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Case report

Dysphagia caused by giant schwannoma of the supraglottic oropharynx: A case report and review of the literature

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HIGHLIGHTS

- Schwannoma is a benign peripheral nerve tumor that usually presents as a slow growing single lesion.
- Giant schwannoma of the supraglottic oropharynx is rare.
- The main symptom is dysphagia due to the location and mass of the tumor.
- MRI is particularly helpful in delineating schwannoma.
- Surgery is the treatment of choice.

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ABSTRACT

Background: Schwannomas are benign, encapsulated, peripheral nerve tumours that arise from the Schwann cell. Approximately 25%–45% of schwannomas occur in the head and neck. The most common site is the parapharyngeal space of the neck. However, schwannoma of the supraglottic oropharynx is rare.

Case presentation: We report on a 35-year-old female who complained of progressive dysphagia, from whom a large schwannoma in the supraglottic oropharynx was excised through a transoral approach. No recurrence was seen after one year follow-up.

Conclusion: Although rare, schwannomas do occasionally occur in the supraglottic oropharynx. When dysphagia is present, a thorough diagnostic procedure should be performed to evaluate the morphology and function of the upper aerodigestive tract. MRI is sensitive and specific in the diagnosis of schwannoma. And the best treatment of choice is complete excision with preservation of functions.

1. Introduction

Schwannomas are benign nerve sheath neoplasms originating from Schwann cells that produce the myelin sheath of peripheral nerves. These tumors have been documented at sites throughout the body, including the head and neck, an anatomic region in which they are particularly common [1,2]. However, a schwannoma of the supraglottic oropharynx is a rare tumor in the head and neck region arising from the sheath of cranial nerves and/or the peripharyngeal plexus [3]. And Reports are often limited to case reports and small case series. They tend to affect women more often than men and are most frequently seen in the third to sixth decades of life. The main symptom is dysphagia due to the location and mass of the tumor. Here we present a case of dysphagia caused by giant schwannoma of the supraglottic oropharynx and review the literature regarding the diagnosis and treatment of supraglottic oropharynx schwannoma.

2. Case presentation

A 35-year-old female patient was admitted to Cangzhou Clinical College of Integrated Traditional Chinese and Western Medicine of Hebei Medical University. The patient was referred to Otorhinolaryngology department with the complaint of dysphagia and pharyngeal foreign body sensation for 3 months. She had reported sore throat and irritating cough when eating. TV-monitored rhinolaryngofiberscope revealed a large mass in the left pharyngeal wall reaching to glottis in the supraglottic oropharynx (The extent

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of disease involved not only the parapharyngeal space but also likely resulted in external pressure on the supraglottic larynx). The mucosa of the pharyngeal bulge was smooth and intact. But the large mass boundary was obscure [Fig. 1**A**]. At that time, the patient refused to accept CT and MRI scan due to have recent fertility requirements.

On march 24,2016, pharyngeal neoplasm resection was performed in order to relieve dysphagia. From the preoperative evaluation, it appeared that the entire tumor could easily be widely exposed and removed without injuring the great vessels and major nerves. A dumbbell-shaped, well-encapsulated, yellowish tumor measuring about 9.5x2.5 cm was removed completely from the pharyngeal space through a transoral approach [Fig. 1**B**]. Pathological examination showed a cellular schwannoma composed of compact (Antoni A) areas and loose meshed (Antoni B) areas. The cells were spindle-shaped and some cells arranged in a fence-like or incomplete swirling. The tumour was made of spindle cells with wavy nuclei showing nuclear pleomorphism with thick walled blood vessels. Few lymphocytes were scattered in between the tumour cells [Fig. 1**C**]. Immunohistochemical stainings with various markers were performed for differential diagnosis with the following results:S-100 (+),SMA (vessel+),Ki-67 (+<1%),CD34 (vessel+),Desmin (−) [Fig. 1**D**].

The pathologic diagnosis was schwannoma.

The patient was started on oral feeds on the first postoperative day. She was subsequently discharged on the third postoperative day. There has been no evidence of recurrence or any other complaint for the last one year.

3. Discussion

Schwannoma (also known as neurilemmomas) is a benign peripheric nerve tumor that usually presents as a slow growing single lesion; it has origin in Schwann cells proliferation [3]; They are solitary, encapsulated, benign tumors arising from the Schwann cells of the peripheral, cranial, and autonomic nerves [4]. And it was first described by Verocay [5]. Schwannoma occurs in overall body areas including the head and neck region; And it usually arise from the cranial (10%) and spinal (25%) nerve roots [6]. They can arise at any site of the peripheral nervous system, with the exception of the olfactory and optic nerves. The nerve origin of the tumour is most likely to be the peripharyngeal plexus. In most of the cases, between 25 and 45%, extracranial schwannoma occurs in the head and neck region. In the oral cavity the lesion is usually present in soft tissues, more commonly the tongue, followed by the palate and buccal mucosa, and may have clinical aspects similar to other benign lesions like mucocele, fibromas, lipomas, and benign salivary gland tumors [7,8]. The most common site is the parapharyngeal space of the neck. The clinical signs and symptoms vary according to the size and location of the tumor, and the nerve of origin. However, isolated schwannoma of the supraglottic oropharynx is rare. Holinger and Johnston [9]found only one case of Schwannoma among 1197 benign lesions of the supraglottic oropharynx. New and Erich [10] found only one case of schwannoma of 722 benign supraglottic oropharynx tumours. They realized that supraglottic oropharynx schwannomas were nearly always solitary, submucosal, well circumscribed and occasionally

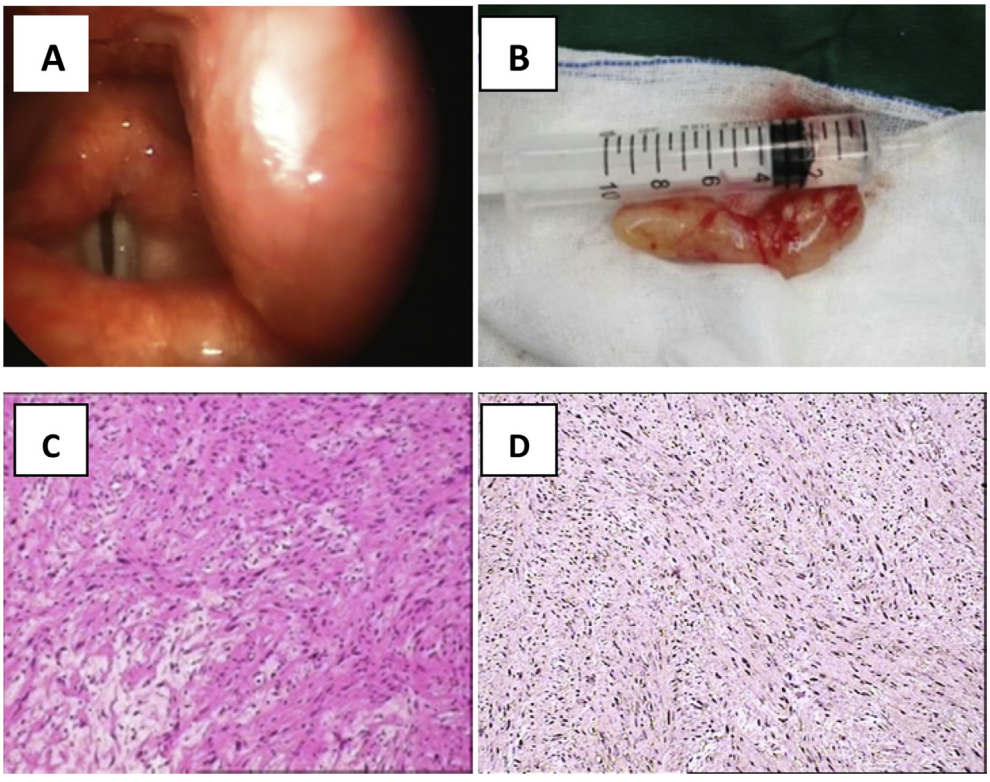

Fig. 1. A.TV-monitored rhinolaryngofiberscope showed a large mass in the left pharyngeal wall reaching till glottis in the supraglottic oropharynx. The mucosa of the pharyngeal bulge was smooth and intact. But the large mass boundary was obscure.
Fig. 1B. A dumbbell-shaped, well-encapsulated, yellowish tumor measuring about 9.5x2.5 cm was removed completely from the pharyngeal space through a transoral approach.
Fig. 1C. Histopathological examination showed a cellular schwannoma composed of compact (Antoni A) areas and loose meshed (Antoni B) areas. The cells were spindle-shaped and some cells arranged in a fence-like or incomplete swirling. The tumour was made of spindle cells with wavy nuclei showing nuclear pleomorphism with thick walled blood vessels. Few lymphocytes were scattered in between the tumour cells (magnification, x10).
Fig. 1D. Immunohistochemical stainings with various markers were performed for differential diagnosis with the following results:S-100 (+),SMA (vessel+),Ki-67 (+<1%),CD34 (vessel+),Desmin (−)(stain, hematoxylin and eosin; magnification, x10).

painful. Dysphagia and shortness of breath are the usual presenting symptoms of supraglottic oropharynx schwannomas mentioned in the literature. In this case, where the tumour was exceptionally large reaching to glottis in the supraglottic oropharynx, complete tumor removal was the preferred choice of treatment. Not only did an accurate histologic diagnosis have to be determined, but also the dysphagia and pharyngeal foreign body sensation had to be resolved. If the tumor was left behind, it could grow progressively larger, possibly leading to upper airway obstruction.

As schwannomas are inherently slow growing and noninvasive, most patients present with an asymptomatic mass without nerve dysfunction; When schwannomas are located in the supraglottic oropharynx, they may cause dysphagia and dyspnea or impair phonation [3]. Differential diagnosis includes: abscess, hematoma, fibroma, neurofibroma, hemangioma, malignant schwannoma, lipoma and so on. In fact, it is difficult to determine the nerve of origin without conducting imaging studies before surgical exploration. Among the imaging modalities, CT and MRI are by choice. MRI with and without gadolinium enhancement is particularly helpful in delineating schwannoma [11,12]. This case refused to accept CT and MRI scan due to have recent fertility requirements. On MRI, schwannomas show low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Following the administration of gadolinium, the solid part of the tumor is strongly enhanced, while the cystic component remains unchanged. Schwannomas can be distinguished from paragangliomas by the contours of the tumor, destruction of adjacent tissues, and vascular stains shown on magnetic resonance angiography. Furthermore, the location and relationship between the tumor and adjacent structures allow the nerve of origin to be predicted preoperatively, thus allowing surgical risks to be evaluated [13]. Ryuji Yasumatsu et al. believe that MRI is sensitive and specific in the diagnosis of schwannoma [2]. In fact, although preoperative CT and MRI may provide information regarding the diagnosis of schwannoma, it can only be confirmed by postoperative histopathologic examination [4].

Surgery is the treatment of choice. An endoscopic approach can be attempted for smaller tumours but for large tumours, external approaches like lateral pharyngotomy are used. In this case, where the tumour was exceptionally large reaching the supraglottic oropharynx, we were able to remove the tumour by transoral approach.

4. Conclusion

The purpose of this paper is to call attention to the fact that a common soft tissue tumor like schwannoma can also present as a giant supraglottic oropharynx tumor resulted in dysphagia. Although rare, schwannomas do occasionally occur in the supraglottic oropharynx. When dysphagia is present,a thorough diagnostic procedure should be performed to evaluate the morphology and function of the upper aerodigestive tract. MRI is sensitive and specific in the diagnosis of schwannoma. And the best treatment of choice is complete excision with preservation of functions.

Ethical approval

Ethical Approval was given.

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Authors' contributions

Li Guojian, substantial contributions to conception and design of the study, acquisition of data, or analysis and interpretation of data; Xin Xiao, Wang Xia, Ren Chongxi, drafting the article or making critical revisions related to important intellectual content of the manuscript; and final approval of the version of the article to be published.

Conflict of interest statement

No potential conflict of interest relevant to this article was reported.

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Ren Chongxi.

Consent of patient

The consent of the patient has been obtained.

SCARE criteria

This case has been reported in line with the SCARE Criteria [14].

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