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Paragangliomas of head and neck – A surgical challenge

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ABSTRACT

Paragangliomas of the head and neck region are rare and predominantly asymptomatic tumors. These neoplasms arise from widely distributed paraganglionic cells, which originate from both mesodermal elements of third branchial arch and the neural crest residues. Despite the rare occurrence of paragangliomas, problems encountered in their diagnosis, unclear malignancy and treatment ensure that they still remain in the focus of head and neck surgeons.

This is a retrospective study of the medical records of patients treated in the ENT Department of the 5th Military Hospital in Krakow during the period 2010–2014. All the preoperative, intraoperative and postoperative data were carefully analyzed for each patient.

Thirteen patients (16 tumors) were treated during the study period. All the patients with a preoperative suspicion of paraganglioma underwent computed tomography angiography. The whole cohort of patients was treated surgically.

Paraganglioma should be always considered in the differential diagnosis for painless neck masses. These tumors require thorough radiological pre-operative evaluation and skilled operative technique. Surgical treatment occurs to provide good cure rates with minimal recurrence and morbidity rates.

1. Introduction

Paragangliomas (PGL) of the head and neck region are rare (overall estimated incidence 1/300 000) and predominantly asymptomatic tumors (Erickson et al., 2001). These neoplasms arise from widely distributed paraganglionic cells, which originate from both mesodermal elements of third branchial arch and the neural crest residues (Wieneke and Smith, 2009). Glomus tumors in the region of head and neck derive from the parasympathetic and sympathetic paraganglia. The sympathetic paraganglia are distributed within the prevertebral and paravertebral sympathetic trunks or in the connective tissue of the pelvis. Parasympathetic paraganglia are found entirely in the head and neck region, along the branches of the vagal and glossopharyngeal nerves. The most frequent locations are as follows: bifurcation of the common carotid artery (Fig. 1), jugular bulb, tympanic cavity and along the vagal nerve (Albsoul et al., 2009). Unusually they can be found in the larynx, trachea, orbit, nasal cavity and paranasal sinuses, nasopharynx and thyroid. According to the World Health Organization (Barnes et al., 2005) the extra-adrenal paragangliomas are categorized as sympathetic and parasympathetic types. In the head and neck region they are classified as carotid body paraganglioma, jugulotympanic paraganglioma, vagal paraganglioma, laryngeal paraganglioma and miscellaneous paraganglioma.

Head and neck glomus tumors in the vast majority (95%) are non-catecholamine secreting and usually diagnosed due to the presence of painless neck mass or after involvement of the lower cranial nerves leading to dysphonia, dysphagia and hearing disorders (Baysal and Myers, 2002).

Parasympathetic paragangliomas are in 30% familial and inherited in the autosomal dominant model with genomic imprinting. To date, 14 genes have been identified in which mutations are involved in the pathogenesis of the paraganglioma or phaeochromocytoma (Lalloo, 2016; Bennedbeak et al., 2016). Most common genetic syndromes burdened with an increased risk of

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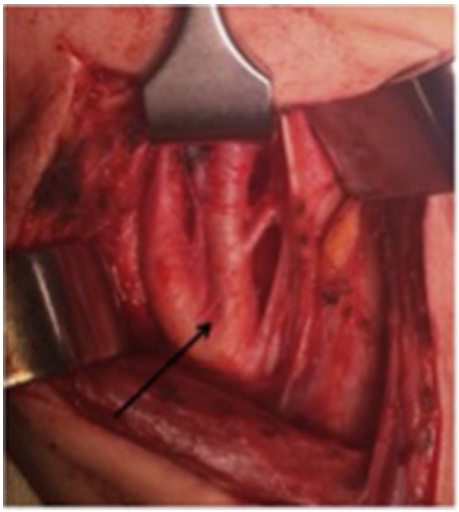

Fig. 1. Carotid body. Intraoperative picture: arrow indicates the localization of the carotid body – bifurcation of the common carotid artery.

paraganglioma are Multiple endocrine neoplasia type 2A and 2B, von Hippel-Lindau disease, Neurofibromatosis type 1, the Carney syndrome (Carney, 1999), the Carney–Stratakis dyad (Carney and Stratakis, 2002) and Hereditary Paraganglioma Syndrome (mutations in the mitochondrial succinate dehydrogenase complex subunits) (Martins and Bugalho, 2014). Parasympathetic tumors are more often familial than sympathetic types. Malignancy rate is estimated to 14–50% for sympathetic paragangliomas and 1–13% for parasympathetic (Barnes et al., 2005). In this type of neoplasms the clinical behavior, malignant potential and prognosis cannot be predicated from the histopathological examination.

Carotid body tumors are the most frequent PGL of the head and neck.

Despite the rare occurrence of paragangliomas, problems encountered in their diagnosis, unclear malignancy and treatment ensure that they still remain in the focus of head and neck surgeons. Because of the difficulties according to their high vascularity, proximity and possibility of the infiltration of the carotid arteries and cranial nerves, and risk of the extension to the skull base, they present a surgical challenge (Gad et al., 2014).

2. Material and methods

This is a retrospective study of the medical records of patients treated in the ENT Department of the 5th Military Hospital in Krakow during the period 2010–2014. All the preoperative, intraoperative and postoperative data were carefully analyzed for each patient.

Thirteen patients (16 tumors) were treated during the study period – 11 (84,6%) females and 2 (15,4%) males, aged between 27 and 77 (average 51,61). 3 tumors (23%) were localized on the left side and 8 (61,5%) on the right side. There was one case of bilateral tumors and one of the multifocal paragangliomas. The average duration of symptoms before surgical treatment was 12 months. Only 3 (23%) patients had other symptoms in addition to the presence of the neck mass: dysphagia (due to the tumor pressure or CN IX and X palsy), dysphonia (due to CN X palsy), tender lateral neck mass. All of the studied cases were non-catecholamine secreting tumors. There were 8 (61,5%) cases of concomitant cardiovascular diseases and 3 (23%) cases of the respiratory system disorders. Only two out of thirteen patients admitted to being an active smoker and one had a history of the nicotine addiction in the past. Familial history of paraganglioma/pheochromocytoma occurred in one patient.

All the patients with the pre-operative suspicion of paraganglioma underwent computed tomography angiography (CTA) study. No regional lymph node metastases were found in the preoperative imaging. The whole cohort of patients was treated surgically. 15 (94%) out of 16 of the analyzed tumors were diagnosed as carotid body tumors (CBT). One case of paraganglioma (in the patient with multifocal tumors and the positive family history of neuroendocrine tumors) was diagnosed as the vagal paraganglioma. According to the modified Shamblin's classification (Fig. 2) (Shamblin et al., 1971; Luna-Ortiz et al., 2006) there were: 2 (13,3%) cases of class I tumor, 6 (40%) tumors of class II, 6 (40%) tumors of class IIIA and one (6,7%) of class IIIB. Pre- and postoperative laryngoscopy was performed to evaluate the function of the laryngeal and vagal nerves.

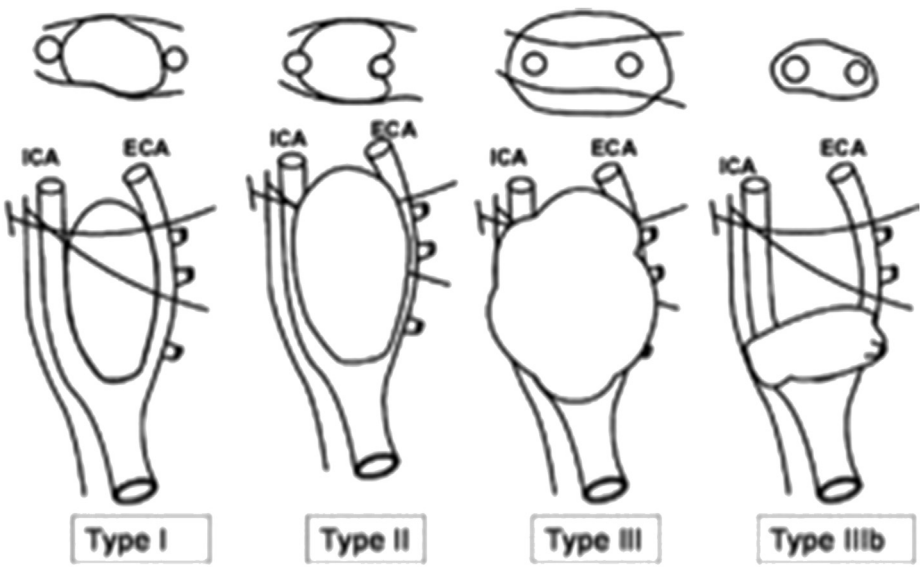

Fig. 2. Schematic diagram of the modification to Shamblin's classification of CBT by Luna-Ortiz et al. (2006). The division depends on the relationship of the PGL tumor to the wall of the carotid vessels (ECA – external carotid artery, ICA – internal carotid artery), superior laryngeal nerve and hypoglossal nerve.

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