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Intraparotid facial nerve plexiform neurofibroma: A surgical challenge

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Abstract

Plexiform neurofibromas are benign tumors grow on nerves. It is rare to find these tumors arising from facial nerve within the parotid gland in the head and neck region. These tumors may grow to a considerable size which cause facial disfigurement and pressure symptoms while making resection of such lesion difficult at times due to unclear margins. We report a case of intraparotid plexiform neurofibroma originated from the facial nerve. It was successfully removed by surgery with no recurrence or facial nerve dysfunction after 24 months.

Keywords

benign tumors; nerves; parotid gland; neurofibroma; surgery

Introduction

Neurofibromas represent 0.4% of all salivary gland neoplasms [1]. Plexiform Neurofibromas are benign rare tumors that grow on nerves and usually associated with neurofibromatosis. They are rare occurrences in salivary glands especially when arising from the facial nerve [2,3]. Due to its exceedingly rare nature we present a case of intra-parotid facial nerve neurofibroma in a 5-yr-old boy with its management and follow-up.

Case Presentation

A 5-yr-old boy presented with two years history of painless, slowly growing right neck mass which was not accompanied by fever, dysphagia, night sweating and weight loss. On thorough history no prior medical or surgical problems were identified and no family history of neurofibromatosis. Examination of the neck revealed diffuse right parotid swelling and right upper cervical neck mass measuring 5x4 cm which was firm, non-tender, immobile, and attached to deeper structures. The rest of neck examination was unremarkable for any lymphadenopathy. There were no signs of neurofibromatosis like café-au-lait spots were present over the body. Endoscopic examination of nose and nasopharynx was normal. Chest and abdominal examination were clear and revealed no hepatosplenomegaly which was confirmed by ultrasound. Laboratory investigations and chest x-ray were within normal limits. MRI neck with IV contrast revealed non-enhancing isointense lesion with surrounding soft tissue on T1 weighed MRI image in the parotid gland with extension into parapharyngeal space and which was pressing on the Rt jugular vein. The lesion enhanced post contrast in T1 and on T2 weighted MRI images. (Figure 1; A&B). Superficial parotidectomy was performed and the lesion was found to be arising from tail of the parotid

with close adherence to the cervical and marginal mandibular nerve branch of the facial nerve. The mass was removed from the nerve and all the branches of facial nerve were preserved. Due to technical difficulties, the nerve stimulator was not used. Further extension of the mass in para-pharyngeal space was removed with ease using blunt dissection (Figure 2). Patient was discharged with mild facial nerve dysfunction (marginal mandibular nerve). Histopathology showed a plexiform multinodular tumor mass composed of hypo cellular proliferation with myxoid background; containing spindle cells, fibroblasts and rare mast cells. Occasional nuclear palisading is noted. No appreciable mitotic activity is noted. The tumor extends to the cauterized resection margin. No tumor necrosis is present. The adjacent salivary gland tissue shows mild chronic inflammatory changes (Figure 3,4,5) and immunohistochemical stain of S-100 confirmed that the tumour was plexiform neurofibroma (Figure 6). A 3-month follow-up facial nerve function was returned to normal. After24-monthsof follow-up there are no signs of recurrence.

Discussion

Primary neurogenic tumors of the facial nerve are exceedingly rare with most of them occurring intratemporal; the much rarer intraparotid involvement of such lesions are a diagnostic challenge. These benign lesions are divided into Schwanomas (more common) and neurofibromas [3]. The plexiform variety of the later has a high association with autosomal dominant neurofibromatosis type 1 (NF1) for which it is almost pathognomic with 10-15% chance of metastasis [1,4]. A review of literature revealed that there are around thirteen case reports of parotid plexiform neurfibroma; six cases were female and seven were male (Table 1).

The patient may also present with single or multiple lesions without being associated with NF1 same as our case in which there was no association with NF-1 [2]. These lesions are non-circumscribed, thick, irregular benign tubular lesion of peripheral nerve sheaths. They are often compared with bag of worms on clinical examination while it becomes difficult to distinguish it from vascular malformations [4].

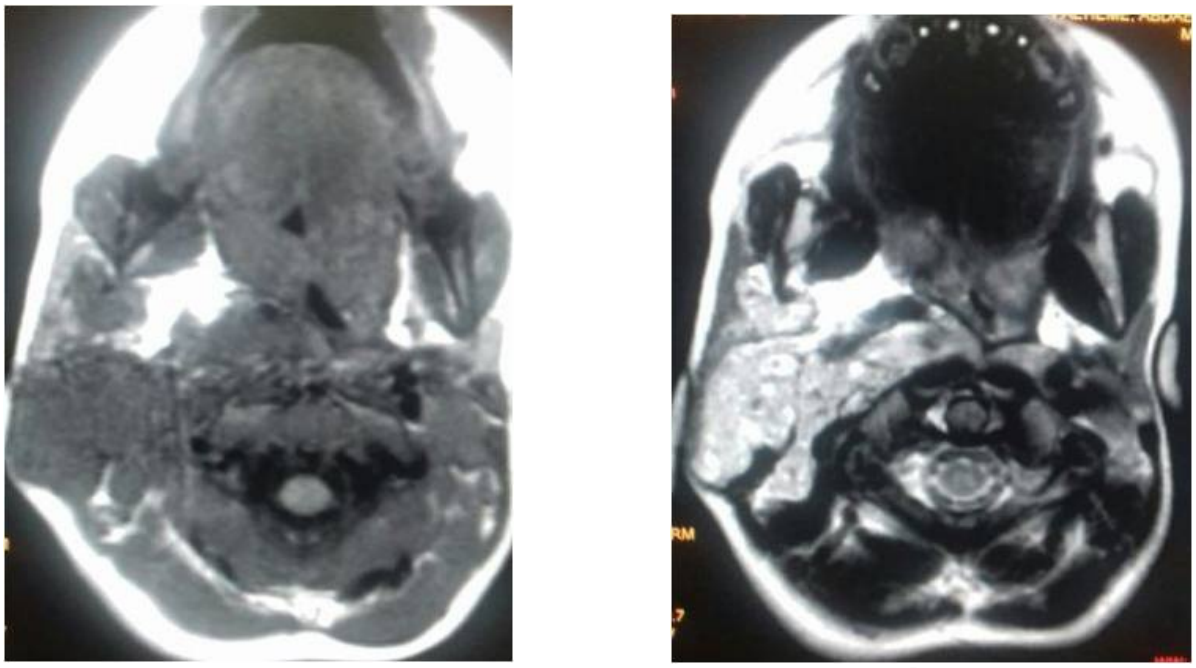
Increase in size with puberty and pregnancy has also been reported [1,5] while this may lead to facial disfigurement, airway, speech and swallowing difficulties ([1,4, 6,7,8]. They are commonly diagnosed at an early age while it is rare to come across such lesions in adolescent [7]. Fine needle aspiration cytology usually has low diagnostic yield in such cases [1] as in our case we were unable to get a satisfactory result. Histologically Schwanomas can be distinguished from neurofibromas; with former having Antoni A and B lesions with Verrocay bodies while in later the cellular pattern is dispersed in a much looser architecture with the absence of above mentioned Verrocay bodies and Antoni A, B lesions [9]. C.T or MRI head and neck are the radiological investigations used for such lesions [8], in our case MRI head and neck was found to be isointense with muscle on T1 and Hyperintense on T2 weighted MRI imaging, enhancing after gadolinium contrast. Complete surgical resection remains the treatment of choice for such cases as limited benefit from chemotherapy and almost no benefit from radiotherapy has been observed in literature [7]. Even though after complete excision of this benign tumor the rate of recurrence can reach up to 20% [10]. This is in part due to invasive nature of the lesion and the location of the tumor; which could pose a real challenge during surgery. Due to lack of a true capsule it is difficult to release the tumor from surrounding tissues, with extensive tumors associated with increase intra-

operative bleeding and more operative time being spent on securing hemostasis so it is advisable to operate these cases at early age [4,9]. Neurological deficits after surgical resection are not uncommon and may reach as high as 10% in head and neck alone [7].

Conclusion

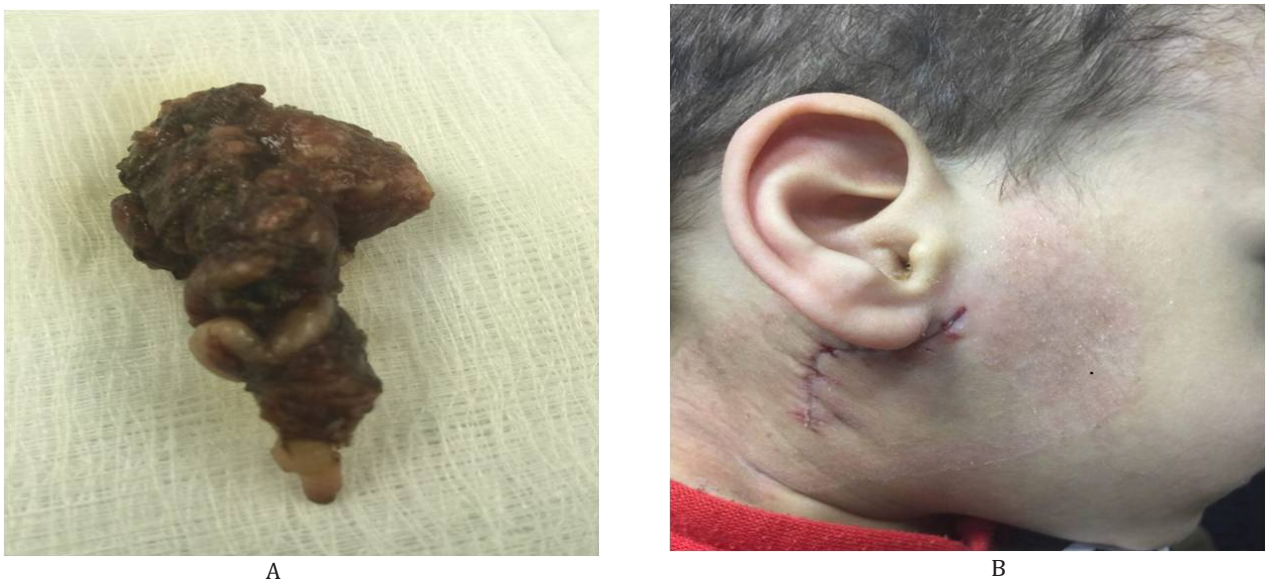
Solitary intra-parotid facial nerve plexiform neurofibroma is a rare diagnostic entity and represents a surgical challenge. As they may grow to a considerable size causing disfigurement, facial nerve dysfunction, and difficult resection due to lack of true capsule. We propose early surgical intervention.

Figures



A B

Figure1 (A): MRI Axial Cut T1 weighted image showing lesion with red arrow. **(B):** MRI Axial Cut T2 weighted image showing lesion with red arrow



A B

Figure 2: Typical gross appearance specimen of plexiform neurofibroma of parotid (A) and postoperative wound after one week of surgery (B).

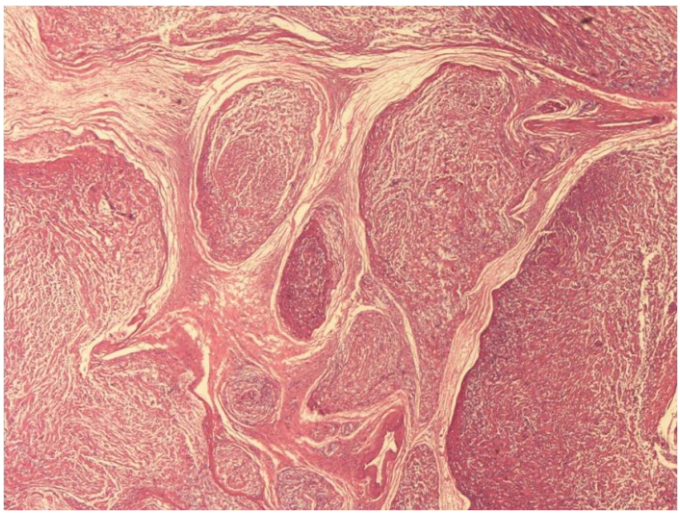


Figure 3: Photomicrograph of the plexiform neurofibroma, with tumor nodules arranged in plexiform pattern (Hematoxylin-Eosin stain, original magnification x40)

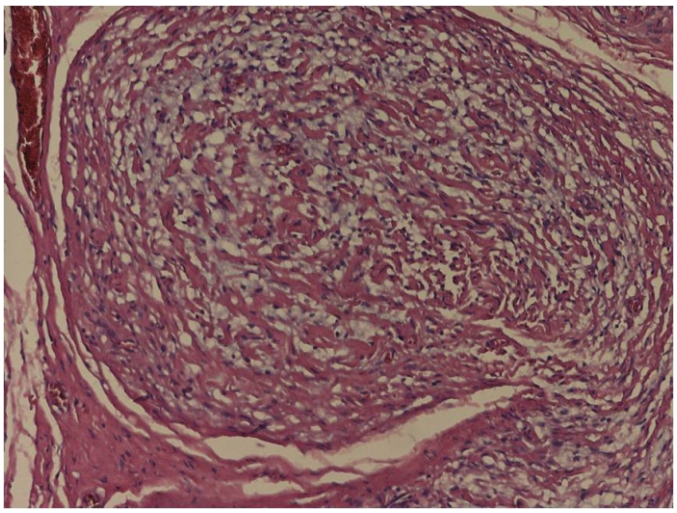


Figure 4: Photomicrograph of the plexiform neurofibroma, composed of spindle cells with thick collagen bundles and myxoid stroma (Hematoxylin-Eosin stain, original magnification x100).

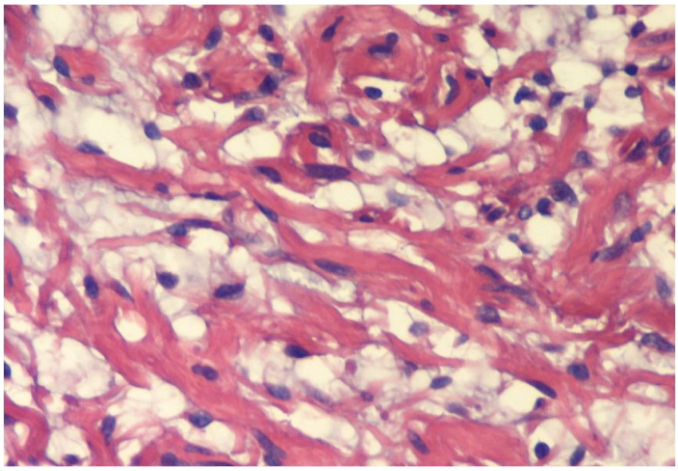


Figure 5: Photomicrograph of the plexiform neurofibroma, composed of elongated spindle cells with wavy nuclei (Hematoxylin-Eosin stain, original magnification x400).

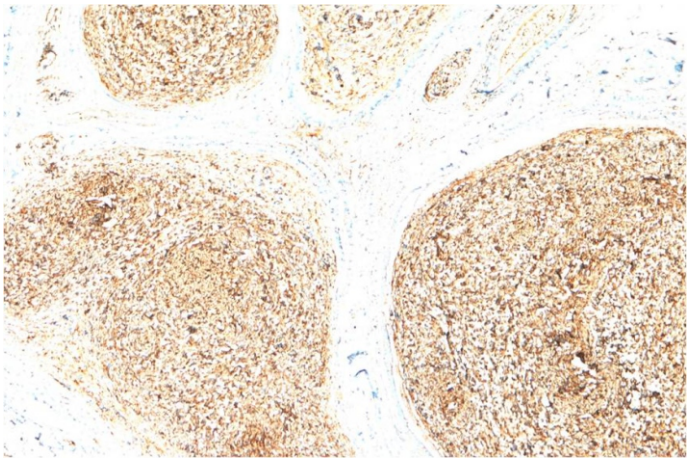


Figure 6: Photomicrograph of the plexiform neurofibroma, showing positive immunoreactivity for S-100 protein (Immunohistochemical staining for S-100 protein), original magnification x100).

Tables

Table 1 : Intraparotid plexiform neurofibroma reported cases in the literature

Literature	Authors	Publication Date	Age	Sex	Procedure
Intraparotid Facial Nerve Plexiform Neurofibroma: A Surgical Challenge. Open Journal of Clinical and Medical Case Reports. Otorhinolaryngol. 2018; 4 (18):1466-1472.	Mubarak Al Shraim, Montasir Junaid, Osamah A. Al-Qahtani , Fawziyah Y. Kilani, Ali S. Al-Qahtani.	2018	5	M	Superficial parotidectomy
Neurofibroma Arising In the Parotid Gland- A Rare Case Report. Journal of Medical Science and Clinical Research. 2016; 4.	Shagufta Qadri, Divya Rabindranath, Azka Anees Khan, Senthil P.	2016	17	M	Surgery
Intraparotid Neurofibroma of the Facial Nerve: A Case Report. Iran J Otorhinolaryngol. 2016 ;28 (87):287-90.	Nofal AA, El-Anwar MW	2016	40	M	Total parotidectomy

Solitary parotid plexiform neurofibroma - diagnostic difficulty in a clinically unsuspected case. Indian J Cancer 2014; 51:373-374.	Mohd Khalid, Khalid S, Zaheer S, Bhatnagar S, Ahmad M. Solitary parotid plexiform neurofibroma - diagnostic difficulty in a clinically unsuspected case.	2014	15	F	Total parotidectomy
Solitary Intraparotid Facial Nerve Plexiform Neurofibroma. International Journal of Clinical Medicine,2014 ; 5 :1125-1129.	Mesolella M, Di Lullo AM, Ricciardiello F, Oliva F, Pianese A, Misso G, Iengo M.	2014	5	M	Partial parotidectomy
Intra-Parotid Facial Nerve Multiple Plexiform Neurofibroma in Patient with NF1. International Journal of Pediatric Otorhinolaryngology. 2008; 72:553-557.	Fadda, MT, Verdino G, Mustazza MC, Bartoli, D, Iannetti G.	2008	10	M	Superficial parotidectomy
Facial plexiform neurofibroma in a child with neurofibromatosis type I: A case report. J Indian Soc Pedod Prev Dent 2007; 25: 30-35.	Patil K, Mahima V G, Shetty S K, Lahari K. Facial plexiform neurofibroma in a child with neurofibromatosis type I: A case report.	2007	12	M	Surgery
Solitary Intraparotid Neurofibroma of the Facial Nerve. Symptomatology, Biology and Management. HNO. 2006; 54: 772-777.	Fierek O, Laskawi R. and Kunze E.	2006	55	F	Surgery
Intraparotid facial nerve solitary plexiform neurofibroma: A first paediatric case report. International Journal of Pediatric Otorhinolaryngology. 2003; 67: 1113-1115.	Souaid JP, Nguyen VH, Zeitouni AG, anoukian J.	2003	17	f	Superficial parotidectomy
A rare case of a facial-nerve neurofibroma in the parotid gland. British journal of plastic surgery. 2002; 689-691.	Kosaka M, Miyanohara T, Mochizuki Y, Kamiishi H	2002	30	M	Superficial parotidectomy
The changing faces of a parotid mass. The Journal of Laryngology & Otology. 1999; 113(10): 938-941.	Hehar S, Dugar J, Sharp J.	1999	32	F	Surgery
Intraparotid facial nerve neurofibroma: a case report and literature review, Otolaryngol. Head Neck Surg. 1990; 102 : 413-415.	Albernaz MS, Pratt MF.	1990	22	F	Surgery

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