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Hemangiopericytoma – a case report of a rare tumour of the parapharyngeal space.

Hemangiopericytoma – opis przypadku rzadkiego guza przestrzeni przygardłowej.

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ABSTRACT:

Introduction: Hemangiopericytoma is a mesenchymal tumour originating from pericytes surrounding the capillary vessels. The etiology of this tumour is still unknown. It may be located in any part of the human body. The most common sites are pelvis and lower limbs and less often it occurs in the head and neck. The characteristic signs of hemangiopericytoma are slow growth regardless of the malignancy level and high vascularity. Surgical excision of the tumour with additional radiotherapy is the treatment of choice.

Case report: A symptom presentation and diagnostic-therapeutic process in a 71-year-old woman admitted to the Department of Otolaryngology of the Medical University of Warsaw due to increasing bilateral nasal obstruction and dysphagia is herein described. Diagnostic imaging showed highly vascularized tumour in the parapharyngeal space also obstructing the left side of the nasopharynx.

After evaluation, tumour resection from the external approach was planned and performed, followed by an uncomplicated postoperative course. Histopathological examination revealed hemangiopericytoma. The patient was qualified for adjuvant radiotherapy. She stays under observation with no signs of recurrence since surgery.

Conclusions: Hemangiopericytoma is a rare tumour with a wide variety of clinical presentations and a relapse-free survival that is difficult to predict. Long-lasting asymptomatic tumour growth results in late diagnosis. Imaging and primarily, histopathological examination are fundamental for the diagnostic process. The best outcome is observed after radical surgical treatment. In order to reduce the risk of recurrence, additional radiotherapy is advised. Local recurrence may appear years after finished treatment, therefore long-term follow-up is essential.

KEYWORDS: hemangiopericytoma, parapharyngeal space, mesenchymal neoplasms

STRESZCZENIE:

Hemangiopericytoma jest nowotworem mezenchymalnym, który wywodzi się z perycytów otaczających naczynia włosowate. Etiologia tego nowotworu nie została dotychczas poznana. Może lokalizować się on w dowolnej części ciała, najczęściej występuje w miednicy i kończynach dolnych, znacznie rzadziej w rejonie głowy i szyi. Charakterystyczną cechą guza jest jego powolny wzrost, niezależnie od stopnia złośliwości, oraz bogate unaczynienie. Leczeniem z wyboru jest operacyjne usunięcie guza z zastosowaniem uzupełniającej radioterapii.

Opis przypadku: Opisano przebieg objawów klinicznych, proces diagnostyczny i terapeutyczny u 71-letniej pacjentki przyjętej do Katedry i Kliniki Otolaryngologii Warszawskiego Uniwersytetu Medycznego z powodu obustronnej niedrożności nosa i zaburzeń połykania nasilających się od 6 miesięcy. Po przeprowadzeniu diagnostyki obrazowej stwierdzono bogato unaczyniony guz obejmujący przestrzeń przygardłową oraz wypełniający nosogardło po stronie lewej. Zaplanowano i przeprowadzono resekcję guza z dostępu szyjnego. W badaniu histopatologicznym rozpoznano hemangiopericytomę. Okres pooperacyjny nie był powikłany. Chora zakwalifikowana została do uzupełniającego leczenia radioterapią. Od czasu zabiegu nie zaobserwowano wznowy miejscowej. Pacjentka pozostaje pod obserwacją.

Wnioski: Hemangiopericytoma jest rzadkim guzem, który charakteryzuje się różnorodnym obrazem klinicznym

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oraz trudnym do przewidzenia rokowaniem przeżycia wolnego od wznowy po zastosowanym leczeniu. Długotrwały, bezobjawowy wzrost nowotworu jest przyczyną rozpoznawania go w zaawansowanym stadium. Diagnostyka opiera się na badaniach obrazowych, a przede wszystkim na wyniku badania histopatologicznego. Najlepsze efekty przynosi radykalne leczenie operacyjne guza. W celu zmniejszenia ryzyka wznowy zalecane jest zastosowanie uzupełniającej radioterapii. Nawroty miejscowe mogą pojawić się nawet po długim czasie od zakończenia leczenia, dlatego konieczna jest wieloletnia obserwacja.

SŁOWA KLUCZOWE: hemangiopericytoma, przestrzeń przygardłowa, nowotwory mezenchymalne

INTRODUCTION

Hemangiopericytoma (HPC) is a rare tumor originating from pericytes that can be either benign or malignant [1]. Pericytes are located along blood vessels and they play a role in regulating capillary lumen, protecting endothelium and supporting angiogenesis [2]. The tumor can develop in any location near capillaries, however, it occurs most commonly in the pelvis and lower extremities. Approximately 30% of cases are found within the head and neck region, more than half of which are found within the paranasal sinuses and nasal cavity, less often inside the orbit, oral cavity, jaws, or parotid glands [3,4]. Until now, only few cases of HPC in the parapharyngeal space have been described [4,5].

The tumor affects both males and females at a similar rate, usually in their 50s and 60s [3]. The etiology is unknown, however, risk factors include hypertension, long-term steroid therapy, pregnancy, hormonal disturbances and trauma [3]. HPC is slow-growing regardless of its degree of malignancy. It has been observed that benign tumors tend to develop in the head and neck region, although this phenomenon has not yet been explained [1].

Malignant and benign types are determined based on pathology study, considering mitotic activity, cell features and presence of nuclear atypia [1]. Clinical features that suggest malignancy include considerable size and necrosis within the tumor [1]. Even benign types of HTC should be closely monitored since there is data in the literature confirming development of distant metastases in such cases.

The clinical course can initially be asymptomatic. With increasing size of the tumor, compression-related symptoms start to develop. In the case of parapharyngeal HPC infiltrating the nasopharynx, nasal bleeding and difficulty breathing are possible manifestations.

The treatment of choice is radical excision of the tumor with normal tissue margins [1-4]. For adjuvant treatment, as well as in the case of unresectable tumors or uncertainty as to complete excision, radiotherapy is applied.

CASE REPORT

A 71-year-old female patient presented to the Department of Otolaryngology of the Medical University of Warsaw due to globus sensation, dysphagia, bilateral nasal congestion and hearing loss lasting for the past six months. The symptoms worsened with time. Furthermore, the patient had difficulty speaking due to nasal speech. She had a history of hypertension, coronary artery disease and type 2 diabetes treated with oral medications.

One month before admission, she underwent biopsy of the tumor under general anesthesia at another facility. During the biopsy, bleeding from the tumor occurred. Pathology study results were non-diagnostic. The patient was referred to our department for further treatment.

On physical examination, dryness of oral mucosa and tongue was noted, as well as anterior displacement of the soft palate by the neoplastic mass in the nasopharynx with consequent restricted mobility, without mucosal infiltration of the palatine arches. The nasophrynx was completely filled with tumor causing bilateral obstruction of choanae.

On fiberoscopy, the tumor almost completely obstructed the choanae. On the right side, we managed to insert the end of the endoscope between the tumor and the lateral wall of nasopharynx visualizing the ceiling and the posterior wall, which were not infiltrated by cancer (Fig.1). The mobility of the tongue and vocal folds was normal.

Magnetic resonance imaging was performed in the patient, which revealed a large tumor sized 38 x 52 x 41 mm with well-defined borders that filled the left parapharyngeal space protruding to the nasopharynx. The tumor showed an intensive non-homogenous contrast enhancement. The suspicion of paraganglioma was raised (Fig.2). Computed tomography scan with vascular option did not reveal any vessels supplying the tumor (Fig.3).

The patient was qualified for surgical treatment. The parapharyngeal space was opened from the external neck approach

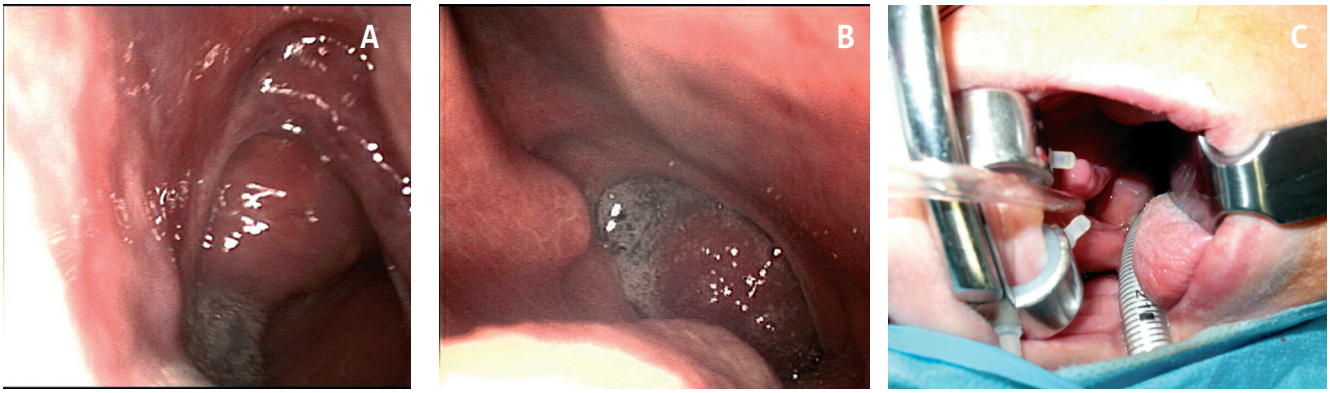

Fig. 1. Clinical presentation of hemangiopericytoma: on fiberoscopy of the nasal cavity, a tumor filling the entire nasopharynx can be seen on the right (1a) and left (1b) side; the intraoperative view from the oral cavity (1c) – the tumor protrudes beneath the soft palate.

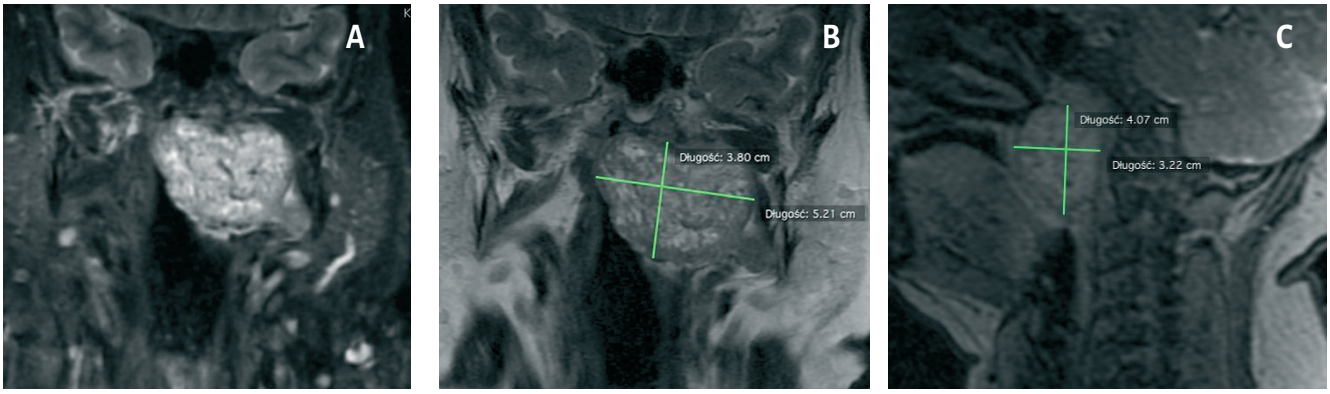

Fig. 2. Magnetic resonance imaging of left-sided parapharyngeal and nasopharyngeal hemangiopericytoma (2a, 2b, 2c).

after dissection of submandibular gland and transection of the stylomandibular ligament. An encapsulated tumor was visualized that fully filled the parapharyngeal space and expanded medially to the nasopharynx. (Fig.4). The posterior head of the digastric muscle as well as the stylohyoid muscle were transected for further access. The neurovascular bundle was dissected from underneath the sternocleidomastoid muscle, the following structures: the internal and external carotid arteries, vagus nerve and internal jugular vein were identified and captured. (Fig. 5). The lower portion of the tumor was resected, and then the intracapsular neoplastic mass was removed in order to reduce its volume. The capsule was separated from the deep portion of the parotid gland and from the cranial base. Due to infiltration of the nasopharyngeal mucosa, the tumor was resected together with nasopharyngeal mucosa and the lateral pharyngeal wall. The pharyngeal mucosa was sutured from the oral access. The nasogastric tube was placed.

After surgery, normal mobility of the soft palate and tongue was observed as well as normal voice. On indirect laryngoscopy, vocal fold mobility was normal and symmetrical on both sides and the piriform sinuses were free. The facial nerve function was bilaterally symmetrical and normal. On pathology study, the diagnosis of hemangiopericytoma was made. Immunophenotyping test result was as follows: CD34 – negative, CD 31 – negative, SMA (smooth muscle actin) – negative, desmin – negative, synaptophysin – negative, chromogranin – negative, CD56 – negative. Infiltration of nasypharyngeal mucosa was confirmed. The margins were free of cancer cells.

Antibiotic therapy including amoxicillin with clavulanic acid was sustained until postoperative day 7. On postoperative day 2, the drainage from the wound was removed. Postoperative healing of the wound in nasopharynx and on the neck was normal.

On day 13, the tube was removed and oral nutrition was resumed. The patient was discharged home in stable condition on postoperative day 14. After oncologic consultation, the patient was qualified for adjuvant radiotherapy at the total dose 30 – 40 cGy, the dose being fractionated.

DISCUSSION

Hemangiopericytoma was first described in 1942 by the team of Stout and Murray [1,2]. Initially, it was classified as a vascular neoplasm stemming from pericytes, i.e. connective tissue cells surrounding blood vessels [2]. Further immunohistochemical studies revealed similarities between HPC and the solitary fibrous tumor (SFT), which suggested common origin of both tumors, which is in accordance with the position of the World Health Organization. There is also a hypothesis stating that HPC derives from pluripotent cells surrounding blood vessels [5]. HPC accounts for 3-5% of all soft tissue tumors and ca. 1% of all vascular tumors [3]. The peak incidence is between 50 and 70 years of age (median age of 45), with no predilection for any sex [3,6]. The etiology is unknown, however, potential risk factors include hormonal disturbances, long-term steroid therapy, pregnancy, hypertension and trauma [3]. The most common locations are lower extremities (35%), pelvis and retroperitoneal space (25%), head and neck (15-30%), chest (14%), upper extremities (10%). Within the head and neck, HPC is most commonly found in the nasal cavity and paranasal sinuses, and less often in the orbits, oral cavity, maxilla, salivary glands, parapharyngeal space [3,7]. So far, only a few cases of HPC in the parapharyngeal space have been reported. At the early stages, the tumor usually does not cause any symptoms, and therefore its early detection is usually accidental. With progression and increasing size, compression- and infiltration-related symptoms arise [8]. On macroscopic view, HPC presents as a brown spongy mass surrounded by pseudocapsule with well-defined borders. It is often accompanied by satellite tumors distant from the main mass [3,9]. On microscopic examination, clusters of tightly-packed cells of oval or fusiform shape with ill-defined borders and dark-staining nuclei are seen. Between the cells, numerous vascular spaces are visible [3,5]. A constantly present marker of pericytoma is vimentin, which can be detected on immunohistochemical staining [3,7,10].

Pericytoma can be benign or malignant. Features indicating malignancy include: hemorrhage and necrosis, high cell density, presence of pleomorphic and immature cells, high mitotic activity (over 4/10), large size of the tumor (over 5 cm or 6.5 cm, according to various sources). However, it should be noted that pathological image does not always correlate with severity of the disease [1,8].

Hemangiopericytoma of the head and neck develops slowly regardless of the degree of malignancy, and is rarely associated with distant metastases to the brain, lungs, mediastinum, thoracic wall, axillae, abdomen and liver [9].

The diagnosis cannot be solely based on clinical presentation and macroscopic features of the tumor. The basis for diagnosis is a detailed pathological assessment. Diagnostic imaging studies are also used, which are significant for evaluation of size and infiltration of surrounding structures prior to surgery. Magnetic resonance imaging allows for the most precise determination of size and infiltration. Computed tomography is helpful in assessing bone infiltration. Angiography is performed in order to confirm vascular nature of the tumor, to evaluate its vasculature and assess the risk of intraoperative bleeding. In the case of large masses or risk of uncontrollable hemorrhage, embolization is possible, however, it is not routinely recommended [4,8,11]. Due to high risk of bleeding, fine-needle aspiration and tissue sampling are not recommended [8,10,11]. Differential diagnosis includes other vascular and soft-tissue

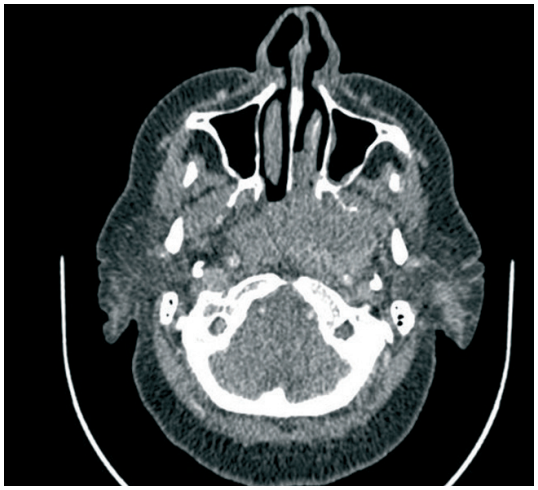

Fig. 3. Angio-CT of left-sided parapharyngeal and nasopharyngeal hemangiopericytoma.

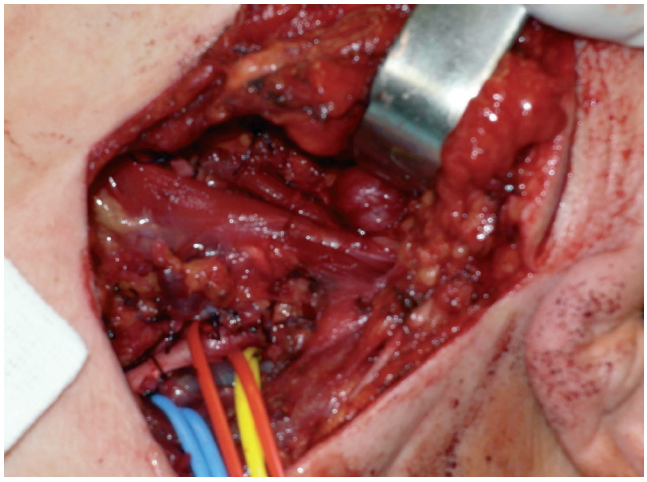

Fig. 4. Cervical access to the parapharyngeal space, after removal of the submandibular gland with neoplastic mass protruding from the space.

tumors, such as juvenile hemangioma, paraganglioma, angiosarcoma, leiomyoma, leiomyosarcoma, schwannoma, mesothelioma, liposarcoma, histiocytoma, chondrosarcoma, synovial sarcoma and cystadenocarcinoma [1].

Radical resection of tumor with normal tissues margins is the treatment of choice [1,3,7,10]. In patients, in whom complete resection was possible, the average 5-year survival rate was 100% [3,6]. In the case of large, not completely resected and potentially malignant tumors as well as recurrence, adjuvant radiotherapy is recommended [1,3,7,10]. In the literature, there have been reported cases of successful chemotherapy in aggressively growing tumors [8,9,11]. Despite common application of radiotherapy and chemotherapy, there are no reliable studies confirming their effectiveness, and thus no recommendations considering the dose or the extent of irradiation [1,3,7,8]. Local recurrence rate of hemangiopericytoma is estimated to be ca. 40% with latency ranging from 63 to 107 months [1]. Therefore, the patients should be subjected to long-term oncologic care and regular check-ups [1].

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