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Case Report

Intraparotid facial nerve schwannoma in a 12-year-old child. Report of a case and review of the literature

Panagiotis Saravakos ^{a,*}, Nektarios Papapetropoulos ^b, Stephanie Vgenopoulou ^c, Eleftherios Karamatzanis ^b, Konstantinos Saravakos ^b

^a Department of Otolaryngology, Head and Neck Surgery, Siloah St. Trudpert Hospital, Pforzheim, Germany

^b Department of Otolaryngology, Head and Neck Surgery, Penteli Children Hospital, Athens, Greece

^c Department of Pathology, Sismanoglio Hospital, Athens, Greece

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ABSTRACT

Schwannomas are benign tumors originating from Schwann cells. The appearance of these tumors in the intraparotid segment of the facial nerve is rare and is observed almost exclusively in adults. We report a case of an intraparotid facial nerve schwannoma in a 12-year-old patient, who presented with a well-defined parotid enlargement and normal facial nerve function. Surgical intervention consisted of parotidectomy with preservation of the facial nerve. A review of the literature is reported.

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1. Introduction

Schwannomas or neurilemomas are benign, generally slowgrowing neuroectodermal neoplasms that originate from Schwann cells. Schwannomas of the seventh cranial nerve can originate from any site along the course of the facial nerve, from the glial-Schwann cell junction at the cerebellopontine angle to the terminal branches in the parotid gland [1]. The majority of facial nerve schwannomas originate in the labyrinthine and tympanic segments of the nerve, while an extratemporal localization in the parotid gland is less common, accounting for approximately 9-10% of all facial nerve schwannomas [2,3]. Intraparotid schwannomas usually affect adults and, to the best of our knowledge, only two pediatric cases have been reported worldwide to date [4,5]. We report a third case of a facial nerve schwannoma in a 12-year-old boy. A preoperative diagnosis of this type of tumor is difficult, and management in the case of the intraoperative suspicion of facial nerve schwannoma is controversial.

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2. Case report

A 12-year-old boy presented with a 9-month history of a slowgrowing, painless mass over the right mandibular angle. Physical examination revealed a well-defined, approximately 2.5 cm \times 2 cm sized, slightly mobile mass. Facial and other cranial nerve examinations revealed no abnormalities. The medical, surgical and family histories of this patient were unremarkable.

Sonographic scan showed a mass with a hypo-/isoechoic pattern, well-defined margins, homogenous echogenicity and acoustic enhancement originating from the superficial parotid lobe. Preoperative magnetic resonance imaging (MRI) examination revealed a 2.5-cm sized parotid tumor. The mass showed a low-intermediate signal density and was isodense to muscle on T1-weighted images, whereas it had a high signal density on T2-weighted images (Fig. 1). After the administration of contrast material, the lesion showed homogeneous enhancement. It should be noted, that the MRI findings were suggestive of a pleomorphic adenoma.

The initial tumor was excised by superficial parotidectomy. Intraoperatively, the mass was well encapsulated and was connected to the mandibular branch of the facial nerve (Fig. 2). The mass did not react with electrical stimuli, as determined using a nerve simulator. The tumor was excised, and the nerve trunk and the main branches of the facial nerve were preserved. Histopathological examination confirmed the diagnosis of a benign

^{*} Corresponding author at: Department of Otolaryngology-Head and Neck Surgery, Siloah St. Trudpert Hospital, Wolfsbergallee 50, Pforzheim DE-75177, Germany.

E-mail address: psaravakos@yahoo.com (P. Saravakos).

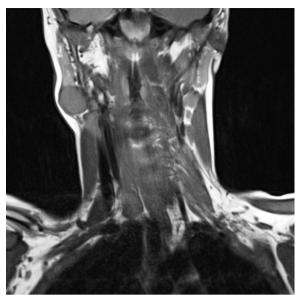


Fig. 1. T1-weighted MRI examination showing the tumor located in the right parotid gland with a hypointense pattern.



Fig. 2. Encapsulated tumor excised from the right parotid gland without facial nerve injury.

schwannoma. Microscopically, the tumor consisted almost exclusively of cellular, palisading spindle cells arranged in an organoid pattern (Verocay bodies) with immunohistochemical expression of the S-100, vimentin and CD68 markers (Figs. 3 and 4). Postoperatively, the patient developed a House–Brackmann (HB) grade II facial weakness that spontaneously resolved after 3 months. During the follow-up examination 1 year after the operation, the facial nerve function was evaluated as HB grade I. No recurrence was noted.

3. Discussion

Salivary gland pathology in children differs greatly from that in the adult group. Most salivary gland lesions in children are developmental (such as hemangiomas), inflammatory or infectious. Salivary gland neoplasms are rare, comprising approximately 5–6% of cancers of the head and neck and 0.3% of all cancers, but in children they account for about 10% of all childhood neoplasms. Malignant parotid lesions are uncommon in the pediatric population, but, when a parotid mass is neoplastic, it has a 30– 60% probability of malignancy. In children, the most common

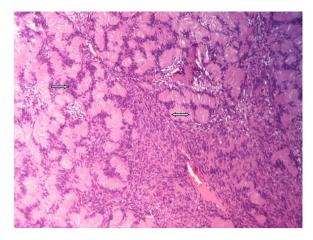


Fig. 3. Characteristic basaloid pattern of dark-staining Schwann cell nuclei (simple arrow) around hyaline stroma (double arrow) in an organoid arrangement; H-EX100.

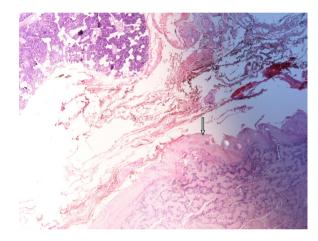


Fig. 4. Salivary gland tissue (upper left) in continuity with the benign encapsulated schwannoma (lower right); H-EX40.

benign epithelial tumor is pleomorphic adenoma, and the most common malignant tumor is mucoepidermoid carcinoma (60– 90%). Adenoid cystic and acinic cell carcinomas follow in frequency. The majority of salivary gland tumors in children occur in the major glands (76.7%). The ratio of occurrence of parotid to submandibular to sublingual tumors in the major salivary glands is 30:6:1. These tumors occur predominantly in girls and at any childhood age. Imaging techniques, such as ultrasonography, CT or MRI may narrow the differential diagnosis, while incisional biopsy should be avoided due to the possibility of tumor spillage and facial nerve damage. Table 1 summarizes the commonest salivary gland neoplasms in children [6,7].

Mesenchymal tumors of the salivary glands can originate mainly from the neural, fibrous or muscular tissue. Benign

Table 1

Common salivary gland tumors in children.

Salivary gland neoplasms in children	
Benign tumors	Pleomorphic adenoma
	Warthin's tumor
Malignant tumors	Mucoepidermoid carcinoma
	Adenoid cystic carcinoma
	Undifferentiated carcinoma
	Cancer originating from alveolar cells
	Sarcoma

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