Eur J Gen Med 2014; Suppl 1: 24-27
DOI : 10.15197/sabad.1.11. 24

Case Report

Familial paraganglioma of head and neck

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ABSTRACT

Paragangliomas are tumors originated from non-chromaffin paranganglia cells derived from the neural crest. These highly vascular tumors are located in the region which extends from skull base to arcus aorta. Head and neck paragangliomas have rare occurence constituting 0.012% of all the body tumors and 0.6% of head and neck tumors. Head and neck paragangliomas occur in sporadic and familial forms. The ratio of familial head and neck paragangliomas is about 10%. Familial form has a characteristic of autosomal dominant inheritance. In this article, four cases having first and second degree of affinity that we followed for paraganglioma diagnosis are presented.

Key words: *Familial, paraganglioma, head, neck*

Ailesel Baş ve Boyun Paraganglioma

ÖZET

Paragangliyomalar nöral krest kökenli non-kromafin paragangliyon hücrelerinden kaynaklanan tümörlerdir. İleri derecede vasküler olan bu tümörler kafa tabanından arkus aortaya kadar uzanan alanda bulunur. Baş boyun paragangliyomaları nadir görülür ve tüm vücut tümörlerinin %0.012'sini, baş-boyunda görülen tümörlerin ise %0.6'sını oluşturur. Baş boyun paragangliyomaları sporadik ve ailesel olmak üzere iki formda görülür. Ailesel baş boyun paragangliyomalarının oranı yaklaşık %10'dur. Ailesel form otozomal dominant geçiş özelliğine sahiptir. Bu yazıda aralarında birinci veya ikinci derece akrabalık bağı olan paraganglioma tanısı ile takip ettiğimiz 4 olgu sunulmuştur.

Anahtar kelimeler: *Ailesel, paragnaglioma, baş, boyun*

INTRODUCTION

Paragangliomas are tumors originated from non-chromaffin paranganglia cells derived from the neural crest. Head and neck paragangliomas have rare occurrence constituting 0.012% of all the body tumors and 0.6% of head and neck tumors. These tumors generally occur in mid-adult life and they have higher occurrence in females. In etiology, familial inheritance and chronic hypoxia are the known risk factors. These highly vascular tumors are located in the region which extends from skull base to arcus aorta (1). Head and neck paragangliomas occur in sporadic and familial forms. The ratio of familial head and neck paragangliomas is about 10%. Familial form has a characteristic of autosomal dominant inheritance. Head and neck paragangliomas can be single or multiple. Generally, multiple tumors occur in familial form. Multiple tumors occur in 30-40% of familial cases, and in less than 5-10% of sporadic cases (2). Although clinical and radiological findings are important in diagnosis, absolute diagnosis is carried out histopalogically. Moreover, high jugular bulb, aberrant carotid artery, carotid aneurysm, arteryvenous malformation and idiopathic hemotympanum should be taken into consideration for diagnosis (3). Despite of slow growing and generally showing benign characteristics, surgical treatment of these tumors should be performed carefully due to being highly vascular and neighboring to important formations (4). In this article, four cases having

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Received: 13.10.2011, Accepted: 28.02.2012

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first and second degree of affinity that we followed for paraganglioma diagnosis are presented.

CASE 1

Fifty three year-old male subject has initially developed pulse-synchronous tinnitus in both ears and hearing loss 28 years ago. 5 years after the initial symptoms, he applied to our clinic upon aching swelling by right ear and ear discharge. During the otoscopic examination, a hemorrhagic-like mass filling both external ear canals was found. Other cranial nerve examinations of the patient with left peripheral facial paralysis and right nervous hypoglossal paralysis were normal. Systemic examination and routine blood tests were normal. There were mixed type hearing losses at both ears in the pure tone audiogram. Magnetic resonance imaging (MRI) data showed a hypervascular mass lesion size of 1.8x3.5x4.5 cm infiltring the temporal bone and jugular foramen initiated from carotid bifurcation extended to the base of the skull on the right side, and size of 3x3.5x8 cm indenting the epiglottis and hypopharynx displaced to the right on the distal left carotid artery. Surgery was recommended to the patient with glomus tumor as a result of a biopsy previously taken in another center from the left external ear canal, patient did not agree to the surgery and radiotherapy (RT) was administered.

CASE 2

Thirty nine year-old male patient has developed hearing loss 25 years ago and it increased over the time. Patient complaining of dizziness from time to time and pulse-synchronous tinnitus has developed left ear discharge and aching swelling by left preauricular region 21 years ago and a facial paralysis has occurred in a matter of months. During the otoscopic examination of the patient, there was a hemorrhagic polypoid-like mass filling left external ear canal and the left thympanic membrane could not be evaluated. Other cranial nerve examinations except facial nerve were normal. Nothing found in systemic examination and routine blood tests. There was a medium mixed type hearing loss at left ear in the pure tone audiogram. Computed tomography (CT) data taken at another center showed that the left portion of the petrous bone of skull base and transverse bulge of C1 were destructive looking on the left side. A mass, located close to the external ear canal filling the back of zygomatic arch in the front, extended to the nasopharynx and filling the nasopharynx slightly on the inside, non-homogeneously and densely staining with contrast agent was also found. RT was administered to the patient with glomus tumor as a result of a biopsy taken at the same center from the left external ear canal.

CASE 3

Forty three year-old male with hoarseness, applied to another center 8 years ago, has been diagnosed with vocal cord paralysis. At that time, a mass size of 4x3.5x3 cm, extends to the anterior commissure obliterating the parapharyngeal fat tissue filled the pyriform sinus on the left side, and size of 4x4x3.5 cm, encompasses internal carotid artery on a bifurcation level on the left side, pushes jugular vein to lateral, was detected in the neck MRI. Patient who did not accept the surgery evidently had been discharged. Patient complaining with hearing loss and pulse-synchronous tinnitus for ten years has applied to our clinic 10 months ago for the right peripheral facial paralysis. During the examination, besides hemorrhagic mass prolapsing the right external ear canal and right facial paralysis, right hypoglossus and right vocal cord paralyses were also detected. Other ear, nose and throat (ENT) examinations of the patient did not revealed pathology. MRI and CT data showed a hypervascular mass lesion size of 1.8x3.5x4.5 cm infiltring the temporal bone and jugular foramen initiated from carotid bifurcation extended to the base of the skull on the right side, and size of 3x3.5x8 cm surrounding common carotid artery, internal and external carotid arteries on the distal left carotid artery, located between left hyoid bone and laryngeal cartilages indenting the epiglottis and hypopharynx displaced to the right. There was a severe mixed type hearing loss at left ear in the pure tone audiogram. Patient was operated. There were no complications after the surgery.

CASE 4

Twenty five year-old female patient complaining with pulse-synchronous tinnitus applied to our clinic 5 months ago. During the examination of patient with dizziness from time to time, a hyperemic lesion at the inferior quadrant of the tympanic membrane was detected.

Pathology was not detected during other ENT examinations of the patient having normal full cranial nerve examination. There was a medium mixed type hearing loss at left ear in the pure tone audiogram. Patient's MRI showed a mass size of 13x19 mm exhibiting evident vascularity located at the inferior surface of the petrous bone in the left hypotympanum. Patient was operated upon the selective embolization with the preliminary diagnosis of tympanic paraganglioma. There were no complications after the surgery.

DISCUSSION

Paraganglioma due to paraganglionic tissues of the temporal bone is generally benign type and it is the most common neoplasm of the middle ear. It is the second most common skull base benign tumors after the vestibular schwannoma (5). Paragangliomas so-called chemodectoma or glomus tumor develops at the paraganglionic tissues located at the bifurcation of the common carotid artery, jugular foramen, middle ear and posterior mediastinum. The most common paraganglioma is the carotid, followed by the jugulotympanic and vagal paragangliomas (6). In paragangliomas, the time passed between the presence of symptoms and the diagnosis can differ from several months to long periods such as 25 years and on average it takes about 3-4 years to diagnose after developing the symptoms (7). The reason for the delayed diagnosis is that symptoms of paragangliomas are not specific to the disease and can be associated with many other diseases. In our patients, upon developing the symptoms, the diagnosis could be made after 4 years in the first case, after 5 years in the second case, and during the first year in both the third and fourth cases. The ratio of cranial nerve involvement is about 35% and the ratio of intracranial invasion is 18-20% in jugular paraganglioma (8). There may be different symptoms and findings in head and neck paragangliomas. These findings are as follows in prevalence order; hearing loss, pulsatile tinnitus, ear discharge, otalgia, dizziness, ear bleeding, cranial nerve paralyses, mass in external ear canal. If the tumor wraps around the base of skull or the facial canal, cranial nerve abnormalities may occur. In frequency order, wrapped around cranial nerves are the cranial nerves 7, 10, 11 and 12 (9). There were pulsatile tinnitus, hearing loss, ear fullness, dizziness in our all four cases, facial and hypoglossal nerve involvement in the first case, only facial nerve involvement in the second case, facial, hypoglossal and vagal nerve involvement in the third case. There was no cranial nerve involvement in the fourth case.

Besides otolaryngological and neurological examinations, audiometer, cranial MRI, angiography and CT are helpful in the diagnosis. Computed tomography is particularly useful in showing the destruction in the temporal bone. MRI shows the vascularity of the lesion and it is not affected by the bone artefacts in the posterior fossa. Therefore, it is more advantageous than imaging with CT (10). It is recommended to perform angiography and MR angiography tests to decide which approach will be used in treatment when the venous condition of the lesion is important (11). Fundamental therapy approach in paragangliomas is the surgical excision following the superselective embolization. However, due to the difficulty of the surgical interferences with cases having common bone destruction and intracranial invasion, radiotherapy or embolization can be a treatment choice for older or debilitated cases (12). Late stage radiotherapy causing malignity emphasizes the importance of permanent embolization. In the permanent embolization, cure can not be the case, only tumor control can be mentioned. Following the permanent embolization, there was a significant regression in clinical and radiological findings along with decrease or loss of patient's complaints (13). Consequently, despite the rare occurrence of familial paraganglioma, a familial study should be carried out in all head and neck glomus tumors especially in subjects with multiple localizations. To detect the tumor in its early stages in patients with family history and to reduce the possible morbidity following the surgical treatment due to tumor growth, monitoring and treatment planning should be executed in these patients at early stages.

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