Cerebral infarction as a postoperative complication of a resection paraganglioma – a case report

Zawał mózgu jako powikłanie pooperacyjne resekcji przyzwojaka – opis przypadku

Krzysztof Kowalik¹, Andrzej Modrzejewski², Konrad Kaźmierczak¹

¹Department of Forensic Medicine, Pomeranian Medical University in Szczecin, Poland; Head: prof. Mirosław Parafiniuk MD PhD
²Department and Clinic of General Surgery, Pomeranian Medical University in Szczecin, Poland; Head: prof. Andrzej Modrzejewski MD PhD

ABSTRACT:

Introduction: Paraganglioma is a rare, usually benign neoplasm originating from the carotid bodies and belonging to a large group of head and neck tumors. It is characterized by slow growth and varied clinical manifestations, often remaining asymptomatic for a long time.

Case report: We present a case of a 19-year-old male patient with carotid body tumor who developed a postoperative left cerebral infraction.

KEYWORDS: cerebral infarction, paraganglioma

STRESZCZENIE:

Wstęp: Przyzwojak to rzadki, zwykle łagodny nowotwór, wywodzący się z ciałek przyzwojowych, należący do licznej grupy nowotworów głowy i szyi. Charakteryzuje się powolnym wzrostem i zróżnicowaną manifestacją kliniczną, często długo pozostaje asymptomatyczny.

Opis przypadku: Przedstawiamy przypadek 19-letniego mężczyzny z przyzwojakiem szyi, u którego doszło w okresie pooperacyjnym do powikłania w postaci zawału lewej półkuli mózgu.

SŁOWA KLUCZOWE: paraganglioma, przyzwojak, zawał mózgu

ABBREVIATIONS

CBT – carotid body tumor
CCA – common carotid artery
CT – computed tomography
ICP – intracranial pressure
ICU – intensive care unit
LN – lymph nodes
MRI – magnetic resonance imaging
PEG – percutaneous endoscopic gastrostomy
TnT – troponin T
US – ultrasound

INTRODUCTION

Paragangliomas (chemodectomas) are rare, usually benign (97%) tumors of the autonomous nervous system with the incidence rate of 2–8 cases per 1,000,000 individuals and malignancy rate of 3% [1]. The tumors develop from the primary neural crest within the paraganglial tissue. [2]. When developing from the sympathetic nervous system within the head and neck, paragangliomas are usually hormonally nonactive [3]. Paragangliomas of the head and neck most often derive from the carotid body located at the bifurcation of the common carotid artery (paraganglioma caroticum), which account for about 70% of all paragangliomas [3]. Carotid body tumors are most common in men whereas vagal paragangliomas are more common in female patients [4]. Paragangliomas of the head and neck may occur bilaterally or multilocally, more frequently in familial (25–50%) than in sporadic form (10%); usually, due to their slow growth and lack of hormonal activity, they remain asymptomatic for long periods [3]. However, lymph node or distant metastases may sometimes be formed and the tumor may damage the cranial base [4]. Clinical symptoms, if present, may be manifested in various forms including non-painful tubers within the neck region, bulbar nerve dysfunction, hearing loss, tinnitus, and raised intracranial pressure syndrome [5, 6].
Paragangiomas of the head and neck occur in two main forms, namely the sporadic form (ca. 90% of cases) and the familial form (ca. 10%) [2]. In children, the tumors develop very rarely [7]. In adults, the mean age at diagnosis is 30–50 years [3, 8].

**CASE REPORT**

A 19-year-old male patient was admitted to the Department of Otolaryngology for elective surgery of a carotid body tumor on the left side of the neck. History included a report of a non-painful tumor being detected by the patient upon shaving four months earlier. Neck ultrasound revealed a well-vascularized tumor sized 4 × 3 × 3 cm located on the left within the CCA bifurcation region; microscopic examination of the collected material revealed the paraganglioma lesion. Neck CT scan revealed the presence of an intensely enhancing tumor originating within the left retromandibular area and extending upwards to the level of the lower margin of the second cervical vertebra and below the level of the mandibular angle. The tumor was closely adjacent to cervical vessels. Blood was supplied to the tumor by pathological vessels originating from the left posterior auricular artery. The supplying vessels were embolized to achieve a reduction in tumor mass by approximately 75%. Follow-up angiography of the left common carotid artery revealed a hypervascularized zone within the CCA bifurcation projection area as well as the presence of tiny, pathological vessels originating from this artery at the bifurcation region to supply the tumor. Due to the location and small caliber of these vessels, an attempt at their embolization was abandoned. Three days later, tumor resection was carried out. The patient was subjected to general endotracheal anesthesia. Following the skin incision, the tumor was reached and found to be well vascularized, with helical vessels about 6 cm in length entwining around its body. At the inferior pole, the lesion was found to infiltrate the carotid artery bifurcation. The superior pole of the tumor reached the left styloid process. In the next stage of the procedure, the branches of the left external carotid artery (facial, lingual, and superior thyroid arteries) were ligated. The superior pole of the tumor was released and the left hypoglossal nerve was found to be strangled by the tumor vessels. The sympathetic trunk taken over by the tumor was removed. The internal carotid artery was atrophic, its lumen being narrow. The operating surgeon made a decision to clamp the common carotid artery using a Satinski clamp for approximately 40 minutes to facilitate the dissection of the well-vascularized tumor. The surgeon closed the vessel to continue dissecting the tumor in the bifurcation region.

During the dissection, an arterial wall rupture occurred within the bifurcation leading to massive bleeding. The attempt at further dissection was abandoned, a Satinski clamp was placed and the suture of the left common carotid artery bifurcation followed as a life-saving procedure. After approximately 40 minutes, the flow was restored and proper CCA pulsation was observed. Hemostatic mesh was placed around the anastomosis and the wound was sutured. After the surgery, the patient remaining in induced coma was transferred to the post-operative room of the Department of Otolaryngology. The patient was circulatorily efficient and mechanically ventilated. On the next day, the patient received the first dose of low molecular weight heparin.

Two days later, a significant increase in arterial pressure was observed in the patient upon recovery from coma along with right-sided hemiparesis and left pupil being dilated with no reaction to light. The recovery was interrupted and the patient was put under general anesthesia. Neck ultrasound scan revealed thrombosis within the left common carotid artery. A head MRI scan was ordered to reveal a large area of fresh ischemia within the zone supplied from the left internal carotid artery in the left hemisphere (encompassing the frontal, parietal, and temporal lobes). In addition, features of moderate cerebral edema were observed on the left side resulting in narrowing of the lateral ventricle and discrete displacement of the ventricular system by about 6 mm to the right. Poor filling of the internal carotid artery was observed within the cranial base and in the intracranial segment in T2-weighted images, suggesting the presence of thrombus within the arterial lumen. Contrast agent was administered during the scan to reveal complete occlusion of the left internal carotid artery and the main trunk of the middle cerebral artery; the patient was hence diagnosed with left hemisphere cerebral infarction. The patient, in grave condition, was transferred to the anesthesia and intensive care unit. Upon admission, the patient remained deeply unconscious, his pupils wide and not reacting to light. The level of consciousness was assessed as GCS score of 3. Laboratory results included prolonged prothrombin time, CRP level of 110, creatine kinase level of 3183 U/L, D-dimers level of 16.02 µg/mL, fibrinogen level of 443 mg/dL, and TnT level of 21 ng/mL. Following neurosurgical consultation and due to the significant increase in intracranial pressure (ICP), the patient was fitted with an intracranial hypertension monitor; the measured ICP value was below 20 mmHg. The patient remained in deep sedation and under mechanical ventilation. Cerebral edema was observed in the follow-up head CT scan. On the next day of hospitalization, the patient’s condition was deemed critical, with neurological examination revealing wide, immobile pupils. A Doppler scan of the brain revealed blood flow within the right cerebral vessels. ICP was at 20–35 mmHg. Another CT scan of the head revealed the ventricular system being displaced by 15 mm to the right with left lateral ventricle compression and features of left hippocampal uncus being wedged in under the cerebellar tentorium as well as features of cerebellar tonsil impingement. Supratentorial as well as infratentorial subarachnoid spaces were compressed. A decision was made to perform an emergency left-sided decompression craniectomy. After the procedure, a 2-mm displacement of the ventricular system was observed in the subsequent head CT scan. The features of uncus being wedged in under the tentorium were still visible. Subarachnoid spaces remained compressed and narrowed. No signs of cerebellar tonsil impingement were observed. Doppler ultrasound of the right lower limb revealed thrombosis of external iliac and femoral veins. On the following days, angio-MRI scans were performed to reveal extensive ischemic lesions with evolution of edema and characteristics of hemorrhage within the left hemisphere as well as the increase in ventricular system enlargement as compared to the previous study.

On the subsequent days of hospitalization, the ICP monitor was removed following the ICP drop to below 10 mm Hg while the patient remained intubated and in severe condition. Right-sided anisocoria and characteristics of right-sided spastic hemiparesis were observed in neurological examination. Due to swallowing disorders, percutaneous endoscopic gastrostomy (PEG) was installed. On the
30th day of hospitalization, respiratory infection with septic symptoms (fever, tachypnoe and tachycardia of 150 bpm) developed. Clinical condition improved following broad-spectrum antibiotic therapy.

Upon discharge from the department, patient's general condition was described as controlled, the patient being minimally conscious with the GCS score of 8. Pupils were evenly sized, wide, not reacting to light, the patient efficiently breathed on his own via the tracheostomy tube and was circulatorily efficient on passive oxygen therapy. Inflammatory marker levels decreased. The patient was transferred for further rehabilitation. Secondary malignant cerebral edema with brainstem ischemia developed. On the 91st day of hospitalization, the responsive yet nonverbal patient was discharged from the ICU for further treatment within the Paraplegia Department. Left-sided cranioplasty was carried out. No significant improvement was observed after more than 10 months of treatment.

DISCUSSION

Despite their low incidence, head and neck paragangliomas pose a serious diagnostic as well as therapeutic problem due to being burdened by a relatively high rate of intraoperative complications. Correct diagnosis, analysis of imaging studies, case-by-case approach, and therapeutic risk assessment are crucial for the success of the management in these tumors. In most cases, surgical resection of the tumor is the treatment of choice. The method is usually curative [9, 10].

Paragangliomas are characterized by a wide spectrum and non-specific nature of symptoms. Frequently, the disease is low-symptomatic and is diagnosed in imaging studies commissioned for other reasons. The most common (76%) symptom reported by patients in relation to the carotid body tumor consists in the presence of painless, usually asymptomatic mass located in the projection of the common carotid artery bifurcation as was in the case of our patient. Other symptoms occur much less frequently (0.1–3%) and are due to the presence of tumor mass compressing the surrounding tissue. These include cranial nerve irritation or palsy, intermittent tinnitus, loss of consciousness, dizziness, dysphagia, neck and head pain. Tumors with a significant mass may make cerebral perfusion difficult, possibly leading to transient ischemic attack or (very rarely) stroke [11, 12]. Differential diagnostics of the condition of a patient who reports a mass located within the projection of CCA bifurcation should include potentially enlarged lymph nodes, pathological hyperplasia of salivary glands and carotid artery aneurysms [13]. Imaging studies of lesions located at carotid bifurcation should begin with duplex Doppler ultrasound scans which, in the case of a CBT, should reveal the presence of a heterogeneous, hypoechogenic mass between the internal and external carotid arteries. Doppler ultrasound also facilitates evaluation of tumor vascularity, with paragangliomas presenting as highly vascularized masses. The gold standard in the preliminary confirmation of the diagnosis consists of angiographic examination which helps in the assessment of the tumor stage as well as in precise determination of the vessel of origin of tumor vascularity [14]. In the presented case, fine needle biopsy was performed which is not recommended in the case of these lesions due to the high risk of bleeding during the procedure [15].

Following the CT scan, the presented lesion was described and classified as type III using the three-grade Shamblin classification system. The classification facilitates a preliminary and measurable assessment of the risks associated with the procedure. Tumors classified as types I and II are easy to remove and bear low risk of intraoperative complications. Surgeries of type III paragangliomas are associated with greater tumor masses, lower accessibility for resection, and greater incidence of complications [16].

The standard treatment of mild paragangliomas consists in surgical treatment aimed at radical resection of the tumor. In some cases, authors suggest the possibility of treatment starting with conventional radiotherapy. In older patients with short life expectancies, observation is suggested, particularly for small, asymptomatic, and slowly growing tumors [17].

In the presented case, preoperative embolization of the vessels supplying the tumor had been performed. This, however, is not necessary and depends on the operator’s preference. Embolization had been shown to significantly reduce blood loss due to the surgery and shorten the procedure duration; however, no evidence is available to support reduction in the number of other complications resulting from the resection [18]. One should also keep in mind the possibility of occlusive material being transported to cerebral arteries, potentially leading to ischemic stroke [19].

In the reported patient, the stage of tumor progression suggested the need for additional studies. If imaging studies indicate the need for temporary intraoperative closure of the CCA or ICA, a balloon occlusion test (BOT) should be considered to assess the effectiveness of cerebral perfusion originating from peripheral circulation. This facilitates predicting the efficacy of cerebral blood supply when only one of the carotid arteries remains patent.

In this procedure, anterior cervical access along the median border of the sternocleidomastoid muscle is recommended. An important element of the procedure consists in timely identification and looping of the CCA, the external and internal carotid arteries and the internal carotid vein. Next, it is essential to identify and dissect the neural structures such as the cranial nerves IX, X, XI, the hypoglossal nerve loop, and the cervical segment of the sympathetic trunk. Resection of these structures may be inevitable in cases of their incarceration within the tumor mass [20].

Prior identification and ligation of vessels supplying the tumor is required. Accurate control of tumor vascularity leads to a reduction in its mass and consistency within a period of few minutes, making the lesion much easier to resect. Tumors classified as Shamblin type III may require reconstruction of the internal carotid artery. Saphenous vein may be used for this purpose. Compared to ligation, arterial reconstruction is associated with a significantly lower risk of ischemic stroke [21]. Shunting may be necessary to avoid temporary ischemia [19].

The most common complication of carotid body tumor consists in damaging the cranial nerves or their branches (up to 25.4% of procedures), the most common injuries being those of the hypoglossal and vagus nerve; a less common complication consists in the
development of Horner syndrome. Correlation was observed between the Shamblin classification and the incidence of intraoperative injuries within the nerve structures; from 5% for type III tumors to 7% for type I tumors. A close correlation is also suggested between the higher classification type and elevated risk of ischemic stroke as observed in the presented case [16]. Electroencephalography [20] or cerebral flow evaluation by means of transcranial Doppler ultrasound [20, 21] may be helpful in assessing proper supply of blood to the cerebral tissue. Implementation of these measures may effectively reduce the number of ischemic incidents.

CONCLUSIONS

1. The surgical team performing CBT resection should be skilled in the reconstruction of damaged internal carotid artery;
2. Stroke may develop when the internal carotid artery remains closed for more than several minutes;
3. General anesthesia facilitates cerebral function being monitored during the closure of the internal carotid artery.

REFERENCES

Krzysztof Kowalik MD; Department of Forensic Medicine, Pomeranian Medical University in Szczecin; al. Powstańców Wlkp. 72, 70-111 Szczecin, Poland; Phone: +48 (091) 4661566; E-mail: krzysio.kowalik@gmail.com