Facial Nerve Hemangioma: A Rare Case Involving the Vertical Segment

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ABSTRACT

Objective:
Report a rare case of facial nerve hemangioma (FNH) involving the vertical FN segment and to discuss the clinical presentation, imaging studies, and management of these rare lesions.

Methods:
A case-report and review-of-literature

Study Retrospective-review

Results:
A 53-year-old man presented with a 10-year history of right hemifacial twitching and progressive facial paresis (House-Brackmann grading score-V/VI). The computed-tomography (CT) and Magnetic-resonance-imaging (MRI) studies confirmed an expansile lesion along the vertical FN segment. Excision and histopathologic examination demonstrated FNH.

Conclusions:
FNHs involving the vertical FN segment are extremely rare. Despite being rare lesions, we believe that familiarity with the presentation and management of FNHs are imperative.

INTRODUCTION

Facial nerve hemangiomas (FNHs) account for 0.7% of all intratemporal tumors. These lesions are benign extraneural tumors that arise from the vascular plexuses surrounding the facial nerve (FN). It is thought that facial weakness is caused either by direct compression of the nerve or the phenomenon of “vascular steal.” In the latter, the FN is deprived of blood supply causing ischemia and weakness.

The geniculate ganglion (GG) is the most commonly involved segment of the FN. As demonstrated in human temporal bone studies by Balkany et al, this high predilection for the GG is likely due to the presence of high perineurial capillary count at the GG compared to other FN segments. The goal of this study is to report a rare case of FNH involving the vertical FN segment and to discuss the clinical presentation, imaging, and management of these rare lesions.

CASE REPORT

A 53-year-old man presented with a 10-year history of right hemifacial twitching. Over the past 6 years, the hemifacial twitching led to right-sided facial weakness, which he described as “difficulty closing his mouth.”

Physical examination was significant for incomplete right eye closure and asymmetry of the right face at rest. His right facial House-Brackmann (HB) score was V/VI. Bilateral ear exams were normal. Audiogram revealed bilateral symmetric high frequency sensorineural hearing loss. The speech reception thresholds and word discrimination scores were normal. CT demonstrated a 10 × 7 × 3 mm bony expansile lesion along the vertical segment of the facial nerve (Figure 1). The lesion expanded up to the jugular bulb medially and expanded the right stylomastoid foramen. On MRI, the lesion had hyperintense MR signal on T2-weighted sequence and enhanced after gadolinium administration on T1-weighted sequence (Figure 2).

The patient underwent a transmastoid and transcervical approach for the excision of the lesion involving the vertical segment of the facial nerve from the outer genu to the stylomastoid foramen, followed by facial nerve reconstruction with greater auricular cable graft. Hematoxylin–eosin staining revealed multiple vascular channels lined with single endothelium surrounded with loose connective tissue (Figures 3). The final diagnosis was consistent with a facial nerve hemangioma.

DISCUSSION

CLINICAL FEATURES:
• The clinical presentation of FNHs varies depending on tumor location.
• Tumors located in the GG most commonly present with FN dysfunction (97%). Hearing loss is a less common presentation occurring in only 6% of the cases. This can be conductive (tumor extension into middle ear) or sensorineural (cochlear fistula formation) in nature.
• Tumors involving the IAC most commonly present with progressive sensorineural hearing loss (90%). Facial nerve dysfunction in these cases occurs less commonly (71%).
• Clinical presentation in patients with vertical segment involvement is variable and consists of progressive FN dysfunction (unlike GG FNHs), and conductive hearing loss, (unlike SNHL in IAC FNHs).

IMAGING STUDIES:
• On CT, FNHs typically present as poorly defined lesions with indistinct bony erosion. Benign bony spicules representing calcification of collagen containing walls of the vascular channels can sometimes give the characteristic “honeycomb” appearance.
• On MRI, FNHs have variable T1-weighted intensities, are hyperintense on T2-weighted sequence, and enhance uniformly after gadolinium administration.
• Since FN dysfunction occurs at very early stages, it is suggested that an IAC lesion measuring less than 10mm in a patient with FN dysfunction is highly indicative of FNH.

TREATMENT:
• The treatment of choice continues to be surgical excision.
• Tumors located at the GG are best approached with a combined transmastoid and middle fossa approach and those located at the vertical segment can be removed using a transmastoid approach only.
• Tumors located in the IAC can be excised using a middle fossa or translabyrinthine approach if there is serviceable or non-serviceable hearing, respectively.
• The timing of surgical excision is controversial. Eby et al. advocate tumor removal within 1 year of onset of progressive facial paralysis if optimal postoperative FN results is to be expected.
• On the other hand, Issacon et al. advocate judicious observation: “delayed surgery may not compromise facial nerve function,” however, “patients with prolonged duration of symptoms may have higher rate of hearing loss than those who undergo surgery early.”

CONCLUSIONS

• FNHs are benign extraneural tumors that arise from the vascular plexuses surrounding the FN.
• Lesions involving the vertical FN segment are extremely rare, and their clinical presentations can be more variable than the FNHs more commonly found in the GG or the IAC.
• Despite being rare lesions, we believe that familiarity with the presentation and management of FNHs are imperative.

REFERENCES