An Unusual Neck Mass in a Child

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A 14-year-old boy presented with a 3-month history of a left-sided parotid mass. It had increased in size over 6 months. The mass was nontender, mobile, and without any cutaneous changes. The head and neck examination, including facial nerve function, was unremarkable.

A contrast-enhanced computed tomography (CT) scan revealed a heterogeneous encapsulated lesion (3 cm × 4 cm) with cystic areas within the left parotid gland. A fine needle aspiration (FNA) biopsy revealed irregular lobules and cords composed of fine fibrillar matter, with scattered round and spindled stromal cells.

Following a superficial parotidectomy, histopathological examination showed a 4.5 cm × 3.1 cm × 2.5 cm mass. Sections demonstrated a well-circumscribed lesion composed of spindled cells predominantly arranged in cords. A rim of salivary glandular tissue was present all around the mass. An immunohistochemical stain for S100 showed diffuse cytoplasmic and nuclear positivity. A diagnosis of schwannoma was made.

Schwannomas are uncommon tumors that arise from cranial, peripheral, or autonomic nerves.¹ Extracranial schwannomas have a predilection for the scalp, face, and the external auditory canal, accounting for about 25% to 40% of all head and neck nonvestibular schwannomas.² They usually present gradually and thus are often diagnosed incorrectly. However, better imaging and cytological techniques have enabled earlier and more definitive diagnosis.

Schwannomas of the parotid gland are exceedingly rare in children. For example, a review of 324 consecutive pediatric salivary gland masses revealed that the most common benign tumor was a hemangioma, accounting for about 60%, followed by lymphovascular malformations representing another 30%.³ Only one schwannoma was seen in this case series. There are few other sporadic reports of nonvestibular schwannomas within the head and neck in children.⁴

In addition to facilitating diagnosis, preoperative imaging facilitates delineation of the tumor from surrounding structures. On magnetic resonance imaging, schwannomas are generally isointense on T1-weighted images relative to skeletal muscle with increased and slightly heterogeneous signal on T2-weighted images.⁵ On CT scans (Figure 1A), schwannomas appear well circumscribed and fusiform with relatively common contrast enhancement. Internal cystic changes are seen with larger tumors, which represent mucinous degeneration, hemorrhage, necrosis, and microcyst formation.⁶

An FNA of the tumor may provide further information to differentiate between benign and malignant causes. However, previous reports have identified specific difficulties associated with interpretation of cytological specimens owing to cystic areas within the tumor and hypocellular specimens.⁷ Satisfactory needle aspirates characteristically consist of spindle cells arranged haphazardly in irregular tissue fragments and in parallel as elongated rasty fascicles, with a myxoid to fibrillar background (Figure 1B). Definitive diagnosis is therefore achieved only by excisional biopsy, which in this instance necessitated a superficial parotidectomy. This was performed with dissection and preservation of the facial nerve. The patient was subsequently discharged home uneventfully.

Histological sections of the specimen demonstrated a well-circumscribed lesion composed of spindled cells predominantly arranged in cords. A high-power image showed extensive Verocay body formation (Figure 1C). An immunohistochemical stain for S100 (Figure 1D) displayed diffuse cytoplasmic and nuclear positivity consistent with the nerve sheath origin of the tumor.⁸

Antoni A and B are cytarchitectural patterns that typify histopathologic diagnosis of schwannomas.³ Type A areas are highly cellular and demonstrate nuclear palisading and associated Verocay bodies, which manifest from prominent extracellular matrix and the secretion of laminin. Type B areas on the other hand are sparsely populated—with myxomatous and cystic changes—and thus may represent degenerated Antoni A tissue.

Schwannomas are benign tumors that respond to meticulous surgical excision with very low rates of recurrence. Postoperative morbidity arises from nerve injury.⁹ Malignant transformation of these tumors is rare and is best treated with wide excision where possible. The role of adjuvant therapy is uncertain; irrespective of the treatment approach, the prognosis, when malignant transformation occurs, is poor with an overall survival of 15%.²

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As detailed above, schwannomas are exceptionally rare benign head and neck tumors in children. They may cause diagnostic dilemmas, which are resolved by characteristic imaging and histopathologic features. Schwannomas should be considered in the differential diagnosis of salivary gland tumors that have (1) atypical appearance on imaging and (2) spindle cells seen in histopathologic examination.

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