Facial Nerve Duplication and First Branchial Cleft Cysts: An Association in an Uncommon Pathology

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First branchial cleft cysts (BCCs) represent an uncommon cause of pediatric head and neck masses and can manifest as fistulae and cysts in the region between the external auditory canal (EAC) and the level of the hyoid bone.\textsuperscript{1} First BCCs are divided based on relationship to the facial nerve: work type I lesions are lateral, and work type II lesions are medial.\textsuperscript{1} Although facial nerve aberrancy is not uncommon in patients with inner ear malformations, there is only 1 report of duplication of the extratemporal main trunk of the facial nerve, also occurring in a patient with type II first BCC.\textsuperscript{2,3} We report a second case, suggesting that there is an association between first BCCs and duplication of the facial nerve trunk. The Ann & Robert H. Lurie Hospital of Chicago Institutional Review Board approved this research.

Case Report

A 16-year-old male presented to the otolaryngology clinic with bilateral neck masses. He was found to have bilateral auricular fistulae posterior to his lobules. They fluctuated in size, and the left-sided lesion became infected frequently. First BCCs were suspected, and a contrasted magnetic resonance image (MRI) of the neck was obtained, confirming bilateral cystic lesions closely associated with the parotid glands (Figure 1). Because he experienced frequent infections, staged bilateral excision was recommended. The left side was the most symptomatic, so it was addressed first. Parotidectomy with complete excision of the BCC has been the mainstay of treatment in our practice to help prevent recurrence. One series noted up to 50% recurrence with limited treatment of BCC without parotidectomy.\textsuperscript{4} A modified Blair incision was made, and the fistula was excised in continuity with the lesion with continuous facial nerve monitoring. A 2-mm nerve was identified emerging from the stylomastoid foramen, which was found to branch once into the temporal and zygomatic branches and activated the orbicularis oculi muscle when stimulated. Inferiorly, a second facial nerve trunk was then identified that branched into buccal, marginal mandibular, and cervical divisions. When stimulated, the orbicularis oris muscle was activated. There was a connection between the 2 main trunks proximal to the first division of both branches. Both nerve branches were dissected back to the stylomastoid foramen and were seen to be exiting separately (Figure 2). The BCC itself was inferior to the lobule, with its tract passing between the duplicated facial nerve trunks in the same plane. The patient had full facial nerve function (House-Brackmann I) postoperatively. Pathology showed a first BCC with surrounding chronic inflammation.

Discussion

Duplications of segments of the facial nerve are rare anatomical variations. Bifurcation anomalies of the facial nerve

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A duplicated facial nerve trunk has been reported for all segments of the intratemporal facial nerve.\textsuperscript{2} We have called the malformation a duplication due to convention, although it also accurate to think of it as a bifurcation within the temporal bone. There has been only 1 previously described extratemporal facial nerve trunk duplication, which was also noted in the setting of a first BCC.\textsuperscript{3} The facial nerve trunk is largely formed by the end of the eighth week, while the mesenchymally derived tissue that it passes through and the muscles it innervates are largely unformed, implying that bifurcation of the nerve may require an error early in fetal life.\textsuperscript{2} First BCC cysts are thought to be duplications of the EAC, as noted in the original Work\textsuperscript{5} classification. We propose that this duplication phenomenon may also affect the mesenchymal elements of the EAC and second branchial arch, thus leading to duplication of the facial nerve trunk, supported by the fact that the BCC tract extended between the separate facial nerve trunks in this patient.

This represents the second case of a duplicated facial nerve trunk, both of which have occurred in type II first BCC. While the exact embryologic mechanism for facial nerve duplication is unclear, this correlation suggests that it may be related to the same process as first BCC formation. Surgeons operating on patients with a presumed type II first BCC must be aware of this association, as careful dissection of the presumed facial nerve trunk should be done from the stylomastoid foramen to the pes anserinus to verify that there is not a duplication and to avoid injury.

**Author Contributions**

Jacob Eide, design, draft, and revision of the manuscript; André Isaac, design, draft, and revision of the manuscript; John Maddalozzo, design, draft, and revision of the manuscript.

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