Facial nerve hemangiomas: A review

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Abstract Facial nerve hemangiomas are rare benign vascular tumors with primary symptoms including progressive facial nerve palsy and hearing loss. They can involve various segments of the facial nerve, especially the geniculate ganglion (G.G.). Preoperative MRI and CT imagining is of great importance for early diagnosis of such lesions. The most accepted treatment for facial nerve hemangiomas is surgical excision and the type of approach depends on tumor location, preoperative hearing level and tumor size. In consideration of the difficulties in differential diagnosis, otologists should pay attention to avoid missed or incorrect diagnosis in hemangiomas of facial nerve.

Key Words Facial nerve; hemangioma; Facial palsy

Introduction

Facial nerve hemangiomas are rare benign vascular tumors which grow slowly and are reported to account for 0.7% of intratemporal tumors and 18% of facial nerve tumors, although their pathogenesis is not completely understood. A new research proposes to reclassify "venous vascular malformations of the facial nerve" instead of "hemangiomas" to be consistent with current terminology.

Facial nerve hemangiomas can arise from any segment of the facial nerve. Pulec first described a hemangioma of the geniculate ganglion in 1969. Facial nerve hemangiomas originate from the venous plexus surrounding the facial nerve. The most common site of occurrence in facial nerve hemangiomas is the geniculate fossa, followed by the internal auditory canal and less frequently, the posterior genu of the facial nerve. As a result of the intense distribution of the capillary plexus in geniculate fossa, internal auditory canal and the vertical section of the facial nerve, geniculate ganglion becomes the most frequent site of the occurrence.

Symptoms

Facial nerve hemangiomas are unusual to be reported especially in China. All of the reported cases complain of facial nerve palsy as the first symptom with or without auditory dysfunction. Even when very small, the tumors can cause weakness of facial nerve function, indicating that the facial nerve fibers are invaded by the tumor. Patients’ facial nerve palsy can be sudden, recurrent or associated with hemifacial spasm. Symptoms often progress for years before the diagnosis is made. Other symptoms include conductive hearing loss, otalgia, pulsatile tinnitus, aural bleeding, vertigo and presence of a polypoid mass in the external auditory canal. The pulsatile tinnitus and vertigo are rare and can be caused by angiomatous invasion of the cochlea or vestibular nerve compression. Conductive hearing loss is caused by tumor extension into the middle ear. An extensive review shows that only 6% of the hearing loss in hemangiomas occur in the geniculate ganglion.

Diagnosis

Other than clinical symptoms and history, gadolinium–enhanced MRI and preoperative high–resolution CT imaging are necessary for timely diagnosis of such lesions. In addition to its ability to demonstrate pathological changes in the nerve, MRI is useful to rule out other causes of facial nerve palsy, such as tumors of parotid or other intracranial lesions. The high–resolution CT of the temporal bone may be of great help in differentiating hemangiomas from other disease entities and may also provide important
anatomical information at the time of surgery.

On CT imaging, hemangiomas show somewhat irregular borders with intrallesional stippled calcifications (Fig. 1). On MRI, they exhibit heterogenous hyperintensity on T2-weighted imaging (Fig. 2.). The differential diagnosis based on the MRI characteristics of the lesion include normal intratemporal facial nerve enhancement, Bell’s palsy, Hunt Syndrome, schwannoma of the facial nerve, and perineural spread of parotid malignancy along the facial nerve.

Fig.1 Axial CT scan of a left temporal bone showing a lesion involving the GG with irregular borders and spicules of bone.

Fig.2 Gadolinium–enhanced MRI images showing a centrally enhancing lesion in the GG, measuring 5mm×10mm in diameter.

Because facial nerve hemangiomas are rare and unfamiliar to most of otologists and symptoms lack specificity, they are easily misdiagnosed or misdiagnosed.

Misdiagnosed as Bell’s palsy

When they are misdiagnosed as Bell’s palsy, steroid therapy has a significant effect on these patients, and it’s possible that steroids can cause shrinking of the tumor. Recurrent facial paralysis is presented when fibrosis occurs within the tumor, and subsequent steroid therapy in these patients has poor results. So the presence of facial nerve function deficit, especially if progressive, persistent or recurrent, should always arouse suspicion of a facial nerve lesion.

Misdiagnosed as otitis media

It is noteworthy that the facial nerve tumors associated with ear symptoms are easily misdiagnosed as middle ear diseases, especially acute otitis media.

Misdiagnosed as external auditory cholesteatoma

Misdiagnosed as glomus nodular tumor

Other differential diagnoses include facial nerve schwannomas, meningiomas, cholesteatomas, and metastatic tumors. Facial nerve hemangiomas can also be mistaken for other vascular masses such as glomus tumour or ectopic tissues of the middle ear. Unfortunately, clinical and radiological evaluation may be insufficient to make a preoperative diagnosis and the diagnosis is often based upon intraoperative biopsy specimen findings and postoperative pathological test results.

Treatment

The most accepted treatment of facial nerve hemangiomas is surgical excision. The type of approach depends on tumor location, preoperative hearing level and tumor size. Because the tumor is extraneural, it is possible to remove the tumor and preserve the facial nerve in some instances. However, it is not possible to preserve the facial nerve in cases with direct nerve infiltration.

Small hemangiomas produce less compression of the nerve, less inflammatory reaction and less fibrosis, which makes it possible for surgical tumor removal while preserving nerve function.

Large hemangiomas are closely attached to the facial nerve, which makes it very difficult to separate the tumor from the nerve. Thus it is often necessary to remove the nerve and repair it by end to end primary anastomosis or interposition with a graft. There are some contradictory articles on the management of these injuries, and the timing of surgery is one of the most controversial. The optimal outcome of a reconstruction with end to end primary anastomosis or interposition with a graft can yield a result better than grade III on the House–Brackmann Scale (H–B I to VI, with I being normal and VI representing complete facial paralysis).

Not all patients show good results of facial function
after reparation. However, most surgeons think that timely diagnosis and surgical resection lead to better postoperative facial function, and early excision of the tumor with preservation of the nerve is the most recommended. Recurrence of facial nerve hemangiomas is rare after complete or partial excision.

Conclusion

Facial nerve hemangiomas are rare but may result in facial palsy independent of their sizes. Isolated facial palsy in a person should be monitored closely using imaging studies even if the initial imaging study is negative. Early decompression of the facial nerve may help preserve its function. Despite the rarity of facial nerve hemangiomas, the presence of a facial nerve function deficit, especially if progressive, persistent, or recurrent, should always arouse suspicion of a facial nerve lesion. For a patient with facial palsy who only partially recovers within six months or a patient with recurrent facial palsy, CT scan and MRI should be ordered to rule out space-occupying lesions along the facial nerve. Accurate and timely diagnosis can help the patient alleviate suffering as far as possible. Surgical treatments should be individualized for each particular patient.

References


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