Bangladesh J Otorhinolaryngol 2013; 19(1): 58-64

Case Report

Recurrent paraganglioma of occipito-temporal bone masquerading as temporal bone malignancy

Sushil Kumar Aggarwal[1], Rajkumar[2]

Abstract:

A 33-year-old male patient presented with left facial nerve palsy, left earache along with left ear bloody discharge for last 4 months. He also had left vocal cord paralysis and impairment of hearing on left side. Contrast enhanced computed tomography (CECT) of temporal bone revealed enhancing mass present in left middle ear and mastoid and also involving left temporal and occipital bone causing destruction of bone, mimicking as temporal bone malignancy. Complete excision of tumor was done and biopsy came out to be paraganglioma. Patient had been operated previously for similar lesion 8 years ago. Paraganglioma with such extensive involvement of bone is not reported in literature and hence we describe this case to distinguish it from temporal bone malignancy. The tumor was not functional clinically as well as biochemically.

Keywords: Glomus Jugulare; Temporal bone malignancy; Recurrent paraganglioma

Introduction

Glomus jugulare tumors are rare, slow-growing, hypervascular tumors that arise within the jugular foramen of the temporal bone. They are included in a group of tumors referred to as paragangliomas, which occur at various sites and include carotid body, glomus vagale, and glomus tympanicum tumors[1].

Jugulotympanic paragangliomas are the second most common temporal bone tumor (after acoustic neuromas). The incidence of jugulotympanic paragangliomas is approximately 1:1,300,000. Unlike carotid paragangliomas, there is a definite female predominance in the incidence of jugulotympanic paragangliomas with a female to male ratio of 4:1, though our patient was male. The median age of presentation is 50-60 years, contrary to our patient who was in third decade of life. Like the carotid paraganglioma, the jugulotympanic paraganglioma has a sporadic and familial form. The familial form is associated with a higher incidence of multicentricity (approximately 25-50%). The incidence of a functional or secreting jugulotympanic paraganglioma is lower than the carotid paraganglioma, occuring in about 1-3%. The malignancy rate is less than 5%.[1, 2]Although, they are histologically benign, they may

1. Senior Resident (ENT), Deptt of Neurosurgery, SGPGIMS, Lucknow-226014, India
2. Professor and Head, Deptt of Neurosurgery, SGPGIMS, Lucknow- 226014, India

Address of Correspondence: Dr Sushil kumar Aggarwal, Senior Resident (Neuro-Otology), Department of Neurosurgery, SGPGIMS, Lucknow-226014, e-mail: doc.sushil.pgi@gmail.com

behave locally as malignant tumors, with extensive invasion of bone, soft tissue, and nerves[3].

Here, we present a unique case of recurrent paraganglioma involving the temporal and the occipital bone and mimicking clinically and radiologiclly as temporal bone malignancy. No such presentation of temporal paraganglioma has been described in the literature till date.

Case Report

A 33-year old male presented to out-patient deptt of our institute with chief complaints of left facial nerve palsy, left earache and left ear bloody discharge for last 4 months. Patient also complained of hoarseness of voice, left ear tinnitus and mild impairment of hearing in left ear. He also had history of unsteadiness while walking with swaying present to left side. On examination, he had left-sided cerebellar signs with impaired tandem gait, left vocal cord paralysis, left facial nerve paralysis and left palatal paralysis. The left tympanic membrane was not visible with some mass present in left external auditory canal (EAC) with bloody discharge coming out of it. Pure tone audiometry revealed moderate sensorineural hearing loss in left ear whereas impedance audiometry showed B-type curve in left ear. Contrast enhanced computed tomography (CECT) temporal bone revealed enhancing mass present in middle ear and mastoid air cells, and also involving occipital and temporal bone with bone destruction (Figure 1 and 2). The squamous part of occipital bone was also thickened on left side. The left cerebellar hemisphere was compressed by this thickened occipital bone. The right cerebellar hemisphere was normal with no signs of hydrocephalus.

Patient was planned for surgical excision and tumor was completely excised by radical mastoidectomy along with petrosectomy and excision of involved squamous part of temporal and occipital bone was also done. There was no intradural extension of tumor and hence brain parenchyma was not involved. Left EAC was completely closed.

Following surgery and after post-operative recovery, patient had complete relief from pain behind the ear as well as unsteadiness. Histopathological examination revealed the tumor as paraganglioma with extensive bony involvement. A post-operative magnetic resonance imaging (MRI) scan of brain and temporal region was done which showed almost complete excision of tumor with brain parenchyma not involved at all. At 6 months of follow-up, patient still had paresis of lower cranial nerves with left EAC completely closed but he has drastic improvement in his pain and general appearance.

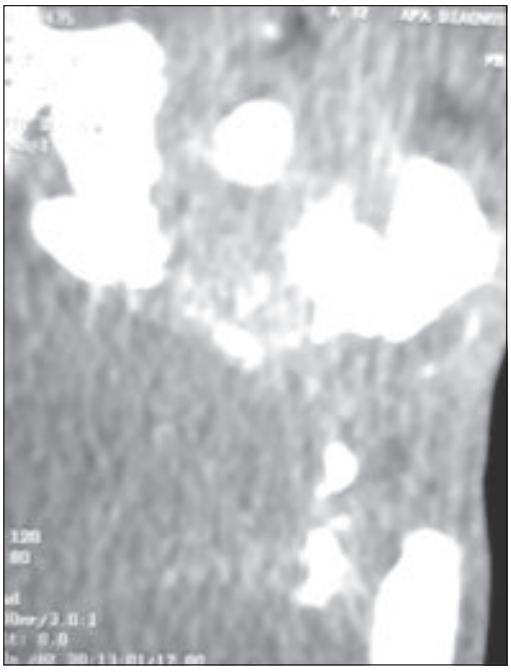

Figure 1: *Axial cut of contrast enhanced computed tomography (CECT) showing mildly enhanced tumor involving occipital bone and mastoid air cells with destruction of bone.*

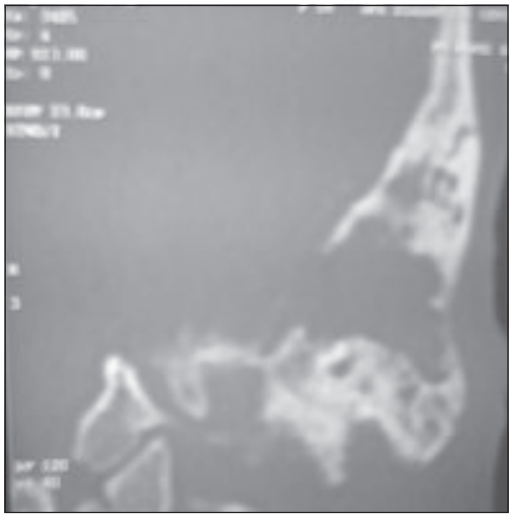

Figure 2: *Coronal cut of bony window of contrast enhanced computed tomography (CECT) showing destruction of mastoid air cells on left side*

Discussion

Paragangliomas are generally benign, slow growing tumors arising from widely distributed paraganglionic tissue thought to originate from the neural crest. Paraganglia are distributed throughout the head and neck and superior mediastinum along the course of the major vasculature. Paraganglia are also found in the orbit, the larynx, and along the course of the vagus nerve[1, 2]. Terms used in the past to describe these tumors based on their histopathologic and anatomical presentations included: glomus tumors, chemodectomas, carotid body tumors, and nonchromaffin tumors. Paragangliomas are most commonly found in the head and neck region. The most common cervical paraganglioma is the carotid paraganglioma (carotid body tumor). Paragangliomas can also involve the vagus nerve, the larynx, the orbit, and the nose and paranasal sinuses. Paragangliomas of the temporal bone include the jugulotympanic paragangliomas including the glomus tympanicum and glomus jugulare[1, 2].

The term paraganglion was first used by histologist Kohn in the early part of this century to describe the carotid body[4] . This term was most appropriate as cells of the carotid body originate from the neural crest and migrate in close association with autonomic ganglion cells. Currently, the correct terminology is paraganglioma based on the anatomical location (e.g. carotid paraganglioma and jugulotympanic paraganglioma)[4].

Guild first described vascularized tisue in the dome of the jugular bulb and on the promontory of the middle ear and named it "glomic tissue" in 1941[5]. On an average, there are three such bodies in each ear. These paraganglioma are usually found accompanying Jacobson's nerve (from CNIX) or Arnold's nerve (from CNX), or in the adventitia of the jugular bulb. Tumors of these paraganglioma are usually seen involving the mucosa of the promontory (glomus tympanicum) or the jugular bulb (glomus jugulare). The blood supply to jugulotympanic paragangliomas is the ascending pharyngeal artery via inferior tympanic and neuromeningeal branches[5].

All paragangliomas are closely related to one another and to pheochromocytomas of the adrenal gland. Their histiologic appearance is similar to the normal histology of the paraganglia. They consist of clusters of Type I or chief cells which are members of the amine precursor and uptake decarboxylase (APUD) family and Type II or sustentacular cells (modified Schwann cells). These two cell types are arranged into clusters with a core of chief cells surrounded by the sustentacular cells embedded in a fibrous stroma. The clusters of cells make up the histologic structure termed Zellballen. Nuclear pleomorphism and cellular hyperchromatism are common in paragangliomas and should not be considered evidence of malignancy[1, 2].

These tumors are very slow growing. They spread locally in a multidirectional fashion along pathways of least resistance within the temporal bone. The air cell tracts are the most important route of spread. Tumors have been noted to spread outside the temporal bone via the eustachian tube, vascular lumens, and neurovascular foramina including the internal auditory canal, though in our patient, the tumor had involved the temporal and occipital bone with mild destruction of the bone. Bone erosion is noted by distinct crescentic lucencies in the bone[1, 2].

The typical clinical course consists of slow continuous growth with few symptoms until the tumor has become far advanced. The patient usually presents with a complaint of pulsatile tinnitis. Other complaints may include aural fullness or hearing loss. Cranial nerve (CNs) deficits may be seen with larger tumors. Deficits of cranial nerves IX and X are most commonly seen but CNs VII, VIII, XI, and XIII can also be affected, as was seen in our case. Otoscopic examination can be normal or a characteristic red or reddish-blue mass may be seen behind the tympanic membrane. The tumor might grow through the tympanic membrane producing a vascular ear polyp, as was encountered in our patient.

Evaluation should begin with air and bone conduction audiometry. Imaging studies should include both a fine cut CT scan of the temporal bone (with axial and coronal images) and an MRI. On CT scan, jugulotympanic paragangliomas characteristically show bone erosion around the jugular bulb and carotid artery. CT scanning also helps delineate the tumor relationship to the facial nerve, the cochlea and to the internal carotid artery. MRI scanning is helpful in evaluating intracranial and intradural extension, presence or absence of flow in the ipsilateral or contralateral sigmoid sinus, and to further define the relationship of the tumor to the internal carotid. Arteriography aids in the detection of multicentric tumors, identifies feeding vessels, allows for embolization of the external carotid artery blood supply to the jugulotympanic paraganglioma, identifies intrasinus and intravenous extension, and provides further information on the adequacy of flow in the contralateral sigmoid and/or internal jugular vein. Additionally, it allows for balloon occlusion testing if prior imaging suggests extensive involvement which may require carotid repair or resection. In our patient, the arteriography was done but the characteristic tumor blush of paragangliomas was absent and hence it was misdiagnosed as temporal bone malignancy. Moreover, the involvement of temporal and occipital bone also misled us radiologically[6].

Two classification systems have been described based on tumor size, petrous apex or carotid artery involvement and intracranial extension. Fisch's classification system has four categories ranging from Type A tumors isolated to the middle ear cleft to Type D tumors with intracranial extension[7]. Glasscock and Jackson's system divides the jugulotympanic paragangliomas into glomus tympanicum and glomus jugulare tumors. Each group is then further divided into four groups (I-IV) based on their size and further extension throughout the temporal bone[8].

Decisions regarding management of these tumors can only be made after the extent of the tumor is defined. Unlike carotid paragangliomas, jugulotympanic paragangliomas are considered radiosensitive[9]. It should be understood that the goal of surgical treatment is the total or near-total removal of tumor while the goal of radiation therapy is the arrest of tumor growth. There is a considerable amount of controversy over the treatment of choice for jugulotympanic paragangliomas. Surgical risks include cranial nerve deficits, vascular injury and bleeding

and cerebrospinal fluid leak (4%). Radiation does not seem to affect the neoplastic component of the tumor (chief cells) that persists chronically after therapy and may continue to secrete catecholamines[10]. Radiation causes diminution of the small vessels, decreased arteriovenous shunting, and proliferative endarteritis[17], but does not seem to affect the larger vessels and causes no overall decrease in tumor vascularity. The overall effect seems to be proliferation of the fibrous stroma[11]. The tumor mass rarely decreases after radiation and may in fact continue to increase[12, 13]. Furthermore, the complications of radiation therapy may include temporary hair loss, mucositis, serous otitis media, stenosis of the external auditory canal, chronic otitis media, osteoradionecrosis, neuropathy of the fifth through twelfth cranial nerves, brain necrosis, and radiation-induced neoplasm or malignant transformation that arises on average 8 to 10 years after therapy[14, 15, 16]. Studies have shown that the risk of osteoradionecrosis is low if the optimal dose of radiotherapy of 35Gy/3weeks or 45Gy/4weeks is used[16, 17]. Hence, we also follow the protocol advocated by Carrasco and Rosenman that control of tumor by surgical eradication rather than alteration of biologic potential with radiation seems safer, especially in the younger patient[15].

Surgical techniques differ depending on the size and extent of tumor growth. For glomus tympanicum tumors limited to the mesotympanum and hypotympanum (Fisch Type A) without involvement of the jugular bulb, a transcanal tympanotomy may be all that is required. For larger tumors that extend into the mastoid (Fisch Type B), a canal wall up mastoidectomy with extended facial recess approach is used. Larger tumors that extend beyond the middle ear or involve the jugular bulb are approached like glomus jugulare tumors[7].

Glomus jugulare tumors that are classified by Glasscock and Jackson as Class I and II or by Fisch as C1 or C2 can usually be resected with an extended facial recess approach. The posterior canal wall can be taken down if necessary for exposure. In 1977, the group of infratemporal fossa approaches for the surgical management of tumors that extend into or beyond the petrous apex, involve the horizontal carotid artery, or involve the foramen lacerum or cavernous sinus, (Glasscock and Jackson Class III and IV or Fisch C3 and C4 tumors) were outlined,[3] with permanent anterior rerouting of the facial nerve and exposure of the intratemporal course of the internal carotid artery (ICA)[18]. These approaches allowed complete removal of even highly advanced lesions, including those situated within the infralabyrinthine and apical compartments of the temporal bone and have become the standard route for excision of these tumors, with significant reduction in morbidity and mortality. This approach allows the exposure necessary for distal control of the distal internal carotid artery. Again, dissection into the upper neck permits identification of the lower cranial nerves and control of the great vessels. Tumors with intracranial extension (Fisch D1-3) may be resected with the infratemporal fossa approach. Retrosigmoid and/or suboccipital approaches may be necessary for tumors with extension into the posterior cranial fossa.In our case, the conventional infratemporal fossa approach was not taken as we were not aware of the diagnosis and the site of origin of tumor was quite different from conventional paragangliomas with more involvement of temporal and occipital bone in our case[19, 20].

Patients are usually kept in intensive care unit overnight after infratemporal fossa surgery. If dysphagia is anticipated, a nasogastric tube should be placed. The

remainder of the post-operative care is directed towards the cranial nerve deficits. Post-operative radiation therapy should be considered in patients with residual tumor. These patients can also be observed with serial MRIs with radiotherapy reserved for evidence of tumor growth.

Symptoms of a secreting tumor should be sought including difficult to control hypertension, tachycardia, tremors or vascular headaches. If there is any suspicion of a secreting tumor, a 24 hour urine collection should be obtained for vanillyl mandalic acid (VMA), metanephrine, nor-epinephrine and epinephrine. If these tests are found to be abnormal, an abdominal CT scan should be obtained to rule out a concomitant adrenal pheochromocytoma, as jugulotympanic paragangliomas are rarely secreting tumors (1-3%)[21].

Paragangliomas of the larynx, orbit and nose and paranasal sinuses tend to be locally aggressive. Laryngeal paragangliomas typically require wide local resection or partial laryngectomy. Orbital paragangliomas are particularly aggressive and rapid recurrence is common after local resection. Nasal paragangliomas usually require wide local excision. Radiation therapy has not been shown to be effective against paragangliomas of the larynx, orbit or nose[10].

Conclusion

Jugulotympanic paragangliomas can have varied presentations in the temporal region and hence, suspicion of paragangliomas should be kept in any case of temporal bone malignancy to avoid intra-operative complications and for preparation of good back-up in case of excessive bleeding intra-operatively.

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