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Ganglioneuroma in The Neck Region: A Case Report

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ABSTRACT

Ganglioneuroma, which rarely occurs in the neck region, is a well-differentiated benign tumor of the sympathetic nervous system. A 39-year-old man presented with neck swelling for a year. A giant tumor was located on the left side of the neck. Schwannoma was suspected on preoperative cytology and core needle biopsy, and the postoperative diagnosis on pathological examination was ganglioneuroma. The origin of the tumor was considered to be the sympathetic nervous system based on the postoperative appearance of Horner's syndrome. The treatment choice for ganglioneuromas is complete surgical resection, contrary to the nerve-sparing resection of schwannomas. Therefore, making the definitive diagnosis before surgical resection could be important. In this report, we focus on the differential diagnosis obtained with radiological findings and the treatment strategy for the tumor with a literature review.

Key words: *ganglioneuroma, head and neck, neurogenic, diagnosis*

INTRODUCTION

Ganglioneuroma (GN) is a benign tumor arising from the sympathetic ganglion. It commonly occurs in the posterior mediastinum, retroperitoneum, and adrenal glands[1–3)]. It is rare in the head and neck region and presents as a slow-growing painless neck mass[1–4)]. It is often initially detected as a neck swelling/mass or abnormal radiological findings. The definitive diagnosis is usually made after surgical resection. Herein, we report a case of GN presenting as a swollen mass in the lateral neck region on the left side. Surgical resection was performed, and histopathology revealed cervical GN. We describe the case with emphasis on the differential diagnosis including schwannoma and review the literature.

CASE REPORT

A 32-year-old man with painless lateral neck swelling on the left side for one year visited our hospital for consultation. He had no relevant medical history, except allergic rhinitis. Physical examination showed an elastic, hard, immobile tumor on the left side of the neck. The lesion showed no other signs, such as tenderness, redness, or rise in temperature. He had no abnormal neurological findings. The blood chemistry results were within the normal ranges.

Neck ultrasonography revealed a fusiform-shaped tumor measuring approximately 100 mm in diameter. It showed an isoechoic, vascular region with clear margin (Figure 1). Flexible laryngoscopy showed deviation of the nasopharyngeal wall and compression by the tumor. No vocal fold palsy or mucosal lesions were seen (Figure 2). Contrast-enhanced computed tomography (CT) revealed a slightly enhanced tumor located behind the common carotid artery (Figure 3). The tumor showed an isointense low signal intensity on T1-weighted magnetic resonance imaging (MRI) and both low and high signal intensities on T2-weighted MRI (Figure 4). The definitive diagnosis could not be made with fine-needle aspiration cytology. Pathological findings on core-needle biopsy suggested a schwannoma with an Antoni B area, as epithelioid cells were CD68- and S100-positive and epithelial membrane antigen-negative (Figure 5). The examination findings were described to the patient in detail, and total surgical resection of the tumor was suggested. The patient and his family provided consent for the surgery, regardless of the possible surgical complications, including palsy of the nerve from which the tumor originated.

A T-shaped skin incision was made on the surface of the tumor. After exfoliating the frontal edge of the sternocleidomastoid muscle, the vagus nerve, hypoglossal nerve, accessary nerve, and carotid artery were identified and separated from the tumor. Most of the tumor capsule did not adhere to the surrounding tissues. However, there was a fibrous nerve-like bundle connected to the surrounding tissues. We totally resected the tumor sacrificing the bundle, as none of the nerves in the bundle

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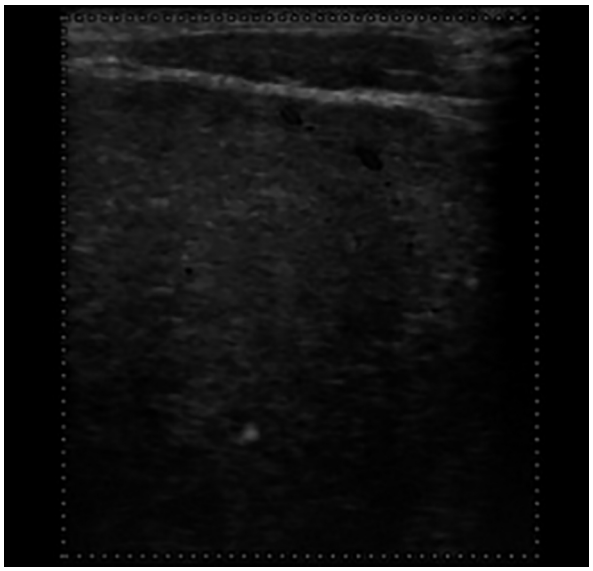

Figure 1 Ultrasonography. Ultrasonography depicts the entire tumor. The tumor appears fusiform-shaped and 10 cm in diameter with clear margin and an isoechoic, vascular region inside.

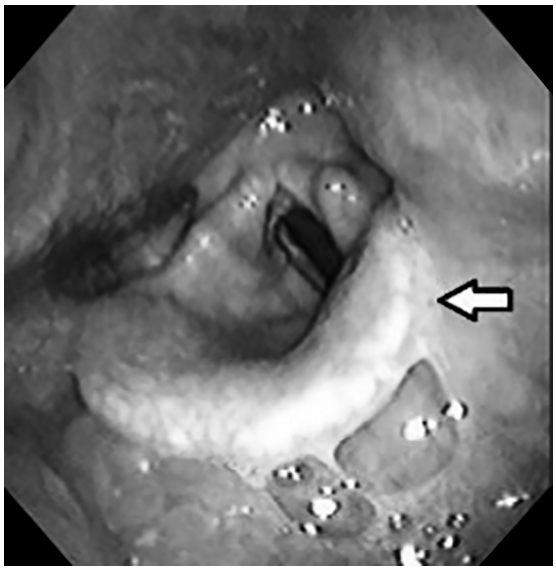

Figure 2 Endoscopic findings. The lateral wall of the nasopharynx is compressed by the tumor (arrow). The mucosal surface remains intact, and no vocal cord palsy is observed.

could be identified (Figure 6).

Macroscopic findings showed a solid, yellow tumor measuring 105 × 70 × 35 mm with clear margin. Microscopic findings on hematoxylin and eosin staining showed a fibrillary skin capsule around the tumor containing the ganglion of the nerve from which the tumor originated. The spindle-shaped tumor cells had grown profusely and showed partial differentiation in the ganglion without neuroblasts. Immunohistology showed positive staining for S100, neural cell adhesion molecule, synaptophysin, and chromogranin A. The Ki-67 labeling index was as low as 1%. The findings were compatible with the pathological diagnosis of GN (Figure 7).

Postoperatively, the patient had slight miosis and blepharoptosis in the left eye. No other neurological disorders, such as hoarseness, dysphagia, or phrenic nerve palsy, were observed. Based on the findings, the tumor seemed to originate from the sympathetic nerve. The patient was discharged from the hospital on postoperative day 8 without major complications. No recurrence was observed at the one-year follow up.

DISCUSSION

Peripheral neuroblastic tumors are classified into three types based on the degree and type of neuroblastic differentiation and the degree of Schwannian stroma development: neuroblastoma, ganglioneuroblastoma, and GN[8]. All these tumors are derived from the neural crest cells but differ in the stage of maturation, including undifferentiated malignant, partially differentiated, and fully differentiated benign cells[7]. GN is a well-differentiated begin tumor, similar to paraganglioma and pheochromocytoma. GN is pathologically diagnosed with a tumor configuration of multiplying ganglion cells, Schwann cells, and fibrous tissues without neuroblast cells[10]. The peak age of onset is in the young-adult age group and no difference between the sexes have been reported[1,3,4,11,12]. The common locations are the posterior mediastinum (39%), retroperitoneum (30%), adrenal glands (22%), and neck region (8%)[3]. In the head and neck region, most GNs originate from the cervical ganglion and a few from the vagus nerve ganglion or carotid bulb[1,3,4,8,12]. As GN is typically asymptomatic, the tumor

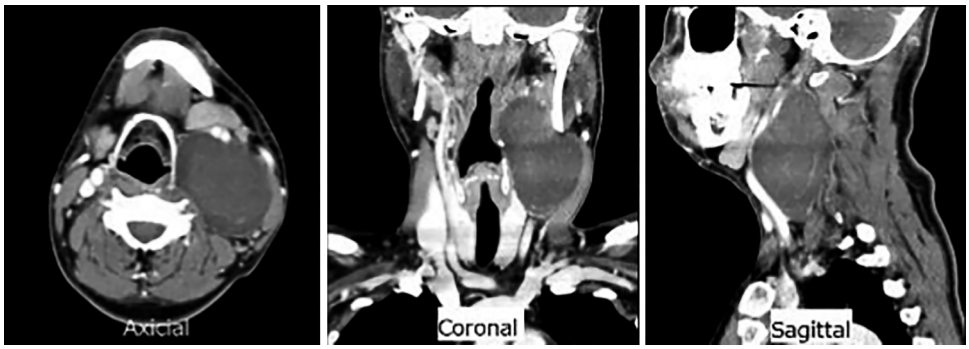

Figure 3 Contrast-enhanced computed tomography. A bulky tumor is located behind the common carotid artery on the left side of the neck. The tumor size is 100 × 50 mm, and the carotid artery has shifted forward.

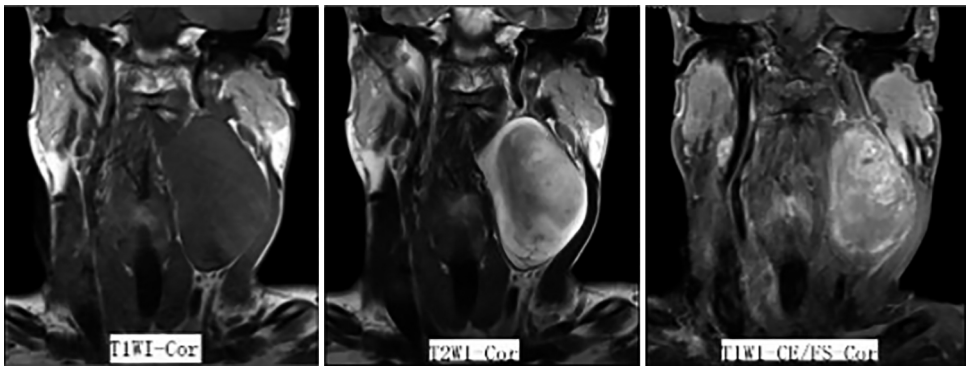

Figure 4 Magnetic resonance imaging (MRI). The tumor shows isointense, low signal intensity on T1-weighted MRI, both low and high signal intensities on T2-weighted MRI, and high signal intensity on Gd-enhanced T1-weighted MRI.

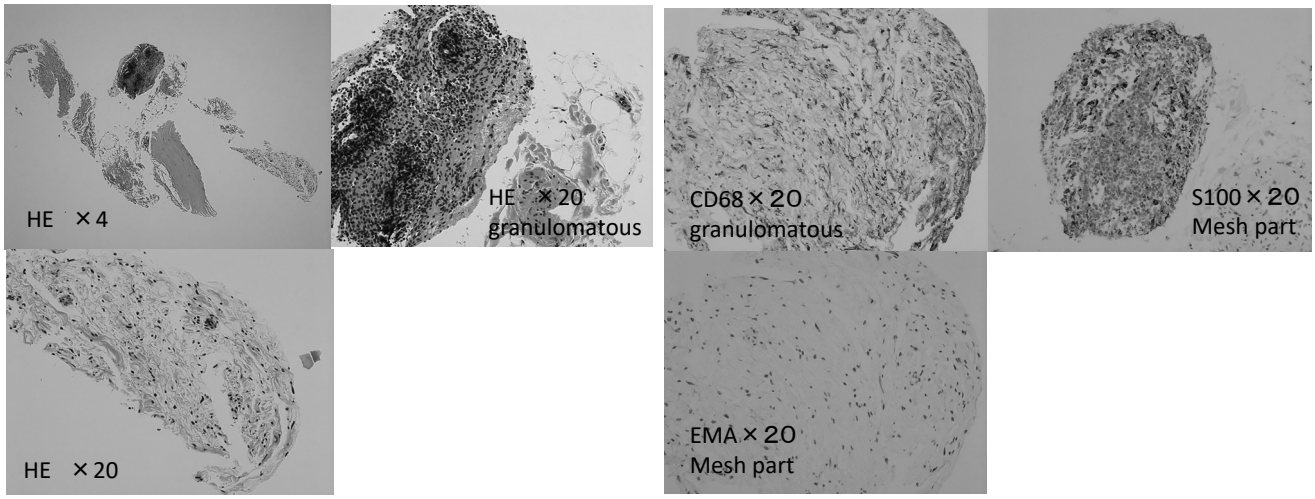

Figure 5 Histopathology of needle biopsy specimen. Left: Hematoxylin and eosin staining. There are granulomatous and reticular parts. Right: Immunostaining shows a CD68- and S100-postive and epithelial membrane antigen-negative granulomatous part.

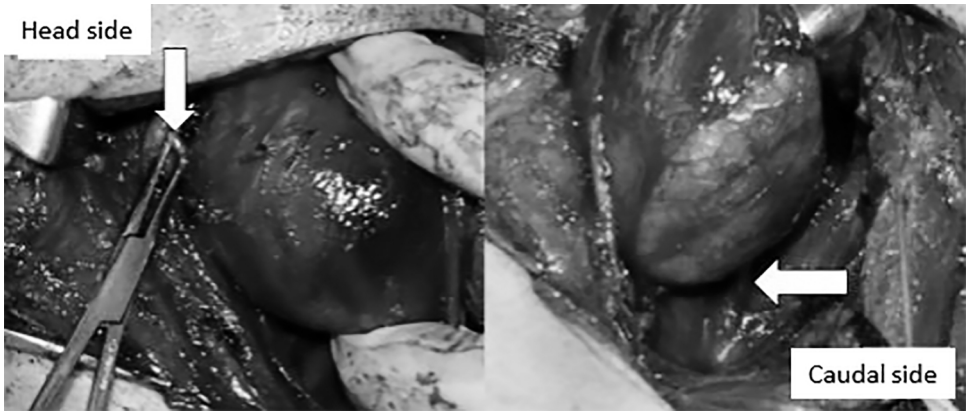

Figure 6 Intraoperative findings. A fibrous cord, which seems to be a series of derived neurons (➡), is observed on both sides of the tumor in the vertical direction.

can be incidentally detected on clinical imaging performed for another purpose. Symptoms may result from tumor invasion of the surrounding organs. In some cases, catecholamine hormones are produced, which may cause diarrhea, high blood pressure, and increased sweating[9,11]. The patient in this case had no symptoms other than the neck swelling.

GN can be detected on CT, MRI, and ultrasonography. Blood and urine analyses may be useful when the tumor secrets hormones. The specific findings of a spring-like cord connected to the spindle-shaped tumor on imaging help diagnose neurogenic tumors. CT shows homogeneous or heterogeneous hypodense areas with punctuated calcification. Some lesions with fat density may present as heterogeneous high signal intensity on T2-weighted MRI. In some cases, a whorled appearance can be found as a non-enhanced or mildly enhanced lesion in the arterial phase of CT or MRI and as a progressively enhanced lesion in the delayed phase. These findings could help distinguish GN from schwannoma, which usually shows

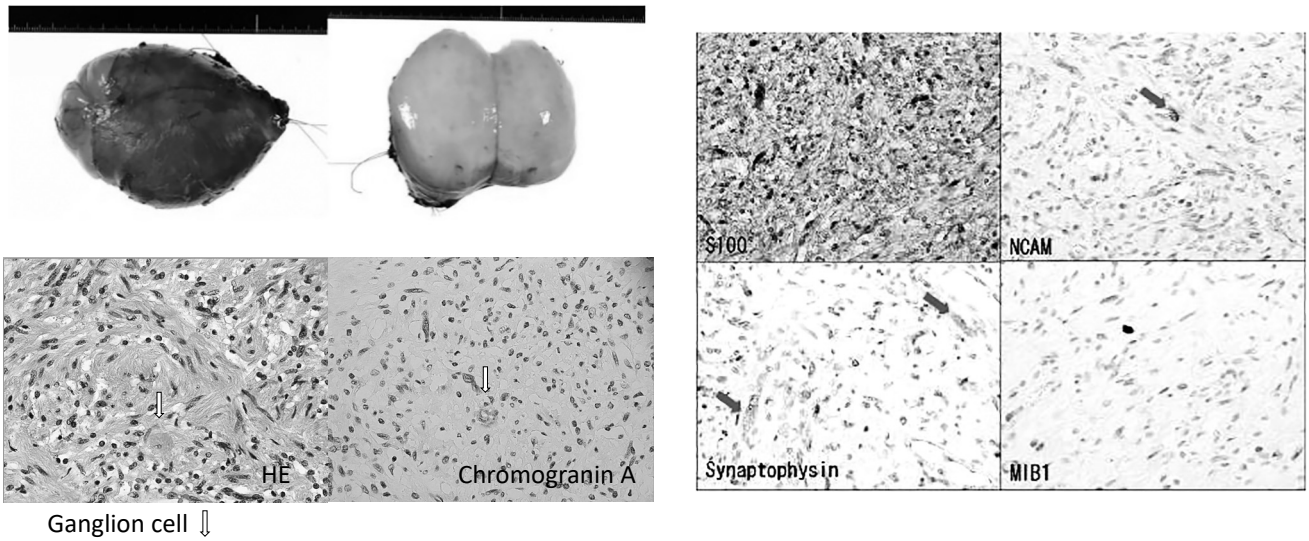

Figure 7 Macroscopic and microscopic findings. Upper left: A pale, yellowish white, solid tumor measuring 10.5 × 7 × 3.5 cm. The cleaved surface has a yellowish white and rubber-like appearance. There is no hemorrhagic necrosis inside. Lower left: Hematoxylin and eosin staining shows complicated proliferation of the spindle-shaped tumor cells. Some cells show ganglionization. Right: Immunostaining shows that most tumor cells are positive for chromogranin A and S100 and that the concave, spindle-shaped scattered cells (⇑) are positive for neural cell adhesion molecule and synaptophysin. The Ki-67 labeling index (the MIB1 positive rate) is approximately 1%.

complex structures because of partial necrosis or bleeding inside the tumor[6]. This case showed low intensity band alignment rather than a typical whorled appearance on T2-weighted MRI. It may be difficult to confirm the diagnosis before the surgery, as the tumor may not show the characteristic findings.

As neurogenic tumor cells generally have strong connections, the definite diagnosis cannot be made with fine-needle aspiration cytology[7,12] and an open biopsy may be required. Core-needle biopsy before the surgery in this case suggested schwannoma with distinctive cytological features excluding malignancy. The biopsy may also have been beneficial for the patient and preparation for the surgery. In this case, the amount of specimen collected with the needle biopsy was not sufficient, so there was a discrepancy with the postoperative diagnosis. However, based on the immunostaining findings, GN was included in the differential diagnosis.

The treatment strategy for GN remains controversial[2,3,5,11,12]. Surgical resection is not mandatory, as GN is benign, and resection may cause neurologic complications. However, it is often difficult to obtain the definitive diagnosis before the surgery, and some neurogenic tumors have malignant potential. Therefore, surgery should be considered in cases of presence of neurological symptoms, increase in tumor volume, hormonal imbalance, and major physical deformities. Additionally, the patient should be informed about the possibility of associated neurological complications and risk of malignancy.

Due to the rarity of GN in the head and neck region, it is difficult to conduct studies with a large sample size to establish evidence-based treatment strategies. However, further studies are warranted, especially for preoperative diagnostic strategies.

CONCLUSION

We reported a case of GN in the neck region. As GN is uncommon in the head and neck region, and treatment strategies have not been established, further investigations are required on strategies to obtain the preoperative diagnosis.

Conflict of interest

The authors have no conflicts of interest to declare.

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