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Case Report
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Intraparotid Facial Nerve Schwannoma in a One-Year-Old Patient

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Introduction

Schwannomas or neurilemomas are benign, generally slow-growing, encapsulated tumors that arise from Schwann cells of myelinated axonal nerve [1]. Approximately twenty-five percent of all schwannomas are described in the head and neck topography [2]. Rarely arise from the facial nerve and most commonly occur in the intratemporal part of the nerve. There are only a few cases reported in the literature of extratemporal facial nerve schwannomas which is manifested by a painless mass in the parotid gland region and can be mistaken for a parotid tumor [3]. The following case is still rarer as it involves an extratemporal facial nerve schwannoma in a pediatric patient.

Case Presentation

A 1-year-old female patient, M.L.V.F., presented at the Head and Neck surgery department in Barretos Cancer Hospital, Brazil, in July 2019, with a slow-growing and progressive right-sided facial mass of 6-months duration. Physical examination showed a mobile mass of softened consistency, approximately 4 cm in the largest diameter in size, in the right parotid gland. There was no facial nerve dysfunction.

A preoperative Computed Tomography scan (CT scan) was obtained and identified an expansive soft tissue formation, at the right parotid region, that medially sloped and compressed the parotid gland on this side, with no signs of adjacent bone invasion. CT scan detected a 4.4 cm × 3.2 cm tumor with well-defined and regular borders, heterogeneous enhancement and hypoattenuating central area suggesting necrosis and liquefaction.

Core-biopsy with immunohistochemical study of the tumor showed a diffuse and significant expression of S-100 and SOX-10 protein and was positive for others biomarkers including BCL2, EMA, TLE-1, CD99, INI-1, CD-56, CD57, suggesting the diagnosis of peripheral nerve neoplasm (Schwannoma).

Due to the diagnostic hypothesis of facial nerve schwannoma in a one-year old child, a close follow-up was recommended. During the follow-up period, the tumor almost doubled in size. Preoperative Magnetic Resonance Imaging (MRI) of head and neck was obtained 16 months later for surgical planning and demonstrated an expansion of the tumor in size, at this moment measuring 6.5 cm × 5.0 cm × 4.5 cm (Figure 1a, 1b).

The tumor resection surgery was performed on a month later, with intraoperative monitoring of facial nerve to avoid sectioning the nerve or its branches (Figure 2). If necessary, the surgical team was prepared to perform a microsurgery reconstruction of the nerve to avoid facial nerve sacrifice and facial paralysis.

Intraoperatively, Blair incision gave an excellent exposure of the surgical field for dissection of anatomical planes (Figure 3). The facial nerve trunk and its terminal branches including mandibular branch, cervical branch, ophthalmic and frontal branches, were identified, monitored and dissected without nerve damage (Figure 4). Intraoperative detailed dissection showed that the tumor emerged from the zygomatic branch of facial nerve (Figure 5). Therefore, the tumor was resected, without need for microsurgery reconstruction (Figure 6). At the end of the procedure, the facial nerve was tested with intraoperative monitoring and showed normal function (Figure 7).

Anatomopathological and immunohistochemical results confirmed Schwannoma result.

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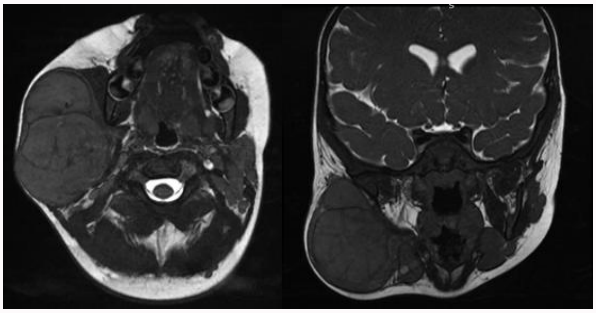

Figures 1A and 1B: Preoperative Magnetic Resonance Imaging (MRI) of head and neck demonstrated an expansion of the tumor in size, measuring 6.5 cm × 5.0 cm × 4.5 cm, axial (1A) and coronal (1B) sections, respectively.

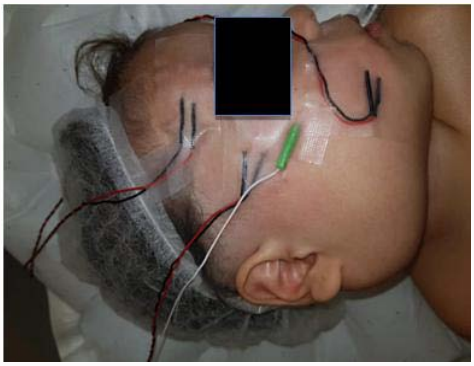

Figure 2: Intraoperative monitoring of the facial nerve.

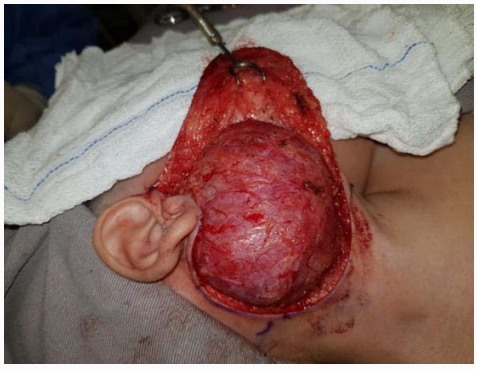

Figure 3: Blair incision wit exposure of the surgical field for dissection of anatomical planes.

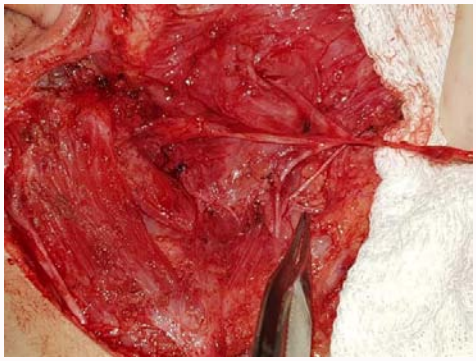

Figure 4: The facial nerve trunk and its terminal branches.

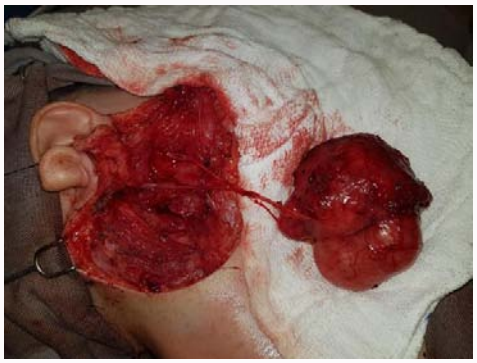

Figure 5: Intraoperative detailed dissection showed that the tumor emerged from the zygomatic branch of facial nerve.

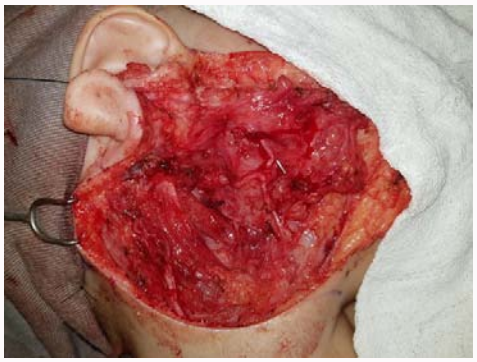

Figure 6: Surgical field after tumor resection, with preservation of the facial nerve and its main branches.

Postoperatively, the patient had a good outcome with normal facial movements and no signs of paralysis. The patient undergoes outpatient follow-up and await further oncogenetics evaluation.

Discussion

Schwannomas are benign tumors with a neuroectodermal origin, arising from Schwann cells of a peripheral or cranial axonal nerve sheath [4]. Head and neck schwannomas represent twenty-five percent of all reported schwannomas [2]. Most schwannomas develop from the eighth cranial nerve, in the vestibular branch.

Solitary schwannoma have been reported in the facial nerve and can be seen anywhere along the course of facial nerve from the cerebellopontine angle to the terminal branches of the medial face. The intratemporal part of the seventh nerve is the most commonly affected. There are a few cases in the literature of extratemporal facial nerve schwannoma. Moreover, schwannomas are very rare in children compared with adults [5].

Classically, schwannomas of the facial nerve appear as palpable solid, mobile, slow growing and painless parotid mass with normal facial nerve functions, but they can grow and compress nervous structures causing facial paralysis [6]. The clinical presentation is generally dependent upon the origin, size and localization of the tumor, the nature of surrounding tissue, and the mass effect imposed on adjacent structures [4,7]. Facial nerve dysfunction occurs in 20% to 45% of extratemporal facial nerve schwannomas while the cases of intratemporal involvement show a higher incidence of facial nerve dysfunction [4].

Magnetic Resonance Imaging (MRI) is the image modality of choice to detect schwannomas and demonstrates the interaction with

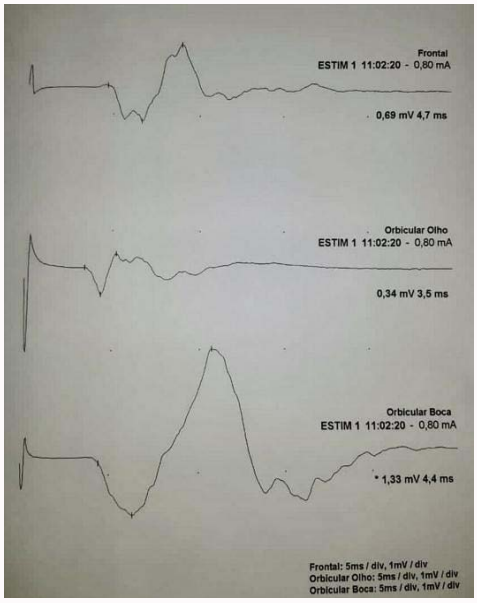

Figure 7: Intraoperative nerve monitoring final test showing facial nerve normal function.

structures surrounding the nerve. Computed Tomography (CT) scan cannot distinguish whether the tumor is a schwannoma or a parotid tumor such as pleomorphic adenoma. Fine needle aspiration cytology is also not diagnostic and in most cases are inconclusive and can lead to a false diagnosis of pleomorphic adenoma. The diagnosis is made mainly intraoperatively [4,8].

In the intraoperative period, the characteristics of the tumor are indicative for the type of neoplasia and its origin, thus the facial nerve schwannoma will be more easily identified. Therefore, intraoperative diagnosis is an important element in decision making [9].

A classification of the intraparotid facial nerve schwannoma was proposed according to its location of origin. In the present case, it would be classified as type B, which means a resectable tumor with only partial sacrifice of the facial nerve, involving one of the peripheral branches or its distal divisions [8].

The literature is controversial regarding the therapy of facial nerve schwannomas. Some authors advocate a wait-and-see policy, while others promote early resection. During the course of a surgical operation, when the tumor can be dissected without damaging the facial nerve or when the tumor affects a branched peripheral nerve, the schwannoma must be resected directly [9,10].

Another characteristic to be considered in facial nerve schwannoma is the difficulty in locating the nerve and its branches during the surgery. Thus, intraoperative monitoring of the facial nerve is used to dissect and preserve the nerve during resection of schwannomas. Studies have shown that monitoring has helped prevent facial nerve damage in about ninety-five percent of patients. However, some patients have no functional recovery and require facial nerve reconstruction or plastic surgery [11]. It is important to highlight the importance of using intraoperative facial nerve monitoring in pediatric surgery, because the nerve structures are smaller and the facial nerve course is more superficial.

Specifically, in pediatric patients, the superficial course of the facial nerve and the absence of surrounding structures are considered main causes of injury to the extratemporal facial nerve in situations such as parotid surgery [12].

Moreover, considering the risk of injury to the facial nerve in children, some anatomical guides can be used for identifying facial nerve trunk such as the anterior border of the sternocleidomastoid muscle. In addition, the parotid gland does not extend posterior to the ramus of the mandible and covers only the lower distal branches of the facial nerve. Because of these anatomical findings in children, the standard techniques for identifying the facial nerve trunk in adults would injure the more superficial located nerve in children [12].

Considering the high risk of injuring facial nerve during resection of the tumor in a pediatric patient, the surgeon should adequately prepare for the possibility of facial nerve sacrifice and discuss rehabilitative options such as nerve graft [13]. According to a study that compared surgical options and outcomes, the better option is the excision of the tumor and involved nerve followed by nerve grafting using the great auricular nerve or the sural nerve [13]. In the present case, there was no need for a reconstructive nerve graft, as the tumor was dissected clearly and affected only the zygomatic branch of the facial nerve. Therefore, the main function of the facial nerve was not affected.

Complete surgical excisions of solitary schwannomas have an excellent prognosis. Pediatric parotid schwannomas can be managed successfully when principles of approach to benign parotid tumors are ensured [14].

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