World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(CASE REPORT)

Plexiform neurofibroma of the parotid gland: Case report

Dounia Berrada Elazizi [1,*], Driss Idrissi [1], Othmane Benhoummad [2], Youssef Rochdi [1] and Abdelaziz Raji [1]

[1] *Faculty of Medicine and Pharmacy of Marrakech, Cadi Ayyad University, Marrakech, Morocco.*
[2] *Faculty of Medicine and Pharmacy of Agadir, Ibn Zohr University, Agadir, Morocco.*

World Journal of Advanced Research and Reviews, 2023, 17(01), 444–449

Publication history: Received on 04 December 2022; revised on 12 January 2023; accepted on 15 January 2023

Article DOI: https://doi.org/10.30574/wjarr.2023.17.1.0053

Abstract

Intra parotid plexiform neurofibroma of the facial nerve is anextremely rare benign tumor, difficult to diagnose and manage, often considered pathognomonic of neurofibromatosis type 1 (NF1 or von Recklinghausen's disease). Its diagnosis is essentially pathological and the treatment is primarily surgical to avoid any degeneration. We report a case of plexiform neurofibroma of the facial nerve centered on the parotid region in a 1-year-old child.

Keywords: Neurofibroma; Parotid Gland; Benign Tumor; Facial Nerve

1. Introduction

Plexiform neurofibroma (NFP) is a benign nerve tumor, most often developed in the cephalic region due to its rich innervation. It is usually considered pathognomonic of Von Recklinghausen's disease, also called neurofibromatosis type 1 (NF1), which is one of the most common autosomal diseases with an incidence of about 1/3000 births [1].The clinical problem of a plexiform neurofibroma is not only related to the significant aesthetic disfigurement and compression of adjacent vital structures, but also because of its potential for malignant transformation, which occurs in about 10% of cases [2]. We report a case of plexiform neurofibroma centered on the parotid region in a 1-year-old male child evolving since birth.

2. Observation

A 1 year old male child, from a first degree consanguineous marriage, followed for Von Recklinghaussen disease, suffered since birth from a progressively increasing swelling of the right parotid region, with café au lait spots in opposite, without other lesions or associated signs or other malformations. Clinical examination revealed a mass in the right parotid and laterocervical region that was soft, well defined, not painful on palpation, mobile in relation to the superficial and deep plane, with café-au-lait spots opposite (figure1), without any cervical lymphadenopathy, neither peripheral facial palsy nor trismus. The ophthalmologic and neurologic examinations were unremarkable. Endoscopic examination of both nasal passages appeared normal.

*Corresponding author: Dounia Berrada Elazizi

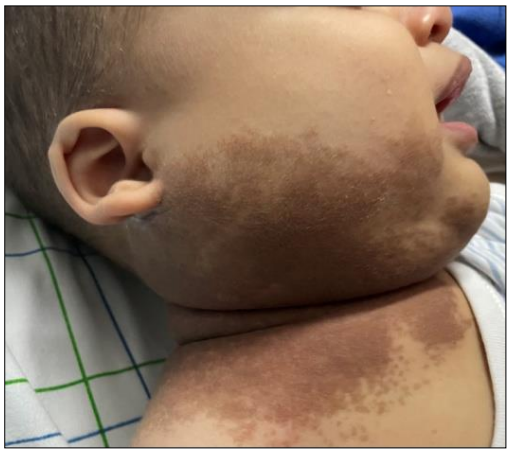

Figure 1 Swelling of the right parotid and laterocervical regionwith café au lait skin spots

The MRI showed a right latero-cervical lesional process centered on the parotid compartment infiltrating inside the para-pharyngeal space, the pterygoid region, further back it comes into contact with the pre and retro-stylian spaces, in front and laterally it invades the masticatory muscles and the mandibular angle, at the bottom it includes the submaxillary gland and extends to the cervical region, behind from the top to the bottom it comes into contact with the golf course of the internal jugular vein, the external auditory meatus and the sternocleido muscle mastoid (figure2).

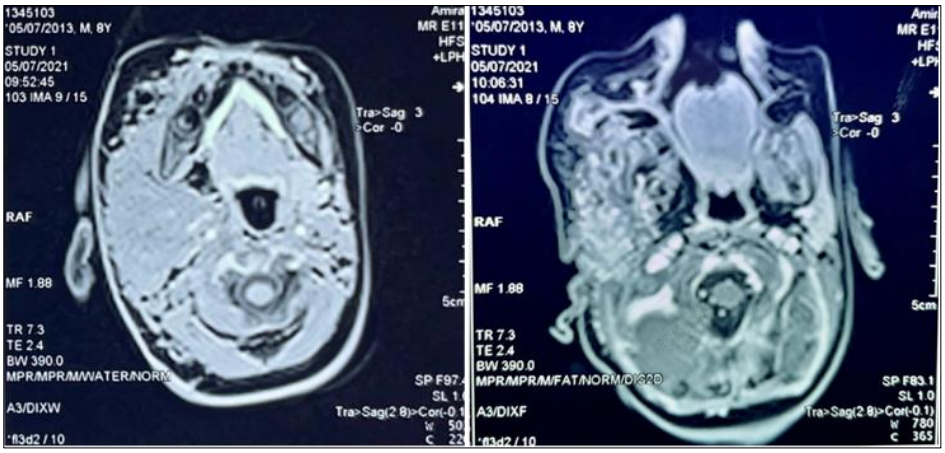

Figure 2 Axial T1 MRI before and after gadolinium injection showing the extension of the plexiform neurofibroma

The surgical intervention was based on the external approach with Redon incision followed by dissection and preservation of the trunk of the facial nerve and its division branches with complete resection of the tumor including a total parotidectomy with excision of a masseter extension, of a cervical extension extended in supra clavicular and a pre-vertebral extension and sub-digastric lymphadenopathy (figure3).

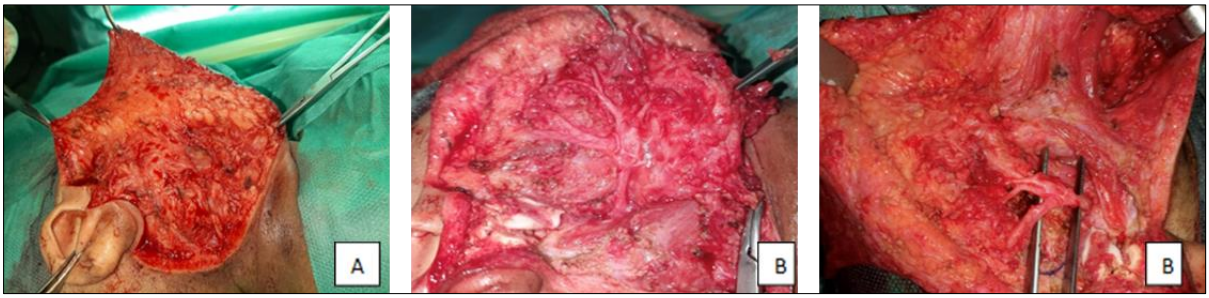

Figure 3 Intraoperative view

- View before dissection
- Total parotidectomy with preservation of the trunk of the facial nerve and its branches

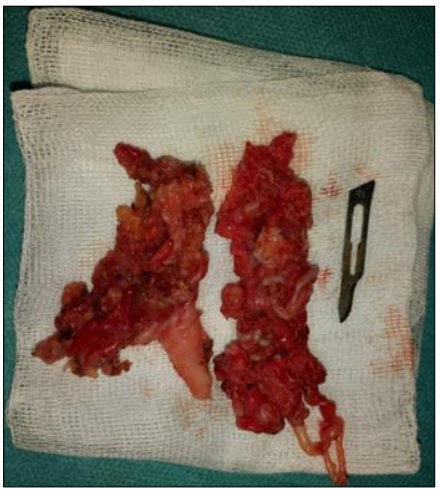

Figure 4 The resected tissue

Indeed, the histological study made it possible to confirm the diagnosis (Figure5) revealing a plexiform neurofibroma dissociating the parotid salivary parenchyma without signs of malignancy with the presence of some reaction-type lymphadenopathy.

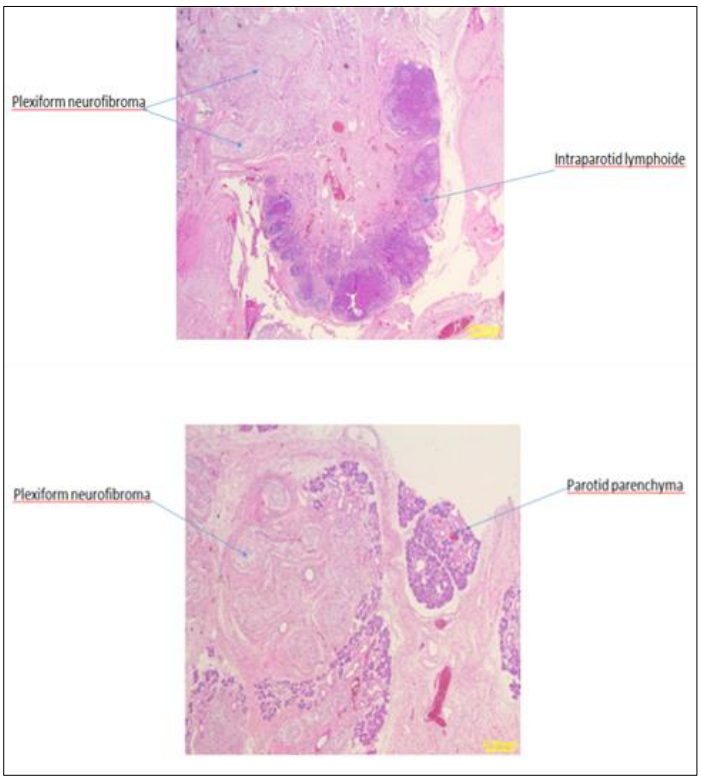

Figure 5 Plexiform neurofibroma of the parotid gland. Thickened nerve bundles are seen growing within and around the glandular parenchyma

The patient presented a facial paresis of the lower territory which regressed after a few days.(Figure6)

Figure 6 Post-operative follow-up

3. Discussion

Plexiform neurofibroma is a benign tumor of the perinerve of the peripheral nerves. It consists of a proliferation of Schwann cells arranged in a myxoid tissue of variable abundance including many fibroblasts and collagen fibers with a so-called "grated carrot" appearance [3]. The presence of anti-neurofilament antibodies within the proliferation can help with diagnosis. The myxoid background is of variable abundance. The cell form can mimic a schwannoma but in immunohistochemistry, the S100 protein is expressed less significantly than in schwannomas. A rather myxoid form can resemble a myxoma.

Plexiform neurofibroma is strongly associated with neurofibromatoses [2], in particular von Recklinghausen's disease (NF1) where it occurs in 24-32% of affected patients [4,5]. NFP belongs to the four types of neurofibromas encountered in NF1 according to the classification of the 1988 consensus conference of the National Institute of Health Development. The diagnosis of the disease is based on the presence of at least two of the following criteria [6,7]:

- At least six café au lait spots larger than 5 mm before puberty´ and more than 15 mm after puberty
- Two or more neurofibromas, one or more plexiform neurofibromas;
- Lentiginous spots of the axillary or inguinal region;
- -Two or more hamartomas of the iris (Lisch's nodules), a glioma of the optic pathways;
- -A characteristic bone lesion (pseudarthrosis of a long bone, spheno-orbital dysplasia, cervical kyphosis).

Formerly called plexiform neuroma or royal tumor, NFP differs from other types of neurofibromas in the importance of its Schwannian component [8]. NFPs develop along the path of the nerve trunks of the different plexuses and their dividing branches, at the craniofacial, thoracic and abdominopelvian stage, with hypodense masses not enhanced by the iodinated contrast [8, 9,10]. Most often, they simulate lymphadenopathy and direct to various differential diagnoses depending on their location [11]. Craniofacial localization exists in 3-7% of cases of neurofibromatosis type I, with NFP developing at the expense of nerve branches of the fifth, seventh or ninth cranial pair [6,11].

The diagnosis of plexiform neurofibroma is essentially histopathological. The clinical aspect is not specific, it may evoke a tumor or vascular malformation, a skin or conjunctiva tumor, benign or malignant [12]. The skin in front of it is classically pathological (hypertrophy, hypertrichosis, brown pigmentation, angiomatous appearance), with two main characteristics: hyperextensibility and lack of elasticity [13]. This skin or subcutaneous tumor, sometimes painful, has a very variable size, ranging from a few centimeters to an entire body segment. Plexiform neurofibroma may be present at birth, but often appears in children between the age of two and five [14].

The imaging is quite unspecific. Ultrasound can still eliminate a rapidly circulating tumor or vascular malformation. MRI has the classic appearance of peripheral nerve tumors (low to moderately intense T1 signal, T2 hypersignal with often a heterogeneous appearance). Hypersignal areas correspond to areas of myxoid tissue or cystic degeneration. The nodular areas of hyposignal correspond to collagen and fibrous tissue and can be enhanced with gadolinium [14].

Neurofibromas can degenerate in 3 to 15% of cases into neurofibrosarcoma. This incidence is 9000 times higher than in the general population [3,4,17,19]. The risk of degeneration alone justifies a systematic excision but this is most often performed in front of a painful tumor and or causing aesthetic damage. Given an often large size, a hemorrhagic character and poorly limited, the excision is most often delicate and cannot most of the time be "carcinological". These

incomplete resections do not appear to increase the risk of neurofibrosarcoma degeneration [20]. As part of NF1, it is advisable to perform annual clinical monitoring [19]. The risk of malignant degeneration of a possible tumor residue must be considered in the face of any rapid increase in the volume of the lesion, the appearance or increase of pain.

The management of plexiform neurofibromas is controversial when facial nerve function is preserved before surgery. Unlike schwannomas which tend to push nerves away and allow its dissection, nerve fibers in neurofibromas pass directly into the tumor making dissection difficult and most often requiring section of the nerve with reconstruction [15].

In our case, the trunk of the facial nerve and its branches was preserved.

4. Conclusion

Intraparotid plexiform neurofibroma is extremely rare. Preserving the nerve during surgery is difficult, almost impossible, and its excision can lead to significant morbidity. The risk of sarcomatous degeneration justifies the removal of the lesion as complete as possible. Annual clinical and radiological monitoring is necessary to detect a possible recurrence or malignant transformation.

Compliance with ethical standards

Acknowledgments

The authors would like to thank everyone who contributes in this study.

Disclosure of conflict of interest

The authors have no conflict of interest to declare.

Statement of informed consent

Informed consent and verbal permission were obtained from the family of patient before the submission of this article.

References

[1] WOLKENSTEIN P, ZELLER J and ISMAILI N Neurofibromatoses Elsevier Paris 2002; 98-755-A-10

[2] Dr. Shabnam Bhandari Grover, Dr. Rohit Kundra, MBBS , Dr. Hemal Grover, MBBS, MD b , Dr. Vishal Gupta, MBBS, MD a , Dr. Rishab Gupta, MBBSImaging diagnosis of plexiform neurofibroma- unravelling the confounding features: A report of two cases 2021

[3] Peltonen J, Jaakkola S, Lebwohl M, Renvall S, Risteli L, Virtanen I, et al. Cellular differentiation and expression of matrix genes in type 1 neurofibromatosis. Lab Invest 1988;59(6):760—71.

[4] Friedman JM, Birch PH. Type 1 neurofibromatosis: a descriptive analysis of the disorder in 1728 patients. Am J Med Genet 1997; 70(2):138—43.

[5] Huson SM, Harper PS, Compston DA. Von Recklinghausen neu-rofibromatosis. A clinical and population study in south-east Wales. Brain 1988;111(Pt 6):1355—81.

[6] Abouchadi A, Nassih M, Rzin A. Orbitotemporal plexiform neurofibroma: about 6 cases. Rev Stomatol Chir Maxillofac2005;106:272–5.

[7] Pinson S, Wolkenstein P. Neurofibromatosis 1 (NF1) or von Recklinghausen's mala-die. Rev Med Int 2005;26:196–215.

[8] Lange F, Herlin C, Frisian L, et al. Management of isolated plexiform neurofibroma in children: about four observations. Ann Chir PlastEsthet2013;58:694–9.

[9] Levy Bencheton A, Mallet S, Rojat Habib MC. Isolated plexiform neurofibroma´ of late onset, outside a context of neurofibromatosis. Ann Dermatol Venereol2010;137:301–4.

[10] Moussali N, Belmoukari S, Elmahfoudi H, et al. A rare cause of dyspne´e: plexiform neurofibroma of the larynx. Arch Pediatr 2013;20:629–32.

[11] Benaissa L, El Kharras A, Bassou D, et al. Left cervical mass. Radiol Sheet 2009;49:207–10.

[12] O'Keefe P, Reid J, Morrison S, Vidimos A, DiFiore J. Unexpected diagnosis of superficial neurofibroma in a lesion with imaging features of a vascular malformation. PediatrRadiol 2005; 35(12):1250—3.

[13] Mimoun N, Razzouq N, Wolkenstein P, Moreno JC, Marty JP, Lantieri L, et al. Evaluation of skin viscoelasticity in type 1 neurofibromatosis patients. Skin PharmacolPhysiol 2006; 19(1):22—7.

[14] F. Lange, C. Herlin, L. Frison, D. Bessis, C. Rouleau-Dubois, M. Bigorre, G. Captier Management of plexiform neurofibroma isolated in childhood: About four observations 2011

[15] M. Kosaka, T. Miyanohara, Y. Mochizuki and H. Kamiis.A rare case of a facial-nerveneurofibroma in the parotid gland 2002