Glomus Tympanicum: A rare case of a painful bleeding ear

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Introduction

Glomus tumors are benign lesions of the middle ear which are slow growing in nature. They are the most common primary neoplasm of the middle ear [1]. Glomus tumor arises from abnormal growth of the paraganglion cells. The head and neck region represent the commonest area where extra adrenal paraganglioma also known as glomus tumors are found [2]. Patients may present with bleeding, pulsatile tinnitus and hearing loss. Surgical resections of the mass are the mainstay of treatment.

Case Report

A 24 year old man presented with left ear bleeding for 6 months in duration which was associated with otalgia, tinnitus and hearing loss. Otoscopy showed a vascular and angry looking mass filling up the entire left external auditory canal which bled on contact. No neurological or cranial nerve deficits were elicited. A high resolution computed tomography (HRCT) and a magnetic resonance imaging (MRI) showed an enhancing mass occupying the left jugular vein, left middle ear cavity, left mastoid air cells and left external auditory canal with infiltration into the left internal jugular vein and erosion of the surrounding bone. Digital substraction angiography showed a left glomus tumor which mass was supplied by the left ascending pharyngeal artery. The patient underwent embolization a day prior to the glomus tumor excision under general anesthesia. Intraoperatively the mass was noted to arise from the medial wall of the left middle ear, occupying the middle ear cavity and extending to the Eustachian tube and the hypotympanum. Part of the mastoid cavity was filled with soft tissue but the mastoid air cells were well aerated otherwise. The jugular bulb was not involved. Facial nerve was intact. Post operatively was uneventful. Histopathological examination of the mass showed features consistent with glomus tympanicum. On follow up 9 months later, patient was well and no evidence of tumor was found (Figures 1,2).

Discussion

Paraganglioma are rare tumors that make up about only 1% of head and neck tumors [2]. Glomus tympanicum (GT) also known as tympanic paraganglioma arises from the tympanic plexus of the Arnold and Jacobson nerve in the middle ear and are rarely functional. This differs from glomus jugulare which

Abstract

Glomus tympanicum is a rare tumor of the middle ear which typically present with pulsatile tinnitus, hearing loss and bleeding. Examination may reveal a vascular mass which bleed on contact. Imaging is important prior to surgical resection.

Statement of the problem: Here we present a rare case of a young man who presented with left ear bleeding and otalgia.

Result: Examination and imaging showed a vascular mass in the left external ear canal, left middle ear and involving the surrounding structures.

Conclusion: A differential of glomus tympanicum must be considered in cases which present with bleeding left external auditory canal mass and otalgia, although the most common presentations are hearing loss and tinnitus.
Glomus tympanicum is normally found in the middle age group with a mean age at presentation of 49-50 with the age range of 17 – 85 years old [2-4]. Our patient is a male patient and is younger than the normal age of presentation. Conductive hearing loss and pulsatile tinnitus which were present in the patient above are the two most common clinical presentations where around 60-80% of patients will present with the above complains [2,3]. Bleeding otorrhea may also be present while ootalgia which was seen in the above patient is an uncommon presentation.

On physical examination, a mass can be found in the middle ear or in the external auditory canal if it has extended beyond the tympanic membrane. Brown sign, blanching of the tumor with pneumatic pressure, is seen in a quarter to up to fifty percent of cases [2,3]. Cranial nerve involvement is rare and if there is involvement of the cranial nerves 9, 10 and 11, glomus jugulare must be suspected. Biopsy of the mass is not advocated as in our case above due to the high risk of bleeding. Histopathological examination shows an encapsulated tumor with an extensive capillary network and chief cells surrounded by sustentacular cells [2].

There are two types of classification for glomus tumor, Fisch and Mattox for middle ear and glomus tumor and the Glasscock and Jackson classification for glomus tympanicum [1,2]. Our patient is classified under Grade IV of the Glasscock and Jackson classification as the tumor involves the middle ear cleft with extension into the mastoid cavity and the external auditory canal but without involvement of the internal carotid artery. The list of important differential diagnosis may include cholesteatoma, inflammatory polyp, high jugular bulb, aberrant internal carotid artery, facial nerve neuroma and malignant neoplasms of the middle ear [4].

Imaging modalities are also important to determine the extension of the tumor and involvement of the surrounding structures. A high resolution computed tomography (HRCT) scan which show air or bone separating the jugular bulb and the tumor points to a glomus tympanicum. Salt- and-pepper appearance maybe seen in contrast enhanced magnetic resonance imaging (MRI) [2].

Treatment options include surgery and radiotherapy which encompasses stereotactic fractionated radiation therapy (SRT), stereotactic radiosurgery (SRS), intensity-modulated radiotherapy (IMRT) and conventional radiotherapy. The decision is made based on the site and the extent of the tumor growth. Local settings and availability of personnel and facilities also play a role in the decision making. In a case series by John A Forrest III et al on 80 patients, the long term tumor control after surgical excision is 92.5%. Post-operative complications are more commonly seen in extensive tumor especially those involving the skull base. Surgical complications include acquired cholesteatoma (1.7%), cerebrospinal fluid leak (0.9%), tympanic membrane perforation (1.7%), hearing loss and facial weakness. In our case, the patient above did not develop any of the above complications. Radiotherapy was previously only considered in cases where there is a tumor residual after surgery or in tumors that were inoperable but a study by Hinerman et al. [5], showed that radiotherapy can give a local control rate of up to 95% with a low complications rate. More conformal techniques of RT such SRS is increasingly becoming the choice of RT in view of shorter treatment time and smaller field size with lower complication rates [6]. SRS has a disadvantage of potential for a marginal miss of tumor. Chemotherapy is not effective in the treatment of paragangliomas.

Conclusion

Glomus tumors although rare must be suspected in patients including those in the younger age group, who present with hearing loss and pulsatile tinnitus with or without a external or middle ear mass. Otolgia is an uncommon presentation. Imaging is needed to delineate the lesion. Surgical resection or radiotherapy are treatment options which can be considered after taking into account the extent of the disease and the availability of the facilities in the local settings.

References


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