



Case report

Giant hamartoma of the nasopharynx – case report

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ABSTRACT

Introduction: Hamartoma is a developmental disorder.

Aim: The aim of this study is to present a case of hamartoma of naso- and oropharynx and discuss the possible surgical approaches.

Case study: A 55-year-old male patient, who had undergone a tracheotomy due to increasing breathing difficulties was admitted to Head and Neck Diseases Clinic in the Clinical University Hospital in Olsztyn with the aim of proceeding further.

Results and discussion: The parapharyngeal space is divided into the pre-styloid and the retrostyloid compartment. Tumours of parapharyngeal space are responsible for 1% of head and neck tumours; 80% of these tumours are benign lesions, most often originating from the parotid gland (50% of all the cases).

Conclusions: The treatment of choice for benign lesions like the hamartoma is a surgical resection. It is important to choose a suitable surgical approach considering morphotic parameters of a tumour, anatomic and topographic conditions.

1. INTRODUCTION

Hamartoma is a developmental disorder of the malformation type, consisting of differentiated cells specific to a given location, yet arranged chaotically. Hamartoma grows gradually, thus not manifesting characteristic nor clinical symptoms and therefore is often detected accidentally.

Tumours of this type are well demarcated from the surrounding tissues, having a non-invasive character.¹ Depending on the amount of the cellular substance two hamartoma forms are distinguished: epithelial and mesenchymal, wherein the latter occurs significantly more often.² Among the mesenchymal tissue cells four main types are identified: osteal, chondral, fibrous, benign non-matrix.^{3,4} In the featured case, the tumour character was chondromesenchymal. The said form was initially described in 1998.^{5,6}

According to the accessible data, the most common tumour locations are the lungs, the liver, the small and large intestines.^{5,7}

Hamartoma of a nasopharyngeal region is described relatively rarely in the literature. In the cranial region it is located predominantly in: the nasal cavity, the paranasal sinuses, in the nasal and pharyngeal part of the pharynx (Table 1). In the review published by Kumar et al., 19 publications on the head and neck regions' hamartomas have been analysed. Thus, most often described were hamartomas of the nasopharynx (31.6%), the nasal cavity (21.5%) and the tongue (15.8%). The tumour rarely occurred in the larynx, the middle ear, the tonsils, on the face, in the pharynx or the posterior wall of the pharynx (5.3%).

In the featured case report, the tumour of 2-piece structure had occupied, seemingly separately, the nasal and the oral part of the pharynx and had been located in parapharyngeal space. Hence in the differential diagnosis of the case, two different tumours occurring in both locations: the parapharyngeal space as well as nasal and oral part of the pharynx, were taken into consideration. In general such tumours of the pharyngeal space constitute from 0.5% to 1.0% of all head and neck tumours, 80% of which are benign neoplasm, most often (50%) stemming from the parotid gland. In the parapharyngeal space occur also the primary tumours of the paranasal

sinuses⁸ and the tonsils as well as tumours originating from the vascular and the neural trunks.⁹

2. AIM

The aim of this study is to present a case of hamartoma of naso- and oropharynx and discuss the possible surgical approaches.

3. CASE STUDY

A male patient, who had undergone a tracheotomy due to increasing breathing difficulties and gasping, reported to otolaryngological outpatient clinic. Thus a 55-year-old man with the endotracheal tube was admitted to Head and Neck Diseases Clinic in the Clinical University Hospital in Olsztyn with the aim of proceeding further.

A month before an admission to the ENT-Clinic, the patient had reported to the emergency unit with symptoms of a severe dyspnea. An urgent inferior tracheotomy was performed together with the biopsy of the middle pharynx's tumour. The histopathological examination showed the presence of an adipose tissue and salivary gland framework. In a laryngological examination, the presence of a tumour on the right side of the neck extending from the auricle to submandibular region was noted. The skin around was painless and well warmed. A tracheostomy was present in the midline. In the imaging exploration performed, the presence of a vast pathological mass with smooth outlines measuring 44 × 77 × 95 mm and predominant adiposo-calcareous component was found (Figure 1).

Based on the medical history, the physical examination and the additional imaging studies performed, the patient was deemed to require a surgical treatment. Extended biopsy was taken into consideration, however, due to the prolonged course of the condition and the stagnant growth of the tumour, as well as failure to come to an unequivocal conclusion after the previously taken biopsy, it was decided the whole mass would be removed and examined. Therefore an incision was made under general anesthesia through the oral cavity approach in the midline, on the posterior wall of the

Table 1. Hamartoma's location described in the literature.¹⁰

Location	N(%)	Authors
Nosopharynx	6(31.6)	Foxwell and Kelham (1958), Bicknell (1967), Birt and Joes (1969), Baille and Batsakis (1974), Majumder et al (1977)
Nasal cavity	4(21.0)	Kacker and Das Gupta (1973), Mukhberji et al. (1980), Dandapth et al. (1985), Suvolia and Agarwal (1987)
Tongue	3(15.8)	Stamm i Tauber (1945), Perri (1956), Ishil et al. (1968)
Larynx	1(5.3)	Clinic et al. (1964)
Middle ear	1(5.3)	Eichel and Halberg (1966)
Tonsil	1(5.3)	Mukhberji et al. (1976)
Face	1(5.3)	Puri et al. (1977)
Hypopharynx	1(5.3)	Subhanna (1986)
Posterior wall of the throat	1(5.3)	The case under report



Figure 1. CT and MR coronal view showing heterogeneous mass of the tumour.

Table 2. The origin of most common parapharyngeal space tumours.¹¹

Study	Number of patients <i>n</i>	Salivary glands		Neurogenic <i>n</i> (%)	Other <i>n</i> (%)
		Benign, <i>n</i> (%)	Malignant, <i>n</i> (%)		
McIlrath i Remine (1966)	101	42(42)	1(1)	28(28)	30(30)
Work and Hybels (1974)	40	14(35)	5(13)	13(33)	7(20)
Maran et al. (1974)	66	17(26)	36(55)	5(7)	8(12)
Som et al. (1984)	97	28(30)	12(12)	28(29)	28(29)
Shoss et al. (1985)	42	10(24)	6(14)	17(40)	9(21)
Carrau and Myers (1993)	77	25(32)	2(3)	36(47)	14(18)
Olsen (1994)	44	11(25)	5(11)	17(39)	1(2)

middle pharynx. The incision line was extended upwards, 3 cm above the hard palate level. An extraction of the soft palate tissue was performed. The tumour was dissected from the parapharyngeal space with simultaneous coagulation of the surrounding minor blood vessels. A neoplasm as a tissue conglomerate of about 10 cm in diameter was located beside the skull base, filling the parapharyngeal space, coming to the prevertebral fascia and spreading to the hyoid bone. However, on account of the possibility of complications, a resection of the second part of the tumour was not performed. The abscission of the affected tissue on the floor of the oral cavity and submandibular region was postponed. Moreover, during the surgery the tracheotomy tube was removed. A histopathological examination of the tumour obtained determined it to have been a neoplasm of developmental disruption nature (hamartoma type), built of cartilage, bone and bone marrow components. Benign hyperplasia of cartilage was observed focally.

The patient stayed in the ENT department for 7 days, in a satisfying condition. After the surgery the patient reported a substantial improvement of breathing, pronunciation and hearing in the right ear, which confirmed that the ultimate cause had to have been an auditory tube orifice obturation. The swallowing and mastication acts were improved as well.

In the following stage, 8 months after the first surgery, the remainder of the tumour was resected from the submandibular region.

4. RESULTS AND DISCUSSION

The parapharyngeal space is divided into the prestyloid and the retrostyloid compartment. The prestyloid compartment contains the pterygoid plexus and the deep lobe of the parotid gland, whereas the retrostyloid compartment contains the internal carotid artery, the mandibular nerve that emerges from the foramen ovale, as well as cranial nerves IX, X and XI. The hypoglossal nerve runs medially to listed structures. When planning a surgery within the oropharynx altered by ongoing disease, the proximity of the internal carotid artery and possibility of severe intraoperative bleeding should be noted (Table 2).

In the presented case study, the tumour occupied the naso- and oropharynx from the skull base to the hyoid bone. The diagnosis of tumours of this location is usually made in advanced stages, due to absence of specific symptoms. Patients with nasal obstruction are usually treated for chronic rhinitis. Another reason for the late diagnosis is that the nasopharynx-

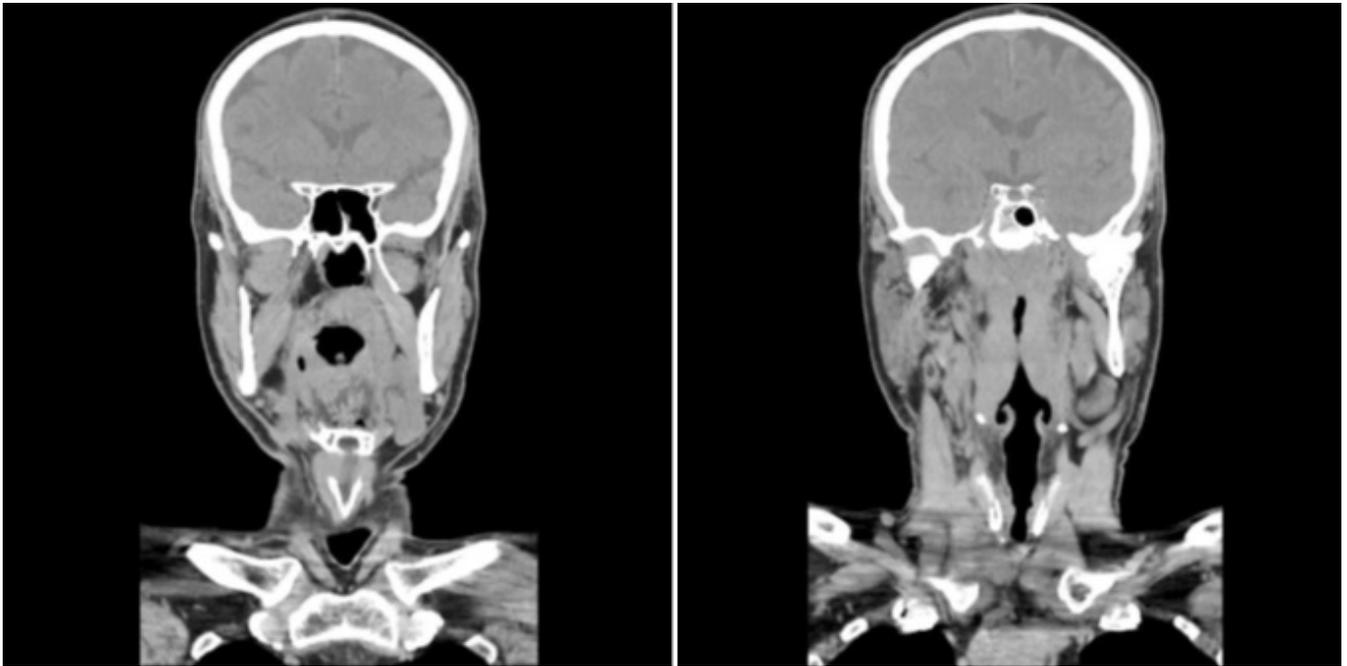


Figure 2. CT scan of the head and neck 10 months after the surgery.

geal tumours often present endophytic growth pattern, which means that macroscopic changes which can be spotted with endoscopy, appear in advanced phase of disease.

Tumours of parapharyngeal space are responsible for 1% of head and neck tumours;² 80% of these tumours are benign lesions, most often originating from the parotid gland (50% of all the cases). Based on the literature, neurogenic tumours account for 29.4% of the cases, lymphomas for 8.3%, inflammatory tumours (as abscess, lymphadenitis) for 4.6% and metastatic tumours for 2.3% .

According to the Olsen metastatic tumours classification,¹² they may be originating from surrounding structures, such as mandible, maxilla, naso- or oropharynx, palatine tonsils, oral cavity or temporal bone, as well as those spreading from vessels and nerves of head and neck region.

Parapharyngeal space might also be a place of distant metastases from thyroid carcinoma, osteosarcoma or adenocarcinoma.

The diagnosis of hamartoma tumour is based on histopathology. The differential diagnosis should include other types of the parapharyngeal space's tumours, e.g. pleomorphic adenoma, which is the most common benign salivary gland neoplasm. The treatment of choice is surgical resection. In order to choose an optimal surgical approach, the tumour's histopathology, size and location should be taken into consideration, as well as its relation to main vessels and nerves of the head and neck region. The other important factor is a surgeon's experience.

Available surgical options are: intraoral, transcervical, submandibular-transcervical or transmandibular approach.^{9,13} In the presented case, the choice of the surgical technique was implied by the lack of histopathological diagnosis. Due to a high probability of benign lesion, mass within the pharynx was qualified for an extended biopsy.

The rest of the tumour located in the submandibular space was postponed for a subsequent surgery.

Surgical treatment from the intraoral approach is a preferred method of treatment for prestyloid compartment tumours. It is chosen particularly for neoplasms: located in the oropharynx, the palatine arches space or the posterior part of the pharynx, under the pharyngeal mucosa, or well demarcated. It is important that the tumour mass should not penetrate the retrostyloid compartment. Due to difficulties in extracting the tumour with a suitable margin, an intraoral approach is recommended for minor volume lesion.

Transcervical submandibular approach is reserved for the tumours of the pre- and retrostyloid compartment. Its doubtless advantage is the possibility of the whole tumour mass exposition within the inferior pharynx. Moreover, having performed a stylomandibular ligament intersection and a mandible forward translocation, one may be able to display the course of the great blood vessels of the area, especially the internal carotid artery along with the branches of the external carotid artery. Such an extension of the operating field is particularly helpful during resections of benign prestyloid tumours of the minor salivary glands.

Transcervical approach is most often used for prestyloid tumour's resection. However, it does not visualize the content of submandibular triangle.

Transmandibular resection of parapharyngeal tumours requires obtaining access to the skull base in order to remove the pathological mass as well as the exposure of vessels of the neck. Tumours that infiltrate the mandible require an extended surgical approach and possibly a mandibulectomy

Removing the pathological masses from the infratemporal fossa is preserved only for the malignant tumours, infiltrating the skull base and penetrating the cranial cavity.

In the presented case, due to the tumour location and a benign disease course, the oral cavity approach was chosen. It was also decided to undertake the approach because of stagnant tumour growth, lack of an infiltration, no enlarged lymph nodes exhibited and lack of histopathological diagnosis. The executed incision in the midline one the posterior and lateral pharyngeal wall allowed avoiding the cut on the skin of the face.

Intraoral and transcervical approaches are most commonly chosen for the resection of benign lesions. They are associated with a minor amount of complications. In the presented case the incision on the posterior part of the wall with access to the right palatopharyngeal arch was limited by the anatomical conditions related to the topography of the area. It is associated with the proximity of the internal carotid artery, which arch or loop may occur in the operating field. Therefore the surgery was undertaken with great precaution, with the help of a zoom microscope magnification.

It should be noted that another possible complication of the following surgery is the temporary dysfunction of the pharyngeal muscles. Moreover, the soft palate resection may cause velopharyngeal insufficiency, which is a disability to close the velopharyngeal space.¹³ In the presented case, the follow-up examination 10 months after the surgery showed preserved function of the palate. The patient's symptoms disappeared (Figure 2).

5. CONCLUSIONS

The tumours of the parapharyngeal space constitute from 0.5% to 1% of all head and neck tumours, 80% of which are benign neoplasm, most often (50%) stemming from the parotid gland. The treatment of choice for benign lesions like the hamartoma is a surgical resection. It is important to choose a suitable surgical approach considering morphotic parameters of a tumour, anatomic and topographic conditions. The possibility of complications during the surgery, e.g. the probability of the great vessels damage, and after the surgery, e.g. rhinolalia and velopalatal insufficiency, should also be taken into account.

Conflict of interest

None.

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