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A rare case of coexistence of two different neurogenic tumors in the parapharyngeal space

Rzadki przypadek współistnienia dwóch różnych nowotworów neurogennych w przestrzeni przygardłowej

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ABSTRACT: Tumors of the parapharyngeal space are rare and account for less than 1% of head and neck tumors. About 20% are neurogenic tumors, of which the most common are schwannomas and paragangliomas. The authors present a case of a 64-year-old patient, diagnosed with cervical spine disorders; he was incidentally diagnosed with a tumor in the parapharyngeal space reaching the internal carotid artery lumen. After the lesion was surgically removed, histopathological examination identified two different tumors of different origin; from the vagus nerve and the cervical portion of the sympathetic nervous system.

KEYWORDS: Schwannoma, ganglioneuroma, parapharyngeal space

STRESZCZENIE: Guzy przestrzeni przygardłowej występują rzadko i stanowią mniej niż 1% guzów głowy i szyi. W ok. 20% to guzy neurogenne, z czego najczęściej są to nerwiaki osłonkowe i paraganglioma. Autorzy prezentują przypadek 64-letniej chorej, u której w trakcie diagnostyki schorzeń kręgosłupa szyjnego przypadkowo stwierdzono obecność guza w przestrzeni przygardłowej, sięgającego otworu tętnicy szyjnej wewnętrznej. Po usunięciu chirurgicznym zmiany, badanie histopatologiczne zidentyfikowało dwa różne guzy o odmiennym pochodzeniu, z nerwu błędnego i części szyjnej układu współczulnego.

SŁOWA KLUCZOWE: Schwannoma, ganglioneuroma, przestrzeń przygardłowa

INTRODUCTION

Neurogenic tumors in the head and neck region are derived from peripheral nerves, autonomic ganglia, chromaffin cells or embryonic neuroektoderm. They can be benign or malignant. Schwannoma (Schwann cell tumor, neurolemmoma, neurinoma) and neuroblastoma originates from peripheral nerves, ganglioneuroma originates from autonomic ganglia, and paraganglioma (chemodectoma) from neuroectodermy-neurothekeoma [1]. Each of these benign tumors has its malignant counterpart: schwannoma malignum, ganglioneuroblastoma, paraganglioma malignum, primitive neuroectodermal tumor and olfactory neuroblastoma.

Tumors originating from peripheral nerves constitute more than 50% of neurogenic tumors of the head and neck, about 45% are schwannomas, 30% - paragangliomas [2]. Laryngopharyngeal tumors constitute less than 1% of tumors occurring in the head and neck, 80% of them are benign and the most common are schwannomas. About half of neuroblastomas originate from

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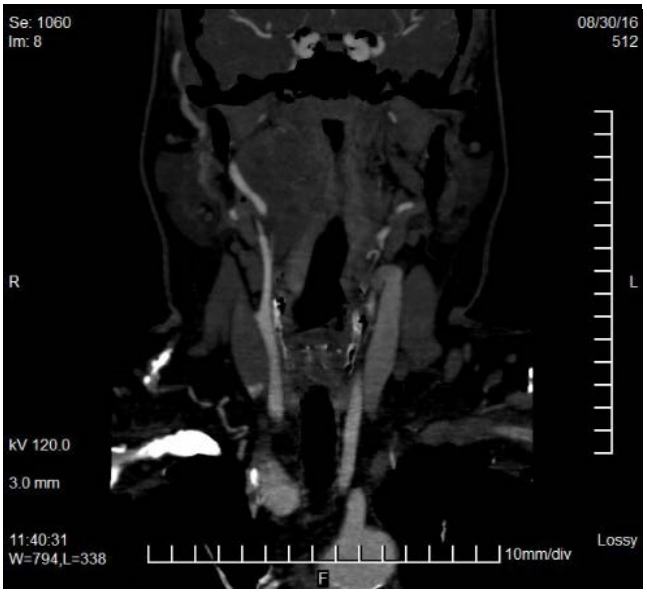

Fig. 1. Angio-CT scan.

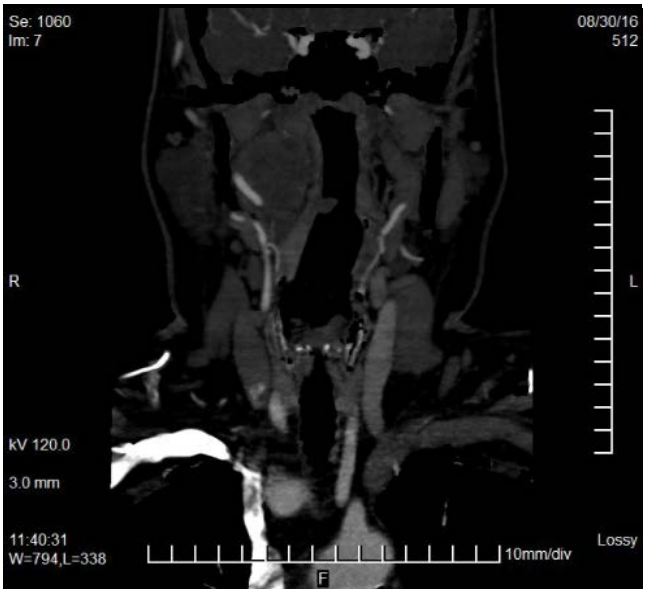

Fig. 2. Angio-CT scan.

the vagus nerve, and further from the cervical portion of the sympathetic trunk [3].

In the parapharyngeal space, the sympathetic trunk lies towards the back of the major neck vessels and the vagus nerve, and to the front from the longus capitis and colli. It is incorporated into the prevertebral plaque of the fascia of the neck and fastened by it, while the string vascular-nervous is movable forward relative to the fascia. The authors present a rare case of two histologically different, asymptomatic, neurogenic tumors in the parapharyngeal space originating from the cervical portion of the sympathetic trunk and the vagus nerve.

CASE REPORT

A 64-year-old female patient was referred from a neurological clinic to treat a tumor in the right parapharyngeal space, diagnosed during diagnostic radiology of the cervical spine. Six months earlier, she reported to a neurological outpatient setting with headaches and numbness in the upper limbs. Arterial hypertension and reduced mobility of the cervical spine were found. As far as the cranial nerves, feeling, muscle strength, movements and pupillary reactions to light are concerned, no abnormalities were found. Despite treatment and normal blood pressure numbers, the headaches persistent. Computer tomography (CT) of the head was performed, followed by angio-ct; an extensive nodular lesion's shift towards the right side of the internal carotid artery was found, extending from the base of the skull at the entry point of the carotid artery to the bony canal, in the temporal bone pyramid down to the nasopharynx level. The internal jugular vein was compressed and shifted towards the back. MRI of the cervical spine confirmed the presence of a tumor with low-intensity signals in T1 sequence and high intensity in T2. The radiologist suggested cavernous hemangioma or paraganglioma. In addition, the NMR study showed prominence of intervertebral disc to the spinal canal at C5-C6 and C6-C7 to a depth of approx. 3 mm,

At the time of admission to the Department of Laryngology and Laryngological Oncology, there were no lesions in the nose and nasopharynx, oropharynx, larynx and ears. In physical examination, the lymph nodes of the neck as well as the nodular lesion of the right thrombotic space were non-palpable. Symptoms of cranial nerve palsy were not observed. The patient did not report any disturbances in vision or hearing. She negated dysphagia.

The patient was qualified for surgery to remove the tumor in the parapharyngeal space. On the right side of the neck, the skin was cut along the anterior edge of the sternocleidomastoid muscle . After thorough assessment of anatomical conditions, the external carotid artery was ligated, the submandibular gland was removed to widen the operating field and provide better access to the lesion. Group III small lymph nodes on the right side were removed. Next, a tumorous 6-cm lesion lying on the fascia, between the vessels, along the vagus nerve was revealed, and further above, a satellite tumor of about 4 cm in size, lying between the internal carotid artery and the prevertebral fascia reaching the base of the skull. Both tumors surrounded by the capsule were removed completely and sent for histopathological examination.

After the procedure, the patient experienced signs of Horner syndrome. The following were observed: narrowing of the palpebral fissure and narrowing of the pupil of the right eye as well as collapse of the eyeball on the right side. General condition and proper local healing of the surgical wound allowed to send the patient home on the 4th day after the procedure.

The obtained result of histopathological examination of postoperative material describes what follows:

1. tumor - well-delineated, partially hyalinization, with hypocellular areas, focally hypercellular nonadenomatous tumor, without signs of pronounced cytological atypia. The image speaks for Schwannom partim cellulare S100 (+), SMA (-/+), v. Gibson (++), Ki 67<5%.

2. a satellite tumor preparation of approximately 4 cm- mature ganglion cell formation (ganglioneuroma).

3. lymph nodes with chronic inflammation.

The patient remains in constant neurological and ENT control. She does not report any problems.

DISCUSSION

Schwannoma is usually located intracranially (7% of primary intracranial tumors) within the vestibulocochlear and trigeminal nerve [4,5,6,7]. Extracranially, although these tumors may have many starting points, we rarely encounter them. They may occur in the nasal cavity and paranasal sinuses, in the floor of the mouth, tongue, throat, larynx or deep spaces of the neck [5, 8]. The case of neuroma within the masseter has been reported [9]. They are most often found in the parapharyngeal space, where the neuromas originate from the vagus nerve and the sympathetic trunk [3,5,8]. The incidence of neuromas is not affected by the patient's age and sex [5, 10].

These tumors are characterized by slow and asymptomatic growth that can, however, cause a mass effect, especially when it concerns limited spaces. Pain and nerve dysfunction occur late in the process of tumor development.

Macroscopically, tumors are surrounded by a capsule, well-circumscribed, solid, their mobility relative to surrounding tissue may be limited. Most often the tumor adheres to the nerve, rarely tumor cells penetrate between the nerve fibers.

On the basis of the macroscopic picture, neuromas can be divided into the following groups [4]:

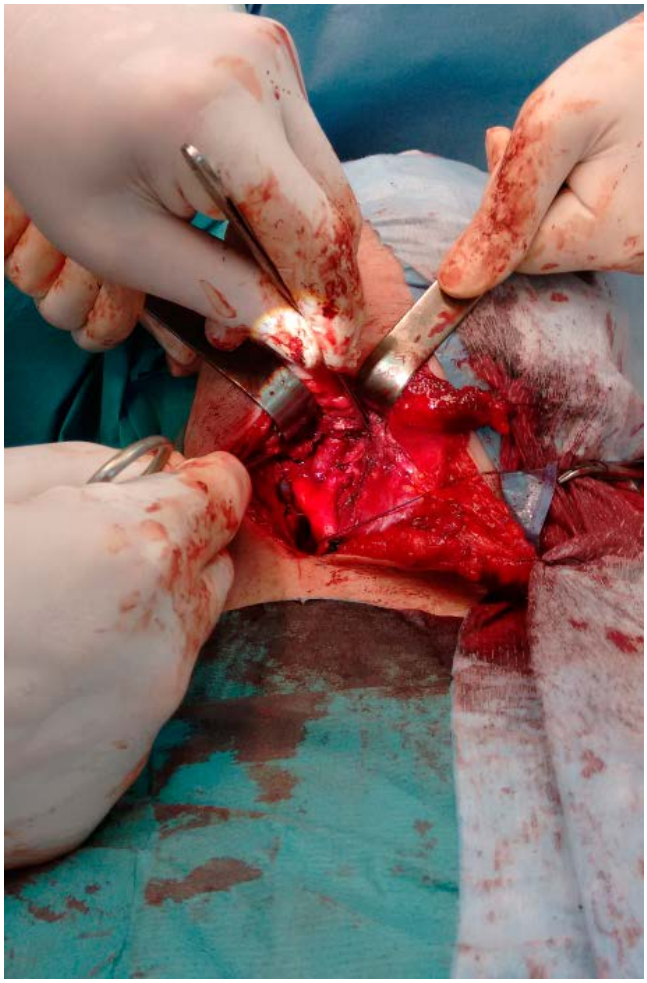

Fig. 3. Division of the right carotis communis artery with the tumor above.

Type I – determination of the tumor origin point is not possible

Type II – a preserved nerve is present in the tumor line

Type III – nerve fibers are delaminated on the surface of the tumor

Type IV – tumor coming out of nerve fibers

Microscopically, the tumor forms homogenous, well-differentiated cells with Schwann cell morphology. Hematoxylin staining distinguished two histological types of schwannomas. Type Antoni A and Antoni B. The histological type of tumor is determined by the dominant type of cellular texture. The site referred to as Antoni A forms clumped cells with a lined, spindle-like shape with palisade arranged cell nuclei. These cells are arranged in vortices separated by non-cellular zones. This cell system creates the so-called Verocay body. As for the Antoni B type, the cells present a loose, irregular arrangement in

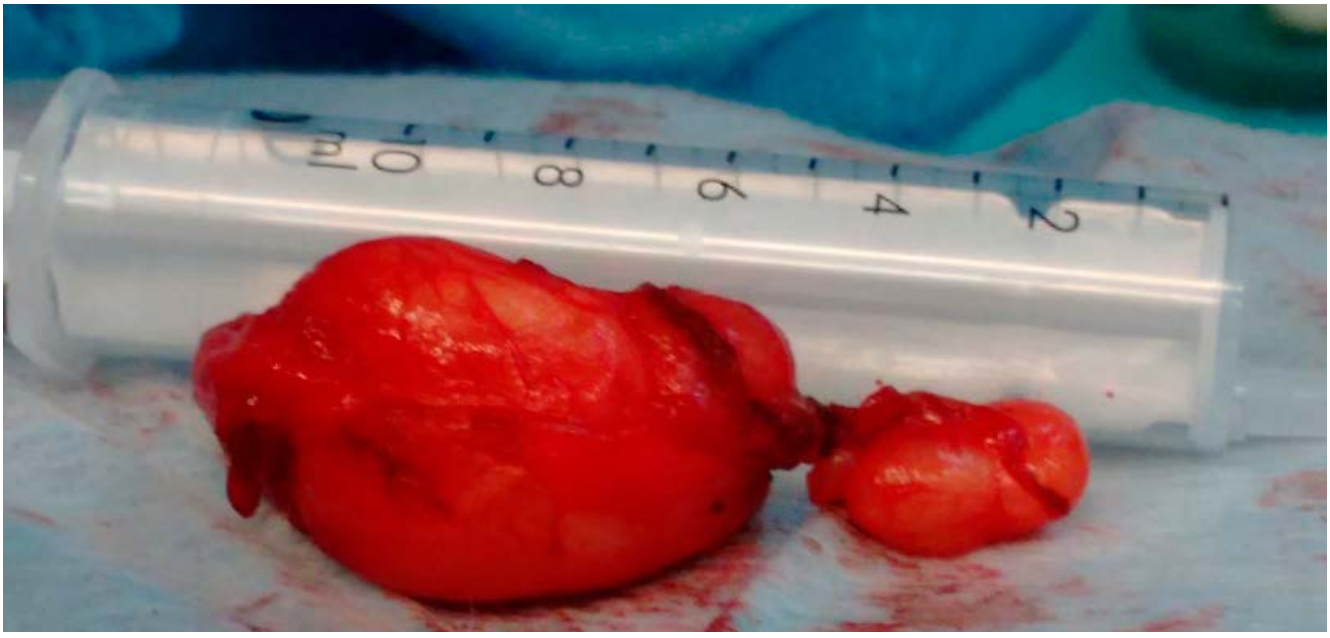

Fig. 4. Deleted tumors.

the connective tissue collagen-mucus stroma. [4,5,10,11,12]. In central tumors, the AA and AB components are represented in the same proportions, whereas in peripheral tumors of up to 5 cm in size, AA is dominant, whereas in the larger tumors AB [6] prevails. The basic immunohistochemical marker for neuroblastomas is S-100 protein and vimentin [6,9,10,12].

These tumors grow slowly and asymptomatically [5]. Most often, it causes discomfort due to nerve compression when the tumor reaches a significant size. MRI allows to establish the precise location of the lesion and determine the relation to adjacent anatomical structures in sites that are particularly difficult to reach [13]. In Bienia's research [2], fine needle aspiration was found to be 100% ineffective and is not the recommended diagnostic method.

Treatment is exclusively surgical, and the confirmation of diagnosis is histopathological examination and immunohistochemical examination [10, 12, 14]. Because these cancers are characterized by high cellular variability, treatment methods such as radiotherapy and chemotherapy are ineffective [3.5].

Surgical resection of tumors due to their neurogenic origin and close location of the large blood vessels of the head and neck is burdened with risk of both neurological complications – nerve paralysis or paresis, as well as perioperative risk of hemorrhage or central nervous system ischemia [2]. Resection should concern the tumor along with a rim of normal tissues [5]. It is usually possible to remove the whole lesion. Such a procedure allows for a complete cure, while non-complete removal of the lesion may lead to its recurrence or malignancy [4, 5, 10]. Tumor obliteration is observed in about 16% of cases [4, 11, 14].

As for tumors relevant to nerves of vital significance, in surgery, the tumor capsule through which nerve fibers may run should be preserved and the tumor should only be attempted to be extracted intracapsularly [2].

An example of two histologically different neurogenic tumors, developing asymptomatically, in a difficult to reach area, originating from various anatomical structures, is presented Imaging examinations made for diagnosis of another disease revealed lesions before the occurrence of complications and their removal was possible.

The encapsulated character of lesions and their complete excision made it possible to cure the patient; unfortunately, she experienced a complication secondary to damage of the sympathetic nervous system in the form of Horner's syndrome. The coexistence of schwannoma with ganglioneuroma is rarely mentioned in the available literature [15]. Therefore, the authors decided that the case which they have come across deserves to be presented.

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