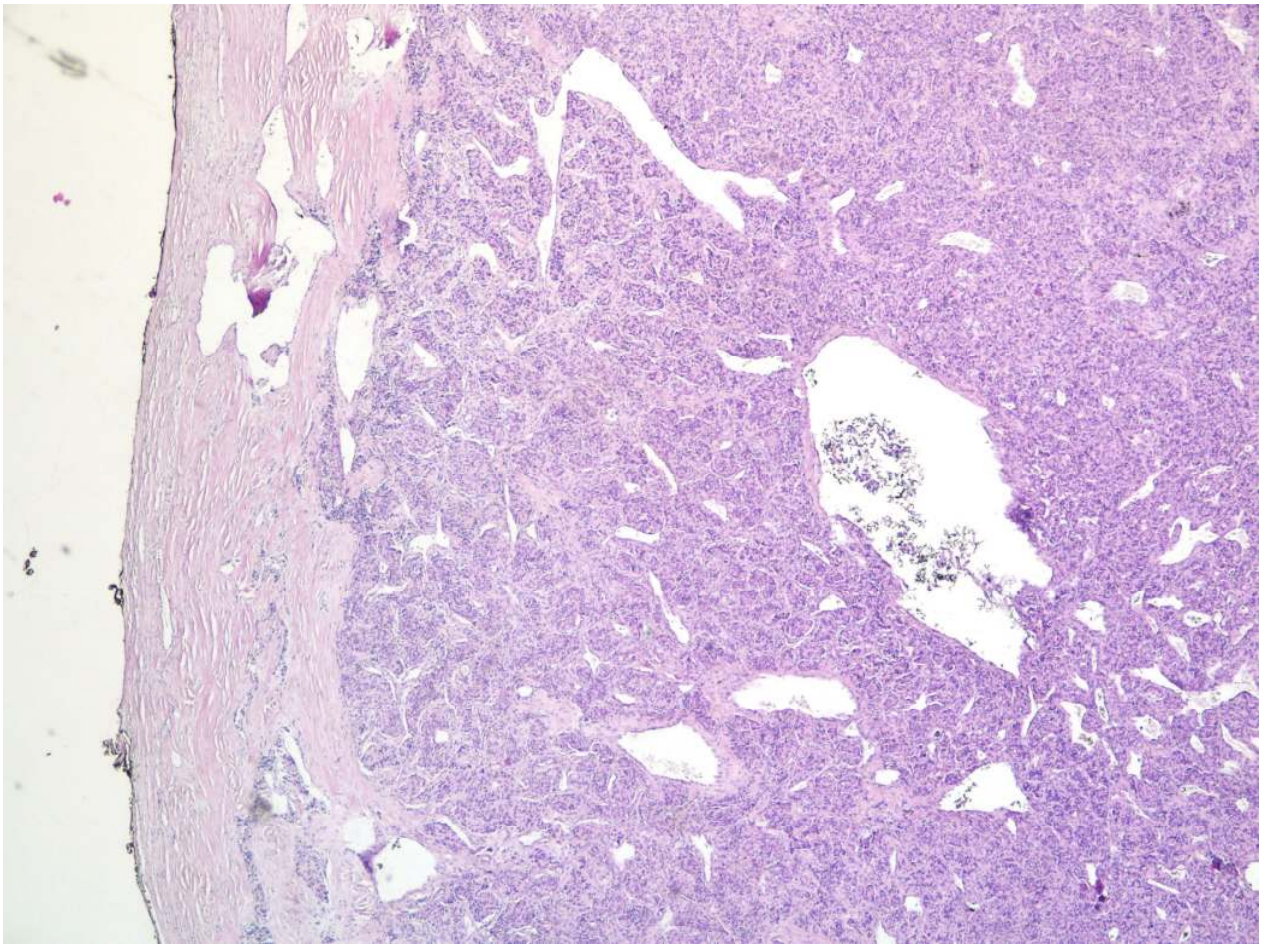
b: Zellballen pattern of paraganglioma (Hematoxylin-Eosinx200).



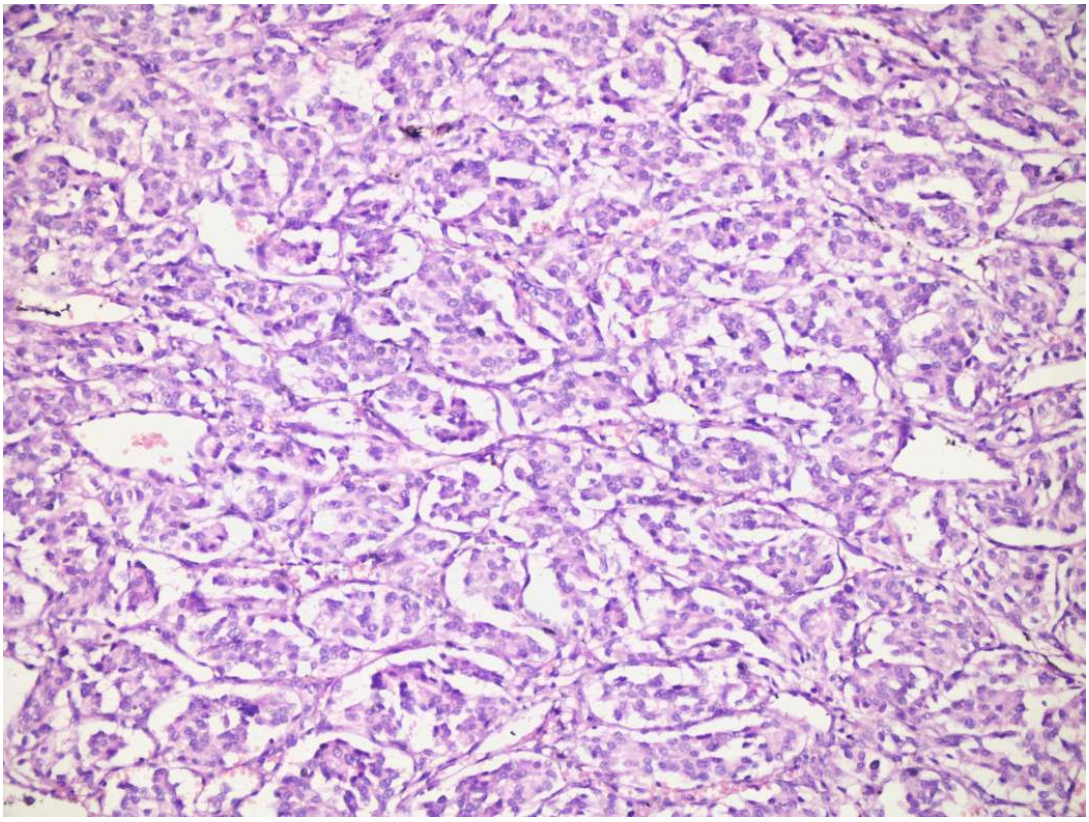
1a



1b



2a



2b