

Schwannoma of the posterior pharyngeal wall – case report

Guz tylnej ściany gardła pochodzenia neurogennego – opis przypadku

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ABSTRACT: The authors present a case report of a 46-year-old man with a tumor of the posterior pharyngeal wall. Based on clinical examination and magnetic resonance imaging, the patient was qualified for surgical treatment. The patient underwent tumor resection under general anesthesia. Histopathological examination of resected tumor revealed the diagnosis of ancient schwannoma.

KEYWORDS: oropharynx, schwannoma, surgery

STRESZCZENIE: Autorzy przedstawiają przypadek kliniczny 46-letniego mężczyzny z guzem tylnej ściany gardła. Na podstawie obrazu klinicznego i badania rezonansu magnetycznego, pacjenta zakwalifikowano do leczenia chirurgicznego. Chory został poddany resekcji guza w znieczuleniu ogólnym z dostępu przez jamę ustną. Na podstawie badania histopatologicznego otrzymanego materiału postawiono rozpoznanie nerwiaka osłonkowego.

SŁOWA KLUCZOWE: chirurgia, gardło środkowe, nerwiak

INTRODUCTION

Schwannomas (*neurilemmoma*, *neurinoma*, *schwannoma*) represent a group of rare, benign neurogenic tumors derived from Schwann cells that produce the sheaths of the cranial and peripheral nerves. Between 25% to 45% of the schwannomas are located in the head-and-neck region [1, 2], predominantly in the parapharyngeal space [2–4] and in the area of cerebellopontine angle, while the less common sites are the oral cavity, nasal cavity and paranasal sinuses. Among head and neck neoplasms, schwannomas account for approx. 1% [1]. The highest percentage of schwannomas originate from the vagus nerve or sympathetic trunk. Oropharyngeal occurrence of schwannoma, and especially on the posterior wall of the oropharynx, belongs to casuistry. Currently available literature from the years 1977–2017 presents only 13 such cases. In this location, schwannomas are more common in females when compared to men, and they usually occur between the 3rd and 6th decades of life [2, 5].

Schwannomas typically occur as a single tumor; nevertheless there are cases of multiple schwannomas, incl. in neurofibromatosis type 2, schwannomatosis, Carney complex [6]. Reports have also included isolated cases of schwannomas of the vestibular nerve after radiotherapy with a latency period of up to 50 years [6].

Symptoms of schwannoma in the oral cavity and oropharynx depend on their location and tumor growth dynamics. Symptoms may include: dysphagia, odynophagia, pharyngeal foreign body sensation, swelling in the oropharynx, phonation disorders or apnea [2]. In the initial stage, schwannomas may be asymptomatic.

Treatment consists of surgical resection of the tumor with the surrounding capsule. In case of the cervical schwannomas, common practices also include intracapsular resection aimed at minimizing the functional disorder of the nerve from which the tumor develops [7]. The selection of the appropriate procedure technique depends primarily on the exact location of the lesion and its size.

CASE REPORT

A 46-year-old patient was admitted to the Department of Otorhinolaryngology, Head and Neck Surgery at the Medical University of Warsaw for surgical treatment of a tumor of the posterior wall of oropharynx on the left side, detected by magnetic resonance imaging (MRI) accidentally. The patient denied any complaints related to the tumor. The patient had undergone bilateral inguinal hernia repair, appendectomy, cholecystectomy and septoplasty in the past.

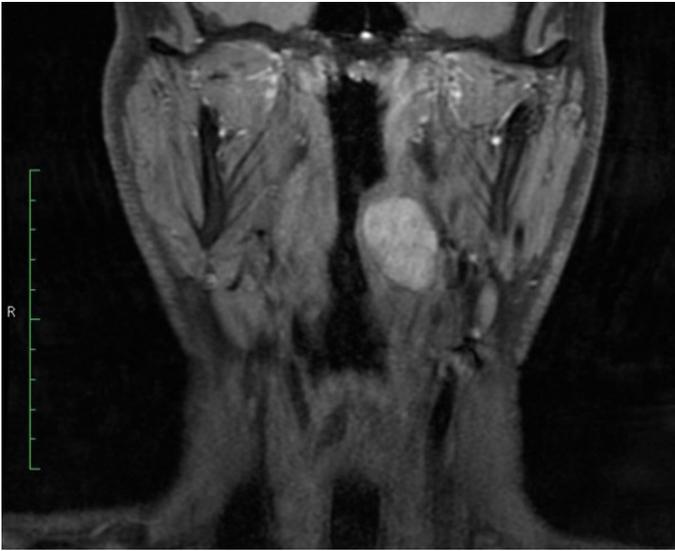


Fig. 1. Frontal MRI of the head in the T2-weighted sequence after administration of contrast agent – hyperintense lesion showing a tumor of the left posterior wall of pharynx.

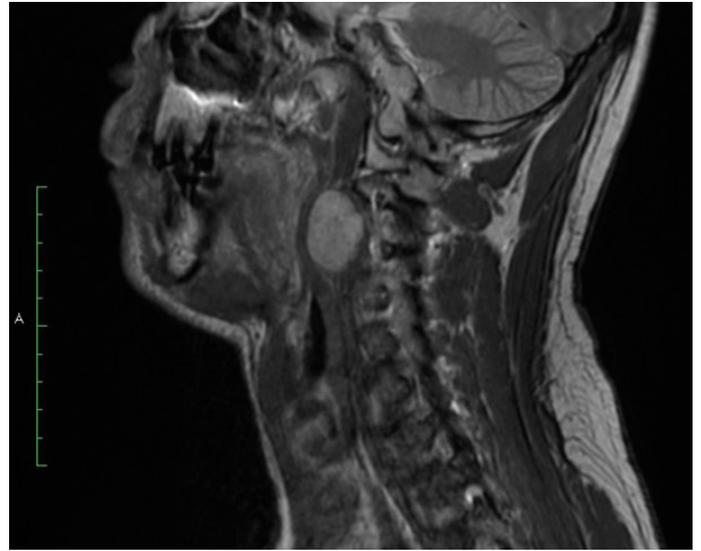


Fig. 3. Sagittal MR of the head in the T2-weighted sequence after contrast administration – tumor of the posterior pharyngeal wall.

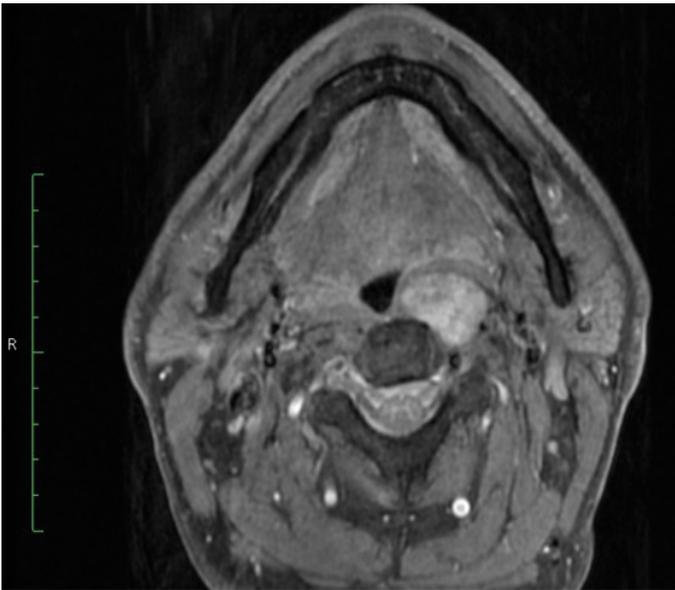


Fig. 2. Axial cross-section MR of the head in the T2-weighted sequence after administration of contrast agent – tumor of the posterior wall of pharynx on the left side

Contrast-enhanced MR imaging of the neck provided by the patient and performed during the initial outpatient diagnosis revealed a well-delimited lesion of 24 x 19 x 32 mm posterior to the left palatine tonsil, hyperintense on T2-weighted sequence, showing contrast enhancement after administration of gadolinium and diffusion restriction. The lesion pushed the tonsil forward; it was situated at a depth of approx. 3 mm. It modeled and narrowed the lumen of oropharynx on the left side. The radiological picture was ambiguous; the possibility of the origin of the tumor from small salivary glands or pathologies of the retropharyngeal lymph nodes was suggested.

Otorhinolaryngological examination showed no significant deviations from the normal state, except for the presence of a tumor located within the posterior wall of oropharynx, which protruded into the pharyngeal lumen. The mucosa above the tumor remained unchanged.

After anesthesiologic consultation, the patient was qualified for removal of the lesion under general anesthesia.

During the procedure, a tonsil retractor was placed to reveal the posterior pharyngeal wall. A convex wall, which was firm and slightly susceptible to pressure during palpation, was revealed on the left side. The mucosa was incised above the tumor, followed by incision of the constrictor muscle. A specimen was taken for an intraoperative histopathological examination. Malignancy was excluded, and the polycyclic tumor with a smooth white capsule of approximately 3.2 x 2.5 x 1.5 cm was completely dissected. After hemostasis was achieved, the lower part of the tumor bed was underpinned and sutures were placed on the muscles and mucosa of the posterior pharyngeal wall.

The final histopathological examination revealed a tumor pattern with signs of severe degeneration, with some features of degenerative atypia. Moreover, during immunohistochemical tests, a positive expression of the S100 protein, no expression of the SMA protein (smooth muscle actin) and no expression of CD34 were observed, which allowed to make the final diagnosis of schwannoma.

There was no complications during the perioperative and postoperative periods. In the postoperative period, the patient received nasogastric tube feeding for 6 days. Healing of the wound was acceptable and the patient's general condition was good. After the feeding tube was removed, no dysphagia was observed. The patient was discharged home on the 6th day after the surgery with a recommendation of follow-up and further care at the Otorhinolaryngology Outpatient Clinic. The postoperative scar in the oropharynx was monitored during the 2-month follow-up period. There was no local recurrence. The patient discontinued further follow-up visits.

DISCUSSION

Schwannomas were first described by Verocay in 1908 [8]. These are sporadic, benign tumors of neurogenic origin. Malignant transformation is extremely rare [5]. Schwannomas can develop from

all cranial and peripheral nerves with a Schwann sheath; therefore, they are not observed in the 1st and 2nd cranial nerves (olfactory and optic nerves, respectively), which do not have such nerve sheaths. Around 25–45% of the schwannomas are located in the head-and-neck region, whereby the most common location is the cerebello-pontine angle and parapharyngeal space. In less than 1% of all head and neck schwannomas, they are located in the mouth and pharynx.

The etiology of the tumor is not fully understood. So far, no external factors have been identified that could influence the development of these tumors.

Symptoms are nonspecific. Due to the low mass of the tumor, the initial clinical course may be asymptomatic.

Differential diagnosis of oropharyngeal tumors developing under an unchanged mucosa should include both inflammatory changes, benign and malignant neoplasms, taking into account i.a. lymphomas, sarcomas, tumors developing from minor salivary glands.

Clinical examination should include a thorough otorhinolaryngological examination, extended with imaging examinations (computed tomography and magnetic resonance imaging). The most common imaging examination, which is considered the gold standard in suspected schwannomas, is contrast-enhanced magnetic resonance imaging [9, 10]. Schwannomas are typically iso- or hypodense in the T1-weighted sequences and hyperdense in the T2-weighted sequences, which corresponds to the Antoni B area identified by histopathology. In addition, ancient schwannomas are strongly enhanced after the administration of a gadolinium contrast medium, which is characteristic of the Antoni A area. However, none of the above-mentioned features is characteristic enough for making a clear diagnosis. Biopsy should be considered if radiographic examination has excluded a vascular lesion and when the location of the tumor allows for a safe biopsy. Experience in the diagnosis of head and neck schwannomas by fine needle aspiration biopsy (FNAB) is still limited. Aspirates in which Antoni A is the dominant pattern can easily be misclassified as other benign or malignant soft tissue lesions [11]. Another challenge is collecting the right amount of quality material. FNAB of schwannomas often includes obtaining hypocellular preparations, which are non-diagnostic, and in the case of cell preparations, the diagnosis of a neurogenic tumor may still be problematic [12]. Despite numerous limitations of FNAB, it is a relatively low-cost method, with a low complication rate, and therefore should be considered in the initial diagnosis [12]. The review of the available literature revealed no case of pharyngeal schwannoma diagnosed on the basis of cytological examination [13].

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Histopathological examination is necessary for the final diagnosis. The microscopic examination of schwannomas in standard hematoxylin and eosin (H&E) staining shows the presence of two main regions of the cellular pattern: Antoni A and Antoni B.

With varying frequency, the tumor pattern of Antoni A contains Verocay bodies, which are spindle cells, characterized by a swirling arrangement resembling palisades. The Antoni B pattern has a slightly looser arrangement of star-shaped cells distributed on a mucus-rich stroma. Additional immunohistochemical tests should be performed in order to confirm the diagnosis. S100 protein is the primary immunohistochemical marker for schwannomas. Vimentin, cytokeratins, CD34 protein, and GFAP are additional markers.

The clinical division created by Enzinger and Weiss [14] considers 4 types of schwannomas: common schwannoma, plexiform neurilemmoma, cellular schwannoma and ancient schwannomas (all of them benign). The ancient schwannoma from the case above was first described by Ackerman and Taylor in 1951 [15]. This tumor is characterized by extensive degenerative changes that may show a pronounced degenerative atypia of the nucleolus, which may be misinterpreted as a malignant lesion.

The procedure of choice is surgical resection. It is the most commonly used method of treating schwannomas, but for functional reasons, with consideration of the patient's age, size, location and growth dynamics of schwannomas, other treatment methods should also be considered, including: watchful waiting, intracapsular tumor resection, and even radiation therapy. The surgical procedure can be performed through the oral cavity by classical resection or with the use of laser therapy [16], or from external access, through lateral pharyngotomy [17].

The prognosis is good. Local recurrence has not been observed in the described 13 cases of schwannoma located on the posterior pharyngeal wall. It can be assumed that the schwannomas in this localization most likely derive from the glossopharyngeal nerve branches.

CONCLUSIONS

Differential diagnosis of tumors of the posterior wall of oropharynx developing under unchanged mucosa requires imaging tests: computed tomography or preferably contrast-enhanced magnetic resonance imaging. Radical surgical treatment of schwannomas in this location makes it possible to cure the disease without considerable functional disorders.

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