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Diagnosis and treatment of Schwannoma of the parapharyngeal space on the example of a 14-year-old male patient

Diagnostyka i leczenie nerwiaka przestrzeni przygardłowej na przykładzie przypadku 14-letniego pacjenta

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Keywords

parapharyngeal space, Schwannoma, Horner's syndrome, transoral approach

Summary

The parapharyngeal space is located on the side of the pharynx wall. It is an inverted pyramid-shaped anatomical area filled with adipose and connective tissue. Schwannoma (neurinoma) is a benign tumour that originates from the Schwann cells that form the myelin sheaths of nerves. This neoplasm in the parapharyngeal space usually arises from the vagus nerve and the sympathetic trunk. Parapharyngeal space tumours are often a complicated diagnostic and therapeutic problem. A 14-year-old patient was admitted to the hospital because of a feeling of discomfort in the throat due to a nodular lesion on the right side. The lesion was removed through transoral approach. The patient remains under laryngological and oncological care. In follow-up examinations 6 months and 1 year after surgery, no features of recurrence were revealed. Due to their rare occurrence, non-specific symptoms, and anatomical topography, the diagnostic and therapeutic process of parapharyngeal space tumours is very difficult.

słowa kluczowe

przestrzeń przygardłowa, Schwannoma, zespół Hornera, dostęp przezustny

Streszczenie

Przestrzeń przygardłowa jest zlokalizowana na bocznej ścianie gardła. Jest to przestrzeń o kształcie odwróconej piramidy, wypełniona tkanką tłuszczową i łączną. Schwannoma (nerwiak) jest łagodnym guzem zbudowanym z komórek Schwanna, które tworzą osłonkę mielinową nerwów. Nowotwór ten w przestrzeni przygardłowej najczęściej powstaje z nerwu błędnego i pnia współczulnego. Guzy przestrzeni przygardłowej są często wyzwaniem zarówno diagnostycznym, jak i leczniczym. Czternastoletni pacjent został przyjęty do szpitala z powodu uczucia dyskomfortu w gardle związanego z obecnością guzowej zmiany po stronie prawej. Zmiana została usunięta poprzez dostęp przezustny. Pacjent pozostaje pod opieką laryngologiczną i onkologiczną. W obserwacji 6- i 12-miesięcznej nie stwierdzono cech wznowy guza. Ze względu na rzadkie występowanie, niespecyficzne objawy oraz topografię anatomiczną proces diagnozowania i leczenia guzów przestrzeni przygadłowej jest bardzo trudny.

Introduction

The parapharyngeal space is located on the side of the pharynx wall. It is an inverted pyramid-shaped anatomical area filled with adipose and connective tissue. There are important anatomical structures such as the internal carotid artery, the internal jugular vein, the cranial nerves (IX, X, XI, XII), and lymph nodes between them. The upper part of this area adjoins the base of the skull, and its apex, located lower, reaches the hyoid bone (1-3). This space is also inaccessible during palpation and ultrasound examinations, which impedes the diagnosis (3).

Parapharyngeal space tumours account for only 0.5% of all diagnosed head and neck tumours, and only 30% of them are neurogenic tumours. Among them, 80% are benign, and only 20% are malignant. Most of the tumours in this location originate from the parotid gland – around 50%. Primary lymph node tumours (8%), inflammatory (4%), and metastatic (2%) growths are also diagnosed (1-3). Schwannomas of the parapharyngeal space usually occur in patients between the ages of 30 and 70 years (1). In the described case, we are dealing with a pediatric patient, which makes his case particularly rare.

Schwannoma (neurinoma) is a benign tumour that originates from the Schwann cells that form the myelin sheaths of nerves. This neoplasm usually arises from the peripheral nerves and in the parapharyngeal space – from the vagus nerve and the sympathetic trunk. Microscopically, Schwannoma is a homogenous, well-differentiated, well-limited, round, and pearly white nodular lesion and has two histological types: Antoni A and Antoni B. It ordinarily reaches a diameter of less than 5 cm (1, 4, 5).

Symptoms of parapharyngeal space tumours are not specific and late-onset. These neoplasms grow slowly and asymptomatically (1, 3). Due to their location, they can cause throat discomfort resulting from foreign body retention, dyspnoea (breathing problems), slurred speech, dysphagia (swallowing impairment), asymmetry of the palatal arches, and damage or displacement of the surrounding anatomical structures (ex. nerves, cervical vessels, tonsils, or lymph nodes). The tumours growing towards the nasopharynx can cause nasal congestion, change of the voice timbre, Eustachian tube obstruction, and secondarily conductive hearing loss or secretory otitis media (2, 4). Patients with malignant tumours are more likely to present with pain, otalgia, trismus, or cranial nerve deficits. The presence of Horner's syndrome before surgery in cases of patients with parapharyngeal space Schwannomas is very rare (1, 6).

Parapharyngeal space tumours are often a complicated diagnostic and therapeutic problem. It is necessary to perform radiological examinations in the diagnostic process: computed tomography (CT) with contrast and magnetic resonance imaging (MRI). They help to visualize and assess the tumour – establish its size and location, determine the tumour's relation to adjacent anatomical structures, or forecast its histopathological type. Particularly, MRI plays a big role in the diagnosis because it allows a good evaluation of soft tissues and does not expose the patient to ionizing radiation. Currently, MRI is the preferred imaging method used in the diagnosis of parapharyngeal space tumours. Furthermore, when vascular etiology is suspected, carotid arteriography should be performed (1-4). There is a controversy about performing a parapharyngeal space tumour biopsy. Naturally, the histopathological examination has great treatment importance – it provides information about the structure, origin, and malignancy of the tumour. However, a needle biopsy is difficult to conduct due to the lesion location – it may result in numerous non-diagnostic cytological findings and can expose the patient to vascular complications (3, 7).

Surgery intervention is the main treatment method for parapharyngeal space tumours. The most common are the transcervical or parotid-cervical approaches – the approach choice depends on information obtained preoperatively. Other possibilities are transoral and transmandibular approaches (3, 8). Depending on the size and advancement of the tumour, the operation may be extended to the removal of the submandibular gland and the mandibulectomy. Because of the neurogenic origin and close location of the large blood vessels, the surgical resection of parapharyngeal space tumours is burdened with a high risk of neurological complications (ex. nerve IX-XII paralysis, central nervous system ischemia) and perioperative hemorrhage (from major cervical vessels) or infection. Additionally, incomplete removal of the lesion can cause a recurrence of the disease. The resection should involve both the tumour and a rim of normal tissue. There are a few described cases in which the function of the affected nerve was preserved after surgery, so the patient should be prepared for the postoperative occurrence of nervous dysfunction (1, 2, 4). If the neoplasm proves to be malignant, oncological treatment may also be included (ex. radiotherapy). Inoperable tumours are qualified for simultaneous radiochemotherapy or palliative radiotherapy (2). However, these cancers are characterized by high cellular variability so oncological therapy might be ineffective (1, 4).

Case report

A fourteen-year-old patient was admitted to the hospital in October 2021 because of a feeling of discomfort in the throat due to a nodular lesion on the right side. In the physical examination neck asymmetry, distortion of the right palatal arch, and enlargement of the right palatine tonsil were described. A tumour biopsy was performed under general anesthesia and later analyzed in a histopathological examination. According to the surgical report, several pieces of the tumour were removed, along with the accompanying lymph node. Perhaps for this reason, the first histopathological examination showed that the specimen was in fact, only a part of an enlarged lymph node. Another incorrect diagnostic trail was introduced by the ultrasound

examination, after which a lymphoma lesion was suspected. Computed tomography of the head with contrast showed a well-defined tumour measuring 35 x 20 x 50 mm, extending from the base of the skull to the level of the hyoid bone, modelling the shape of the posterior wall of the pharynx on the right side, and dislocating surrounding structures – the internal carotid artery, the right medial pterygoid muscle, and the styloid process. An oncological consultation was also conducted. The patient was discharged home with the diagnosis of acute lymphadenitis (fig. 1, 2a, b).

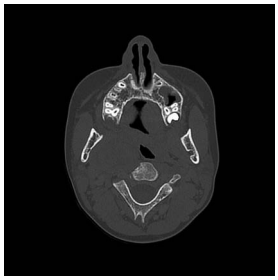

Fig. 1. Tumour on CT scan without contrast

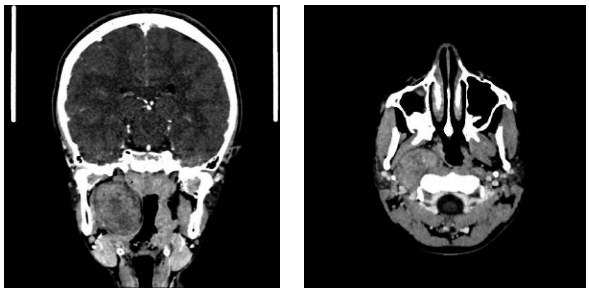

Fig. 2a, b. Tumour on CT scan with contrast

After a few weeks, corrections in the result of the histopathological examination were made – the new diagnosis was Schwannoma, type Antoni A. The patient was admitted to the hospital in December 2021 for an MRI. It determined that the size of the tumour had not changed since the previous hospitalization, and it was suspected that the tumour might have been related to the nerve XII or sympathetic trunk. The patient was discharged home pending resection surgery (fig. 3a-c).

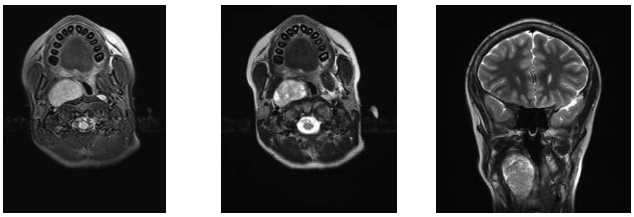

Fig. 3a-c. Tumour on MRI scan

The third hospitalization took place the following week. There were performed additional neurological and ophthalmological consultations, which revealed the symptoms of Horner's syndrome probably caused by the damage to the nerve transmission by the tumour: anisocoria (the enlargement of the left pupil comparing to the right one), a drooping eyelid, and asymmetry of the eyeballs. Dysphagia and slurred speech have also been observed. The lesion was successfully removed using a microscope through transoral approach. After the procedure, most of the symptoms disappeared. During the genetic consultation, chemotherapy was not recommended, but the need for further examinations regarding neurofibromatosis type 2 was advised.

The patient remains under laryngological and oncological care. In follow-up examinations 6 months and 1 year after surgery, no features of recurrence were revealed (fig. 4a, b, 5a, b).

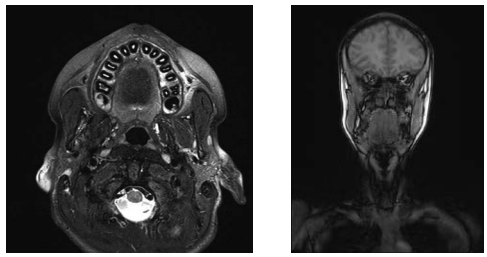

Fig. 4a, b. MRI scans in 6-months follow-up

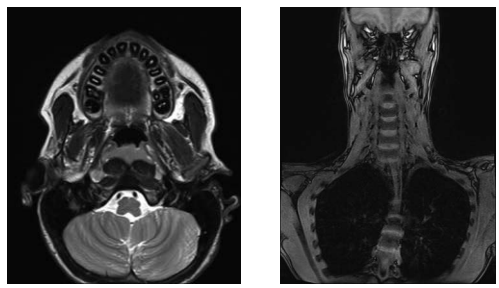

Fig. 5a, b. MRI scans in 1-year follow-up

Discussion

Parapharyngeal tumours are very rare. Schwannoma, described in our patient, is a neurogenic tumour, as 30% of the head and neck tumours, and benign, as 80% neurogenic tumours (1), which makes the described patient a unique case.

The diagnosis of neoplasms located in the parapharyngeal space is particularly difficult since neither otolaryngological examination nor ultrasound investigates this profound area completely.

Computed tomography (CT) allows for the assessment of the location, size of the tumour and possible infiltration of adjacent tissues, such as the larynx and structures of the neck or chest. CT also allows visualization of possible metastases in the lymph nodes. In addition, thanks to the contrast, it is possible to visualize the characteristics of the

tumour, such as areas of necrosis or vascular changes. Tumours in this location may be small, making them difficult to detect, they may have similar tissue density to surrounding structures, resulting in the lack of clear contrast on CT images. This makes it difficult to precisely define the tumour's boundaries, and distinguish it from normal tissue. The parapharyngeal space is also actively involved in breathing and swallowing. This may lead to motion artefacts on ultrasound and CT images, affecting image quality and impeding the assessment of the exact shape and size of the tumour (9-11).

MRI is particularly useful in the evaluation of parapharyngeal tumours due to its high resolution and ability to distinguish soft tissues. It enables accurate visualization of the tumour, assessment of the extent of infiltration of anatomical structures and assessment of metastases in lymph nodes (9-11).

Despite the development of imaging techniques in the diagnosis of the parapharyngeal space, various diagnostic difficulties are still encountered.

Ultrasound allows visualization of lymph nodes and assessment of their size, shape, echogenicity and possible presence of metastases. It is also helpful in monitoring the progress of therapy, e.g. by assessing changes in tumour size. Due to the above, for parapharyngeal tumours, a combination of different imaging techniques such as CT and/or MRI is often used to obtain a more comprehensive assessment of the tumour and its surroundings (12, 13).

Moreover, those tumours present a late onset of the symptoms. Due to their location, they can grow for some time before causing any symptoms. Therefore, they are often diagnosed at an advanced stage when the tumour grows larger or invades adjacent structures. When symptoms do appear, they might be non-specific and confusing. Parapharyngeal tumours may cause symptoms that are similar to other conditions of the throat or digestive tract, such as difficulty swallowing, chest pain, hoarseness, shortness of breath, or chronic cough. These symptoms may be underestimated or attributed to other causes, leading to a delay in diagnosis. Sometimes tumours of the parapharyngeal space have a neurological origin. Their symptoms may be difficult to interpret or may be confused with other neurological conditions (14).

Even though fine-needle biopsy and surgery may be challenging, they remain the main method of diagnosis and treatment. Parapharyngeal schwannomas are characterized by slow growth, low recurrence rates, and often non-invasive nature, thus not requiring immediate surgery and allowing for an observational approach. In the past, most neuromas of the parapharyngeal space were removed perorally, which was associated with the risk of incomplete excision of the tumour, haemorrhage, infection or damage to the surrounding nerves. Currently, transcervical access is the most preferred due to the possibility of recognizing and safely separating the facial nerve branches. On the contrary, transauricular access does not provide control of nerve structures and blood vessels, therefore it is used rather in cases of small, benign and non-vascularized tumours. It is also possible to perform resection with mandibulotomy, i.e. dissection of the mandible, which provides wider access to the operating field and radicalization of the operation. This is particularly useful in malignant, recurrent, large benign and highly vascularized tumours. If the tumour is completely removed, it is very unlikely that the disease will recur (1, 3). Patients after surgical treatment remain under medical supervision. In the 18 months follow-up of our patient, no recurrence was observed, which is coherent with other cases.

A complete surgical excision prevents the parapharyngeal tumour's recurrence and is a recommended treatment (15-18).

Performing biopsies in the diagnosis of parapharyngeal tumours is a controversial topic. Particularly in children, this procedure is very rarely used as it requires general anesthesia and there is uncertainty whether pathological cells can be obtained. This is particularly difficult in the case of a fine needle biopsy, which additionally cannot be controlled by ultrasound. A CT biopsy is possible but rarely used in practice. Excisional biopsy, which is associated with the risk of metastases and adhesions, is even more questionable. Its use for diagnosis is suggested for non-surgical malignancies, metastases and lymphomas (3).

Conclusions

Due to their rare occurrence, non-specific symptoms, and anatomical topography, the diagnostic and therapeutic process of parapharyngeal space tumours is very difficult. Many incorrect initial diagnoses appear there (3).

CT and MRI are the most useful examinations in the diagnosis and the choice of surgical resection procedure of parapharyngeal space tumours (3).

The surgical resection of parapharyngeal Schwannoma is still the basic method of treatment, although it is also a complicated surgery procedure with a high risk of complications such as postoperative neurologic deficits. It is possible to predict the nerve giving rise to a tumour with MRI or CT with considerable precision, and this information allows us to anticipate the function of the nerve that will be affected (1).

Fortunately, we could avoid several complications in the described case. The treatment applied to our patient improved his quality of life, but unfortunately, the symptoms of Horner's syndrome persisted.

Conflict of interest
Konflikt interesów

None
Brak konfliktu interesów

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