Case Report

Solitary Intraparotid Plexiform Neurofibroma: Case Report and Literature Review

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Abstract

Solitary plexiform neurofibroma is a peripheral nerve sheath tumor with proliferation of Schwann cells accompanied by mast cells and fibroblasts laying down collagen. The occurrence of this tumor in the salivary gland of an individual in the absence of neurofibromatosis 1 (von Recklinghausen disease) is rare. We present a case of sporadic solitary intraparotid neurofibromatosis in an 8-year-old female. The 8 cm × 6 cm × 2 cm mass removed through superficial parotidectomy showed the characteristic irregular bundles of Schwann cells, fibroblasts, mast cells, and strands of collagen. Tissue biopsy is indispensable in the diagnosis of this tumor and its differentiation from other more benign condition such as pleomorphic adenoma which it mimics clinically.

Keywords: Facial nerve, intraparotid tumor, neurofibroma, peripheral nerve sheath tumor, salivary gland tumor

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Introduction

Salivary gland tumors range broadly, from the more common benign tumors such as pleomorphic adenoma and Warthin tumor, to the less common malignant ones such as mucoepidermoid carcinoma, acinic cell carcinoma, or squamous cell carcinoma.[1] Tumors of nonepithelial origin constitute 10% of tumors of the salivary glands.[2] These mesenchymal tumors include hemangioma, lymphangioma, neurofibroma, schwannoma, lipoma, and sarcomas.[1]

A neurofibroma is a nerve sheath tumor which has the tumor cells consisting of proliferating Schwann cells and fibroblasts laying down wisps of collagen in a myxoid background.[3] Neurofibromas are one of the features of von Recklinghausen disease of neurofibromatosis type 1 (NF 1) manifesting with Café au lait spots, patches, plaques, or nodules all over the body.[3] Solitary neurofibromas may occur sporadically and have been reported to be more common in the head-and-neck region,[4] including the ear,[5] larynx,[6] lacrimal glands,[7] and the face.[8,9] The occurrence of this tumor in the salivary glands has been very low with only a few cases reported in literature.[10] Here, we present a case of solitary plexiform neurofibroma in a female child and a review of the literature. To the best of our knowledge, this is the first report of the occurrence of sporadic solitary intraparotid neurofibroma in Nigeria.

Case Report

The patient, an 8-year-old female, presented at the hospital with a 7-year history of a small swelling behind the left ear which had over time significantly increased in size. There was an associated hearing loss in the ipsilateral ear and dryness of the mouth. The patient did not notice changes in size while eating, neither was there a positive history of trauma to the site, otorrhea or otalgia, related nasal or throat symptoms. Café-au-lait spots or similar lesions were not observed in other parts of her body, and there was no family history suggestive of neurofibromatosis. In a bid to obtain cure to the ailment, the patient's parents employed the aid of native medications which required making small incisions on the site using sharp blades before herbal preparations were rubbed in. This invariably

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resulted in the presence of small scarification marks on the skin over the swelling. On further examination, the pinna was found displaced outward. The swelling extended from the tragus to the left mastoid tip and angle of the mandible [Figure 1]. It had an irregular surface and was firm, nontender, freely mobile, and not attached to the overlying skin or structures below. There were enlarged cervical lymph nodes on the same side. A diagnosis of a pleomorphic adenoma was considered at this point. This diagnosis was supported by a hypoechoic irregular mass in the lower region of the parotid which exhibited an enhanced flow on Doppler. The regional lymph nodes, especially the postauricular group appeared matted [Figure 2]. Subsequently, a superficial parotidectomy was performed with complete dissection of the mass. The postoperative condition was satisfactory, but she suffered residual incomplete facial nerve palsy. The removed tissue mass submitted to the histopathology laboratory for evaluation.

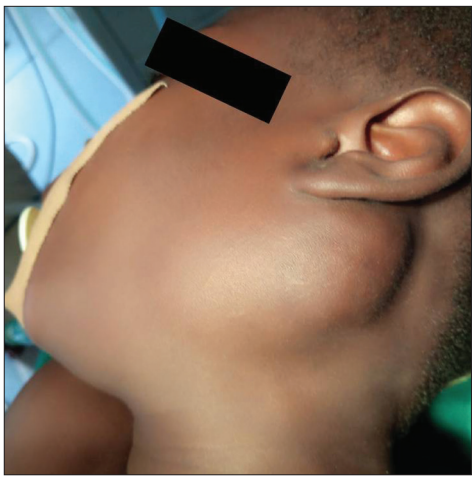

Figure 1: The picture shows a swelling which extended from the tragus to the left mastoid tip and angle of the mandible. It had an irregular surface, firm, nontender, and freely mobile without any attachment to the skin or underlying structures

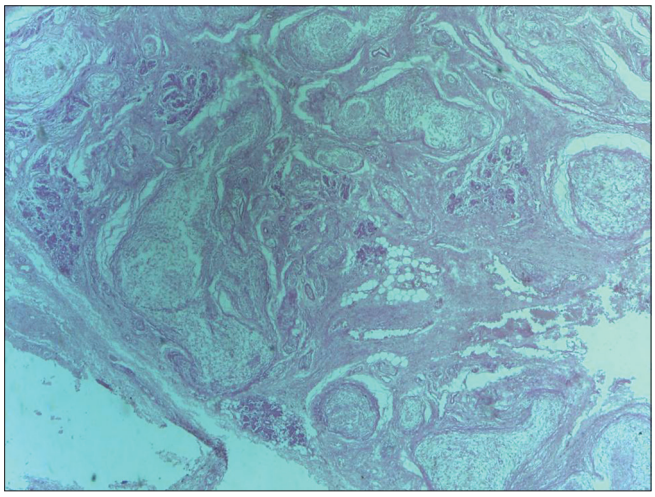

Figure 3: Intraparotid neurofibroma histology. At low power, the tumor shows numerous pale irregular neural bundles, surrounding fibrosis, and a few salivary gland ducts (H and E, ×4)

A firm gray–brown tissue with an irregular shape was received in the histopathology laboratory. It measured 8 cm × 6 cm × 2 cm. The tissue mass appeared spongy on sectioning through it with numerous cystic cavities and solid areas. Histological evaluation showed a salivary gland containing a well-circumscribed lesion consisting of numerous distinct irregular neural bundles of different sizes [Figure 3]. These were made up of groups of wavy spindle shaped or serpentine Schwann cells and strands of collagen disposed in a myxoid background [Figure 4]. The areas surrounding these neural fascicles contained normal salivary gland acini. These observations abrogated the clinical suspicion of pleomorphic adenoma in favor of a solitary intraparotid neurofibroma with a plexiform pattern.

Discussion

Intraparotid neurofibroma is very rare with only a few have been recorded in the English medical literature. Table 1 represents some of the cases reported so far from different

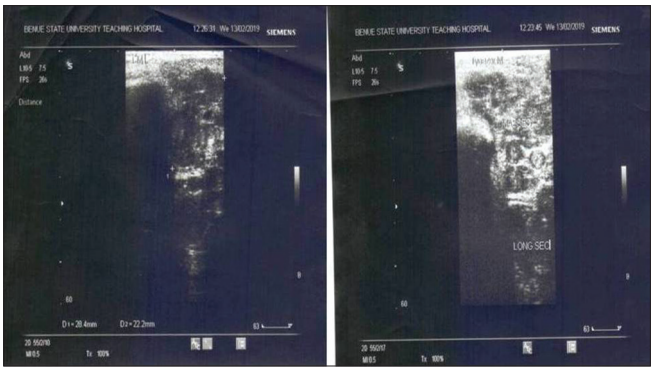

Figure 2: Parotid mass ultrasound scan. A hypoechoic irregular mass was observed in the lower region of the left parotid gland. The regional lymph nodes also appeared matted

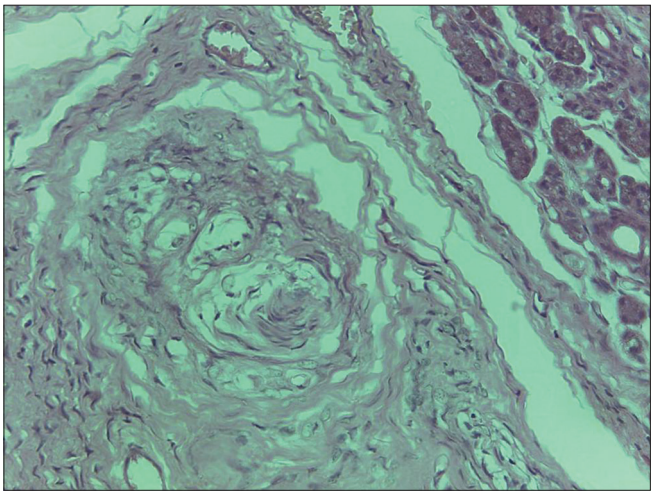

Figure 4: Intraparotid neurofibroma histology. The irregular neural bundles observed in this tumor, shown on this photomicrograph, are composed of spindle cells with curved serpentine nuclei disposed in a pale myxoid background. The top right corner of the image shows crops of salivary gland acini (H and E, ×40)

Table 1: Cases of intraparotid neurofibroma reported in the literature

Author	Year	Location	Age (years)	Sex	Size (mm)	Tumor duration (years)
Wilkinson *et al*.[11]	1971	The USA	59	Female	80×80	3
Malcolm and Lopes[12]	2002	The USA	35	Male	45×15×15	Unknown
Kosaka *et al*.[10]	2002	Japan	30	Male	30 (diameter)	3
Irfan *et al*.[13]	2009	China	35	Male	40×60	1
Shekar *et al*.[14]	2010	India	15	Male	80×50	Unknown
Sethi *et al*.[15]	2011	India	48	Female	30×25	2
Maheshwari *et al*.[16]	2011	India	40	Male	50×40	Unknown
Asha'ari *et al*.[4]	2012	Malaysia	6	Male	Unknown	2
Kumar *et al*.[17]	2013	India	45	Male	80×60×40	4
Mesolella *et al*.[18]	2014	Italy	5	Male	90×40×20	0.3
Rai and Kumar[19]	2015	India	36	Male	90×50	1
Satar *et al*.[5]	2016	Turkey	30	Male	26×31×36	0.5
Khalele[20]	2016	Egypt	48	Male	Unknown	Unknown
Nofal and El-Anwar[21]	2016	Egypt	40	Male	60×30	2
Qadri *et al*.[2]	2016	India	17	Male	50×45×30	0.7
Blioskas *et al*.[22]	2017	Greece	6	Male	60×45×30	1
Shwetha *et al*.[23]	2017	India	35	Male	33×30×30	Unknown
Vhriterhire *et al*. (this article)	2020	Nigeria	8	Female	80×6×2	7

locations of the world over the years. Kosaka *et al.* included a review of seven cases in a case report.[10] The list of the cases presented in Table 1 is by no means exhaustive but reflects the fact that this rare tumor is increasingly being recognized and reported in peer-reviewed literature. Seven (38.9%) of the 18 cases reported in the literature emanated from India, two were from each of the United States of America and Egypt (11.1%), whereas only one (5.5%) was reported from each of the remaining countries [Table 1]. This report represents the first case of intraparotid neurofibroma in the pediatric age group to be published in the peer-reviewed literature in Nigeria (to the best of our knowledge).

The tumor has been observed to occur over a wide age range. Mesolella *et al*. reported a case of neurofibroma in the right parotid gland of a 5-year-old boy, whereas Wilkinson *et al*. reported an occurrence of this tumor in the left parotid gland of a 59-year-old woman.[11,18] The index case was first noticed when the patient was just 1-year-old, but sought medical attention and had it removed at the age of 8 years. In contrast, most cases reported in the literature showed that solitary plexiform intraparotid neurofibroma mostly occurred within the fourth decade consisting of 6 (33.3%) out of the 18 cases considered in this review [Table 1]. Moreover, the tumor seems to be more common in males (15/18, 83.3%) than females (3/18, 16.7%) based on the reported cases presented in Table 1.

Since most cases of intraparotid neurofibromas arise from the facial nerve or its branches, facial paresis or paralysis has been reported to be associated with about 20% of cases.[10] Facial paresis sometimes followed as a complication of the parotidectomy in a patient who previously had full functions of the facial nerve.[13,15,21] While some instances might have a long-lasting severe residual loss of facial nerve functions, Irfan *et al.* recorded just a mild paresis, whereas in the case studied by Shwetha *et al.*, the patient experienced simple transient neuropraxia.[13,23] The patient in this case was left with a similar incomplete facial paralysis. Nerve axons often penetrate through the tumor making it a herculean task to separate the tumor without sectioning of adjoining nerve twigs.[10]

The diagnosis of a solitary intraparotid neurofibroma often proves challenging in the absence of obvious features such as the cutaneous manifestations of von Recklinghausen disease or facial paresis. It presents like any other slow-growing salivary gland tumor and relies heavily on tissue biopsy histology to differentiate it from the commonly occurring benign pleomorphic adenoma. Histologically, it is composed of a mixture of S-100 positive Schwann cells, fibroblasts, perineurial cells, mast cells, collagen, and endothelial cells.[24] The tumorigenesis of plexiform neurofibroma is dependent on the interplay of factors in the microenvironment such as secretion of stem cell factor by the Schwann cells which recruit mast cells, abnormal c-kit signaling by the mast cells, and loss of heterozygosity of NF 1 gene in Schwann cells as observed in experimental studies.[25,26] Features indicative of malignant change such as atypia and necrosis occur only very rarely in salivary gland plexiform neurofibroma and were not found in this index case presented.

Conclusion

Only a number of cases of salivary gland solitary plexiform neurofibroma have been reported worldwide. Presented in this article is a report of the occurrence of this rare condition in an 8-year-old female without NF 1 (von Recklinghausen disease). The clinical resemblance of pleomorphic adenoma by this benign neoplasm makes tissue histology an indispensable tool for its accurate diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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