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Skull base plexiform Schwannoma of possible chorda tympani nerve origin in a three-year-old patient☆

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1. Introduction

Plexiform schwannomas (PS) are rare variants of Schwann cell tumors that are characterized by a plexiform pattern of intraneural growth that is often accompanied by a multinodular pattern. Such schwannomas are rarely associated with conditions such as neurofibromatosis type I. A subset of PS can affect superficial soft tissues of the head and neck [1]. Schwannomas originating from the chorda tympani nerve are particularly rare tumors with less than two dozen cases reported to date [2].

2. Case report

A 3-year-old male with no significant past medical history presented with a 6-month history of progressive left pre- and retro-auricular swelling. Following multiple inconclusive ultrasound findings and failure of the swelling to resolve, the patient's family made the decision to pursue fine needle biopsy of the area, which was consistent with schwannoma. The patient was then referred to our tertiary referral center for further evaluation.

Physical examination was notable for firm swelling at the left tail of the parotid area which extended to the postauricular region, with normal facial nerve function pre-operatively. The patient and family denied any other symptoms or stigmata of neurofibromatosis. Given no reported hearing changes reported preoperatively, no formal preoperative audiogram was performed.

Preoperative imaging demonstrated a soft tissue mass centered in the left parotid region with deep extension into the left parapharyngeal space and superior postauricular extension into the region overlying the mastoid air cells. There was bony remodeling of the mastoid cortex and posterior ear canal wall on computed tomography (CT) (Fig. 1). The lesion appeared to widen the facial recess without involvement of the Fallopian canal. On magnetic resonance imaging (MRI), the mass measured 3.7× 4.2 cm in the axial plane and up to 6 cm in the craniocaudal dimension.

Following discussion in our multidisciplinary tumor board, the family elected for an open parotid and transmastoid surgical resection. Intraoperative findings included superior extension of the tumor into the postauricular crease and significant thinning of the cortical mastoid bone. The tumor extended through the posterior ear canal wall and was adherent to the posterior ear canal skin. To fully remove the tumor, the mastoid tip, part of the posterior canal wall skin, and tympanic ring were resected. The facial recess was widened secondary to the mass (Fig. 2). Notably, the tumor appeared to originate from the chorda tympani nerve near its take off from the facial nerve. This finding was supported by inspecting the middle ear space and noting that the chorda tympani

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nerve moved when the mass was pulled inferiorly. The mass was closely aligned with the main trunk of the facial nerve but did not appear to originate from the facial nerve, which was fully delineated in the vertical segment, decompressed down through the stylomastoid foramen, and followed extratemporally via continued parotid dissection. Following total tumor removal, the ear canal defect was reconstructed with temporalis fascia and a free cortical mastoid bone graft, with non-absorbable canal packing placed. At the end of the procedure, the left facial nerve was confirmed to be intact anatomically and physiologically with stimulation. No other lesions were noted intraoperatively.

Surgical pathology confirmed a 7.5 cm PS (Fig. 3). Hematoxylin and eosin (H&E) staining demonstrated a circumscribed spindle cell proliferation with a plexiform growth pattern and immunohistochemistry (IHC) staining was diffusely positive for S100 and SOX10, confirming schwannoma (Fig. 4).

There were no acute postoperative complications. Follow-up demonstrated no cranial nerve deficits but was complicated by delayed stenosis of the external auditory canal, which ultimately resolved following stent placement. A three-month postoperative CT scan showed no residual tumor, and postoperative audiogram showed normal hearing thresholds and tympanometry.

3. Discussion

We report one of the youngest cases of PS and, to our knowledge, the first to possibly originate from the chorda tympani nerve. Chorda tympani schwannomas can present with a variety of signs and symptoms related to the involvement of the chorda tympani nerve and adjacent structures in the middle ear. Historically, patients have presented with a mixed or conductive hearing loss. However, symptoms such as tinnitus, otorrhea, earache, or facial paresis due to the tumor's proximity to the facial nerve have also been reported [2–4]. Interestingly, the growth pattern of this patient's lesion extended inferiorly and anteriorly into the deep parotid space, ultimately presenting as a facial mass.

Tumors arising from the chorda tympani nerve pose unique diagnostic and therapeutic challenges due to their intimate relationship with the facial nerve. High-resolution imaging techniques are crucial for precise tumor localization, assessment of size and extension, and identification of the relationship with surrounding structures. Confirmation of a PS is achieved through histopathological examination of biopsied or resected tumor tissue, though it should be noted there are no pathological tests which can reliably characterize the nerve of origin. Therefore, surgical visualization of the nervous origin of the tumor is critical.

Surgical resection is the primary treatment modality of skull base PS and should aim to achieve complete tumor removal while preserving cranial nerve function and minimizing postoperative complications. The surgical approach may vary based on tumor characteristics and the surgeon's expertise, with endoscopic approaches reported to be successful in some cases [3–5]. In this patient, the tumor was adherent to the posterior external canal wall skin and its extension into the facial recess required a careful approach with removal of the mastoid tip and resection of part of the posterior canal skin to allow for complete resection.

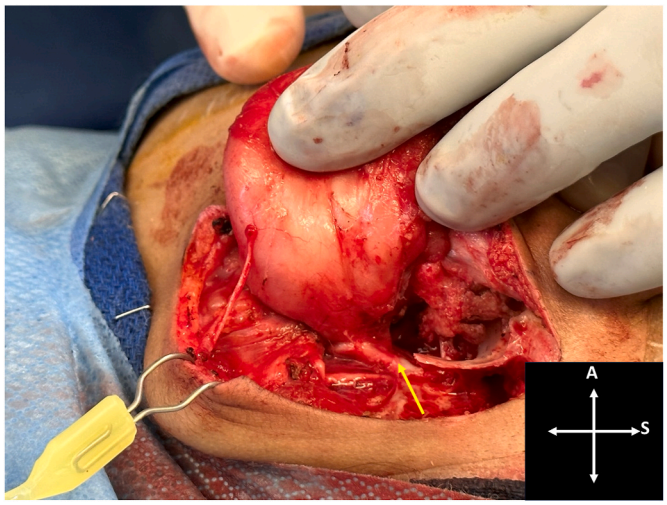

Fig. 2. Intraoperative view. Left postauricular approach showing the tumor being dissected away from the facial nerve (yellow arrow) and the associated mastoidectomy defect with facial recess dissection. Figure legend demonstrates anterior (A) and superior (S) orientation. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

4. Conclusion

This case is the first report of PS of apparent chorda tympani nerve origin in a pediatric patient. PS in young children are exceedingly rare tumors, and schwannomas of the chorda tympani nerve even more so with less than two dozen reports to date in the English literature. There are no pathognomonic signs or symptoms of PS of the chorda tympani nerve. Accurate diagnosis relies on high-resolution imaging and histopathologic evaluation. Surgical resection appears to be the standard of care, with the choice of surgical approach depending on tumor and patient-specific characteristics.

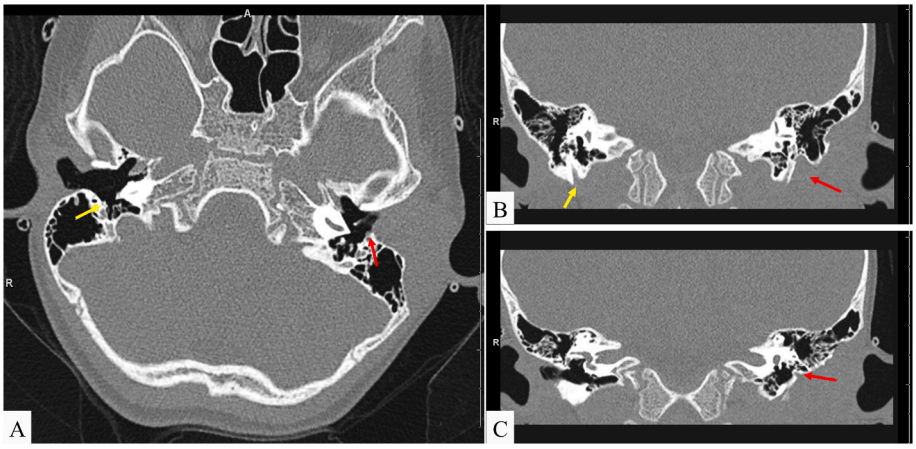

Fig. 1. (a) Axial, and (b, c) Coronal CT demonstrating cortical bone remodeling from the lesion and its relationship to the vertical segment of the facial nerve (red arrows). Yellow arrows represent the normal contralateral side for comparison.

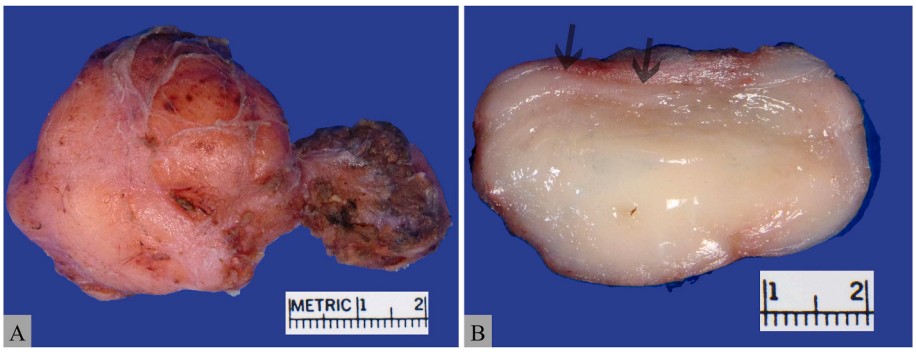

Fig. 3. Gross pathology. (a) Outer surface of lesion, well circumscribed and surrounded by a thin fibrous plane/capsule. (b) Cut surface of lesion with a white-tan and glistening appearance, as well as possible plexiform bundle (arrows).

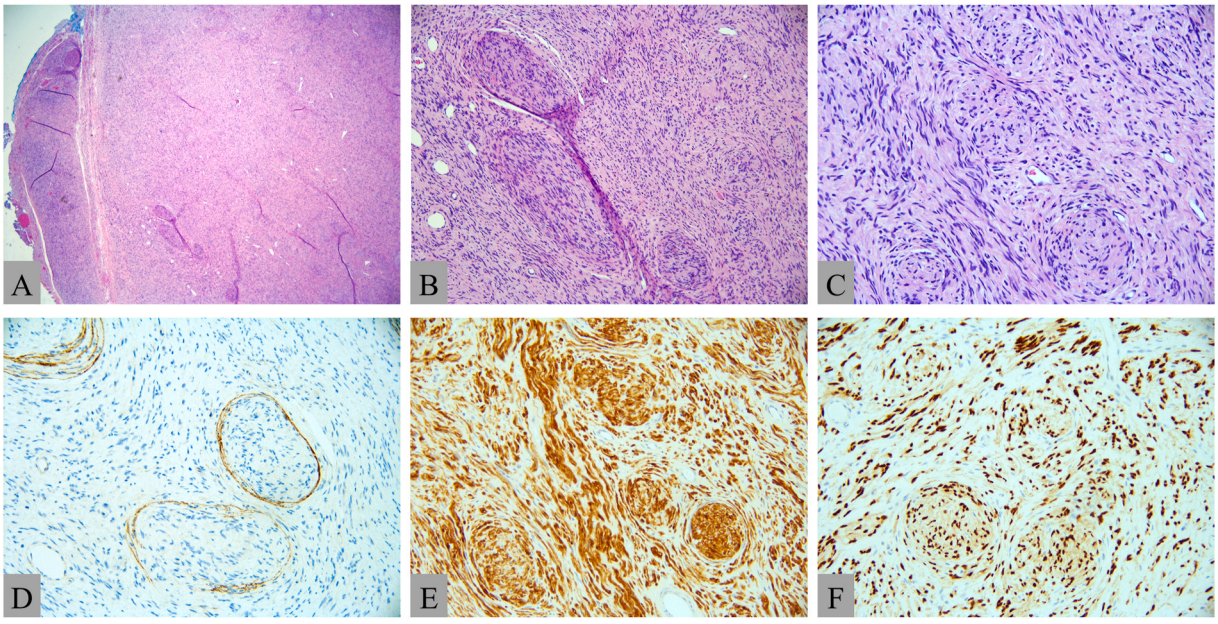

Fig. 4. Histopathology. (a) 20x H&E staining demonstrating a circumscribed spindle cell proliferation with a plexiform growth pattern and tumor bundle at left periphery. (b, c) 100x and 200x H&E staining, respectively, demonstrating smaller intralesional bundles consistent with plexiform growth pattern. (d) Immunohistochemistry (IHC) staining for EMA highlighting perineurium of intralesional bundles. (e, f) IHC for S100 and SOX10, respectively, consistent with schwannoma.

Disclosure statement

The authors have no relevant grants or funding from funding agencies in the public, commercial, or not-for-profit sectors to report.

Ethical statement

This case report adheres to the ethical guidelines of *Otolaryngology Case Reports* and the University of Chicago IRB. This case report has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki). The manuscript is in line with the Recommendations for the Conduct, Reporting, Editing and Publication of Scholarly Work in Medical Journals and aims for the inclusion of representative human populations (sex, age and ethnicity) as per those recommendations. The terms sex and gender are used correctly. Informed consent was obtained from the patient described in the case presentation and identifying or personal health characteristics were omitted.

CRediT authorship contribution statement

Nihar Rama: Writing – review & editing, Writing – original draft, Visualization, Formal analysis, Data curation. **Aaron Wallace:** Writing – review & editing, Data curation. **Maximilian Hemmrich:** Writing – review & editing, Data curation. **Nicole A. Cipriani:** Writing – review & editing, Formal analysis, Data curation. **Olga Pasternak-Wise:** Writing – review & editing, Formal analysis, Data curation. **Nishant Agrawal:** Writing – review & editing, Formal analysis, Data curation. **Terence Imbery:** Writing – review & editing, Supervision, Formal analysis, Data curation, Conceptualization.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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