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# Diagnostic peculiarities and difficulties of parapharyngeal space tumours

Prieryklinio tarpo navikų diagnostikos ypatybės ir sunkumai

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## Introduction

Parapharyngeal space tumours, most of them benign, account for some 0.5% of head and neck. Since they are rare, most clinicians will only come across a small number of them during their career. Nevertheless, it is important to highlight a few aspects of their diagnostics: symptoms are various and usually nonspecific, and signs manifest when tumour is rather advanced.

## Materials and methods

This article summarizes diagnostic aspects of retrospective analysis of 6 parapharyngeal space tumour cases, treated in National Cancer Institute between 2004 and 2016. We revise the scientific literature and analyse the diagnostic sequence, placing special emphasis on describing difficulties and peculiarities of diagnosing parapharyngeal space tumours.

#### Results

3 benign and 3 malignant tumours were diagnosed. Patients reported a wide variety of symptoms, most of them being nonspecific. The vast majority of patients underwent preoperative fine needle aspiration biopsy (FNAB), and radiological imaging (computed tomography (CT) and/or magnetic resonance imaging (MRI) was performed for all of them.

## Conclusions

All clinicians who come across these cases should take into consideration that symptoms of parapharyngeal space tumours are usually nonspecific, and signs appear after a period of silence. In diagnostic sequence, fine needle aspiration biopsy is mandatory to avoid histological surprises. Radiological imaging, especially MRI, is useful in predicting the origin, side, size and, to some extent, the type of the tumour.

Key words: parapharyngeal space, tumour, fine needle aspiration

## Įžanga

Prieryklinio tarpo navikai, dažniausiai nepiktybiniai, sudaro apie 0,5 % galvos ir kaklo srities navikų. Jie yra reti, todėl daugelis gydytojų per savo praktiką su šia patologija susiduria nedažnai. Svarbu išskirti kelis prieryklinio tarpo navikų diagnostikos ypatumus: simptomai yra įvairūs ir nespecifiški, o objektyvūs požymiai pasireiškia, kai navikas yra vėlesnės stadijos.

#### Medžiaga ir metodai

Šiame straipsnyje apibendrinami retrospektyviosios šešių prieryklinio tarpo navikų atvejų analizės diagnostiniai aspektai. Visi aptariami pacientai buvo gydyti Nacionaliniame vėžio institute nuo 2004 iki 2016 metų. Straipsnyje apžvelgiame mokslinę literatūrą ir nagrinėjame diagnostikos seką, atkreipdami dėmesį į prieryklinio tarpo navikų diagnozavimo ypatumus bei sun-kumus.

#### Rezultatai

Buvo diagnozuoti trys gerybiniai ir trys piktybiniai navikai. Pacientai skundėsi įvairiais, daugiausia nespecifiniais, simptomais. Daugumai pacientų buvo atlikta priešoperacinė aspiracinė biopsija plona adata, visiems – priešoperacinis radiologinis tyrimas (kompiuterinė tomografija ir (ar) magnetinio rezonanso tomografija.

#### Išvados

Visi gydytojai, susiduriantys su šia patologija, turėtų atkreipti dėmesį, kad prieryklinio tarpo navikų simptomai dažniausiai būna nespecifiniai, o objektyvūs požymiai pasireiškia navikui jau pažengus. Kalbant apie diagnostikos seką, aspiracinė biopsija plona adata yra būtinas tyrimas, siekiant išvengti histologinių netikėtumų. Radiologinis tyrimas, ypač magnetinio rezonanso tomografija, yra naudingas vertinant naviko kilmę, dydį, iš dalies ir tipą.

Reikšminiai žodžiai: prieryklinis tarpas, navikas, aspiracinė biopsija plona adata

# Introduction

The parapharyngeal space is difficult to recognize among numerous important anatomical structures in the neck. However, it is a well-defined space located on both sides of the neck, deep to the mandibular ramus, the pterygoid muscles and the parotid gland in the upper neck immediately lateral to the pharynx [1].

Parapharyngeal space is described as an inverted pyramid with the base formed by the skull base and the apex pointing to the greater cornu of the hyoid bone [2].

The fascia running posteriorly from the styloid process to the tensor veli palatini muscle divides the parapharyngeal space into two compartments. The prestyloid (anterior) compartment is filled with fat, connective tissue and part of the retromandibular portion of the deep lobe of the parotid gland, while the poststyloid (posterior) compartment contains the internal carotid artery, internal jugular vein, cranial nerves from IX to XII, sympathetic chain, and lymph nodes [1, 3].

Tumours may arise from each of mentioned structures and any clinician, who encounters parapharyngeal space lesions, should give thought to this pathology.

The purpose of this report is to highlight diagnostic peculiarities and difficulties of parapharyngeal space tumours and to describe our experience in process of formulating diagnoses.

# Patients and methods

The study design was a retrospective analysis of 6 patients with benign or malignant tumours who underwent

surgical resection between 2004 and 2016 in National Cancer Institute at the Department of Otolaryngology, Head and Neck Surgery and Oncology. Patients were referred from different health care institutions, consulted in our outpatient clinic and referred for surgical treatment. Two surgeons performed all operations. Two patients underwent postoperative radiotherapy. Patients were followed up at the outpatient clinic.

# Results

Between 2004 and 2016, 6 patients with benign or malignant masses of parapharyngeal space underwent surgical resection in our institution. There were 4 males and 2 females with the average age of 51.5 (range 36-65). Before treatment patients complained with the following: feeling of foreign body in the pharynx when swallowing (2), headache (1), throaty voice in the mornings, which used to become normal in a few hours (1), lasting coryza and stuffy nose (1), tumorous formation on the wall of pharynx (3) and palate (1), intense pain of pharynx especially during the night time (1), loss of weight (1), general weakness (1), difficulties in breathing through nose and mouth (1). Complaints had the duration of 3 weeks to 2 years. One patient had been complaining with resent symptoms, which appeared due to benign tumour recurrence, for 2-3 months; however, primary diagnosis had been established 12 years ago. During preoperative physical examination, various signs were detected. In one case only, a lump on the right submandibular region of 2 cm in diameter was observed

extraoraly, while intraoral deformation reached up to 9×5×7 cm in size. In all cases, deformation of pharyngeal area (nasopharynx, palatopharyngeal and palatoglossal arch, pharyngeal wall, palate midline displacement, expulsion of tonsil) was observed. In one case, enlarged lymph node was also palpated in the left parotid area (malignant tumour diagnosis). All patients preoperatively underwent radiological examination, usually MRI (for two patients both MRI and CT were performed and one underwent only CT). Well defined tumour with clear, smooth borders was detected (Figure 1, 2). Majority of patients underwent fine needle aspiration biopsy and no open biopsies were performed. In two cases, preoperative diagnoses were consistent with final diagnoses (pleopmorphic adenoma). In one case, preoperatively malignant tumour was suspected, but postoperatively pleomorphic adenoma was diagnosed as well. Another patient was treated from "pharynx inflammation" until final diagnosis of metastatic oropharynx carcinoma was made at our institution after histological examination of resected parapharyngeal tumour. In case No. 6, preoperative fine needle aspiration biopsy was not performed but diagnosis of myoepithelioma was established after postoperative histological examinations following previous surgical treatments. After latest palliative resection, inversion of myoepithelioma to myoepithelic carcinoma

was determined. Neck ultrasound evaluation was performed for two patients, but this examination was not informative. All patients were treated surgically those with malignancy received postoperative radiotherapy. Clinical data about presented cases is summarized in Table 1.

## Discussion

Wide variety of structures causes diversity of histopathologies in parapharyngeal space, and poststyloid tumours can arise from each of these structures. Meanwhile, prestyloid tumours mostly develop from salivary gland. It is useful to determine whether tumour is in either pre- or poststyloid compartment, as knowledge of this is helpful in determining possible diagnosis (Table 2) [4].

The anatomical relationships and histological diversity of parapharyngeal space tumours present difficult diagnostic and management challenges.

Parapharyngeal space tumours account for 0.5% of all head and neck masses but they are common enough to warrant a high index of suspicion [5].

Most of parapharyngeal space tumours are benign (70–80%), and 20–30% are malignant [6, 7].

The biggest part (40–50%) of parapharyngeal masses is composed of salivary gland tumours, the second in



**Figure 1.** CT, axial view. Well defined tumour with clear, smooth borders, dislocating adjacent structures (wall of pharynx), in the right parapharyngeal space



**Figure 2.** CT, sagittal view. Dislocation of adjacent structures (wall of pharynx). Tumour bordered with thin fat density boundaries

Table 1. Clinical summary of patients in the study

Patient No.	Sex	Age (years)	Symptoms	Duration of symptoms	Physical exa- mination	Preoperative FNAB / final diagnosis	Preoperative radiological examination MRU/CT
1	М	36	Dysphagia	3 weeks	Lump of 2 cm in diameter at the submandibular region, de- formation of palatopharyngeal arch, midline of the soft palate pushed to the left, right tonsila expulsed by lump 9×5×7 cm in size	Pleomorphic adenoma / pleomorphic adenoma	Well-defined pre- styloid tumour, 4,5×4,5×7 cm in size
2	F	44	Headache, tumorous formation	2 years	Tumorous formation in left pharyngeal wall 5 cm in diame- ter, medial displacement of soft palate and palatoglossal arch	Pleomorphic adenoma / pleomorphic adenoma	Formation in left parapharyngeal space 49×33×25 mm in size
3	М	54	Dysphonia, lasting coryza	_	Deformation of nasopharynx and palatopharyngeal arch on the left	Adenocarci- noma of low malignancy level / pleomor- phic adenoma	Tumorous forma- tion in left pa- rapharyngeal space 27×42×50 mm and possible metasta- ses in II neck zone 9 mm lymph node
4	М	64	Pain of phar- ynx, tumorous formation, gen- eral weakness, loss of weight	4 months	Tumorous formation in the left parapharyngeal area, behind fauces, extending to nasopha- rynx	Atypical squ- amous cells/ squam-ous cell carcinoma (mts)	Pathological forma- tion on the left side in nasopharyngeal – oropharyngeal level, 33×24 mm in size
5	М	46	Tumorous formation, dysphagia	6 months	Deformation due to tumour 7×5 cm in size observed intraorally in the right side of oropharynx	Pleomorphic adenoma with atypical cells / high malig- nancy ductal adenocarcinoma in pleomorphic adenoma of salivary gland	Tumour expanding to right oropharynx, narrowing pharynx, 55x×45 mm in size
6	F	65	Tumorous formation, dyspnea	Recent complaints for 2–3 months. Ill for 12 years	Painless soft and hard palate deformation on the left, extend- ing to nasopharynx. Enlarged lymph node in the left parotid area	Myoepithe- lioma / myoepi- thelic carcinoma	Tumor in palati- ne / parotid area 52×68 mm in size, enlarged regional lymph nodes

Table 2. Contents and primary lesions of the parapharyngeal space	[4	]
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	Prestyloid compartment	Poststyloid compartment
Contents	Deep lobe of parotid gland, minor salivary glands, lymph nodes and parapharyngeal fat	Carotid artery, jugular vein, cranial nerves IX, X, XI and XII, lymph nodes, cervical sympa- thetic chain and glomus tissue
Primary lesions of the parapharyngeal space	Salivary gland lesions, lymphomas and lipomas	Paragangliomas, nerve sheath tumours, lym- phomas, connective tissue

frequency are neurogenic tumours (17–25%), 10–15% of parapharyngeal lesions are paragangliomas [7]. Our experience coincides with data found in literature: majority of tumours originated from salivary gland tissue. In one of presented cases, final diagnosis of metastatic oropharynx carcinoma was made after histological examination of resected parapharyngeal tumour.

Most of benign tumours of salivary gland in the oral cavity present themselves as painless submucosal swelling. Those from the parapharyngeal space may also manifest additional symptoms, like otalgia, neuralgia, foreign body sensation in the throat, difficulty in swallowing, voice changes, trismus, otologic symptoms, obstruction of the Eustachian tube, 9th, 10th, or 11th cranial nerve deficits, and, in rare cases, obstructive sleep apnea [8, 9].

Sometimes such nonspecific complaints as ill-fitting dentures may be the first sign of benign prestyloid lesion [10].

In the cases we present, a wide variety of complaints was observed: feeling a foreign body in the pharynx when swallowing, headache, throaty voice in the mornings, lasting coryza and stuffy nose, intense pain, especially in the night time, general weakness, loss of weight, difficulties in breathing through nose and mouth, with feeling of tumorous formation as the most common complaint (66.6% of cases), which corresponds to data found in literature. Diversity of non-specific symptoms may mimic other diseases, challenge determination of correct diagnosis and prescription of appropriate treatment.

Parapharyngeal space is a difficult place for clinical examination as it is surrounded by the muscles of mastication, mandible, and parotid gland. Symptoms are usually noticed when the tumour is larger than 2.5-3 cm [9, 10].

Submucosal swelling in the lateral pharyngeal wall with or without extension to retromandibular fossa or the submandibular trigone is the classical finding of benign parapharyngeal swellings [12, 13, 14, 15].

However, in most cases, patients with parapharyngeal space tumours are present with a neck or oropharyngeal mass, which does not cause symptoms detectable on physical examination as only the inferior and medial boundaries of the parapharyngeal space are distensible [10]. Therefore, often extraoral examination does not reveal any pathological changes (as was the case in the majority of cases we present).

Tumours often grow to at least 3 cm in diameter before they are detected, so patients may have these masses for long periods of time before symptoms arise [15]. In such cases, cervical swelling and smooth submucosal mass that anteromedially displaces the lateral pharyngeal wall, tonsil, and soft palate, revealed by intraoral physical examination, are recognized by chance [2, 7, 16]. Often, previously mentioned masses are misdiagnosed for an infection or a tonsil tumour [2, 16]. In one case we present, before admitting to our institute, parapharyngeal tumour was misdiagnosed and treated as paratonsilar abscess by incision. In our experience, all cases were presented with intraoral deformation due to tumorous masses, mostly more than 5 cm in diameter. Precise extraoral and intraoral examination is important in predicting the size of tumour; however, radiological imaging is essential for determining exact extent of masses, as well as their localisation.

Fine needle aspiration cytology is one of options for obtaining biopsy sample for diagnosis [17]. Literature shows that peroral or transcervical fine needle aspiration, performed under CT or ultrasound-guidance, is accurate in 90–95% of cases [16]. In parapharyngeal space tumours, 25–60% of intraoral fine needle aspirations may not be informative because of lack of adequate cellular material, excessive bleeding or other technical problems. The positive predictive value to identify benign tumours is 90% when performing fine needle aspiration under CT or ultrasound-guidance, but drops to 75% for malignant ones [18, 19].

Our experience shows that fine needle aspiration is sufficient as a method for determining the nature of tumour. Core needle biopsy is also a choice of modality. However, literature shows that fine needle aspiration is more advantageous when compared to core needle biopsy is terms of potentially increased risk of tumour spillage, capsule rupture and tumour recurrence, which ultrasound-guided core needle biopsy is associated with (depending on needle diameter and sample volume) [20]. In our opinion, open biopsy is not necessary. Other authors claim that open biopsy is not advisable as it increases the risk of bleeding, breakage of the capsule and, accordingly, the seeding of the lesion [21].



**Figure 3.** MRI, coronal view. Well defined tumour in the left parapharyngeal space, no relationship with parotid gland

Imaging studies (CT scan and MRI) are an important diagnostic means used to predict the origin, side, size and, to some extent, the type of parapharyngeal tumours. MRI with gadolinium has been shown to be superior to CT scan, and may be used in the investigation of parapharyngeal space tumours (Figure 3). It enables to reliably distinguish various structures: deep lobe parotid tumour, tumour of neurovascular origins, and minor salivary glands. Fatty layer between structures in T2 regimen is important diagnostically: if it is visible between the tumour and the parotid deep lobe, it shows that the tumour had separated from the lobe (Figure 4). Contrariwise, absence of this plane indicates that the tumour originated in the parotid deep lobe or, less frequently, was invading it [16, 22, 23, 24].

Contrast enhancement is characteristic of vascular and neurogenic tumours. Angiography is also useful for

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Figure 4. MRI, axial view. Well defined, heterointensive with cystic component tumour, deformating pharynx from the left

all enhancing lesions or vascularised masses, especially in relationship with carotid bifurcation [16].

This examination was not performed in any of presented cases as none of the mentioned lesions was suspected.

# Conclusion

Tumours of the parapharyngeal space are rare. Most cases are treatable, and high level of alertness is needed to diagnose them correctly. All clinicians who come across these cases should take into consideration that symptoms of parapharyngeal space tumours are usually nonspecific, and signs appear after a period of silence. In diagnostic sequence, fine needle aspiration biopsy is proven to be efficient and safe. Radiological imaging, especially MRI, is useful in predicting the origin, side, size and, to some extent, the type of the tumour.

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