



A rare cause of cervical swelling: Solitary plexiform neurofibroma

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ABSTRACT

Introduction: Plexiform cervical neurofibromas are benign neoplasm, extremely rare, difficult to diagnose and to manage. Only some cases have been reported in the literature.

Case presentation: We report the case of a 60-year-old man admitted for a lateral neck mass, for which the surgical indication was the increase in volume of this mass, as well as the aesthetical impairment, the surgical exploration found the tumor attached to the cervical plexus. The excision of the mass was performed without damaging nerve. The pathological study was in favor of a plexiform neurofibroma.

Discussion: Even though Plexiform cervical neurofibroma are extremely rare, and their diagnosis are not often primary evoked in front of any growing mass of this region, the surgeon must keep in mind the existence of these neoplasms as a differential diagnosis of a neck tumor.

Conclusion: Surgery remains the gold standard in the treatment of these locally invasive tumors. It is essential that the surgeon keep in mind the possibility of these tumors as a differential diagnosis of a neck tumor.

1. Introduction

We present a case in accordance with SCARE 2020 criteria [1]. Plexiform neurofibroma is a rare, poorly defined benign tumor of the peripheral nerve sheath. They can be single or bilateral, sometimes stepped, superficial or profound. Often considered pathognomonic of neurofibromatosis type 1 (NF1 or von Recklinghausen disease), it can be solitary without NF1.

Formerly known as "plexiform neuroma" or the plexiform neurofibroma is a "royal tumor" and differs from the other types of neurofibroma by the importance of the component schwannian [2]. The diagnosis of plexiform neurofibroma is essentially anatomopathological, particularly without the suggestive context of type I neurofibromatosis. The clinical aspect is not specific, and may evoke a vascular tumor or malformation, a skin or connective tissue tumor, benign or malignant [3]. Generally, these tumors are slow-growing. Their symptomatology is variable and depends on their topography [4].

2. Case report

A 60-year-old male patient was referred to our otolaryngology department. The patient had an irrelevant past medical history with no known allergies. He had a history of smoking (25 pack-years). He reported with a long standing swelling in the right supraclavicular region. The swelling persisted for more than ten years.

On clinical examination, it was found that the swelling was firm in nature, non-tender, and non-pulsatile. It was located at the level 5 region and measured 5 × 4 cm, (FIG: 1). No other neck masses. Dermatological, ophthalmological and MRI of the nervous system were normal.

CT scan revealed a large well-defined mass located in the right supraclavicular region. It measured 7 × 3.5 cm (transverse × anteroposterior measurements). (FIG: 2). Further, MRI scan revealed a low T1 signal with a high T2 signal with heterogeneous enhancement. No other neck masses were found and lymphadenopathy was also absent.

The surgical exploration found the tumor attached to the cervical plexus. (FIG: 3). The excision of the mass was performed without damaging nerve. (FIG: 4).

In the pathological study, the tumor was poorly limited,

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Fig 1. Long standing swelling in the right supraclavicular region.



Fig 3. Surgical exploration showing the tumor attached to the cervical plexus.

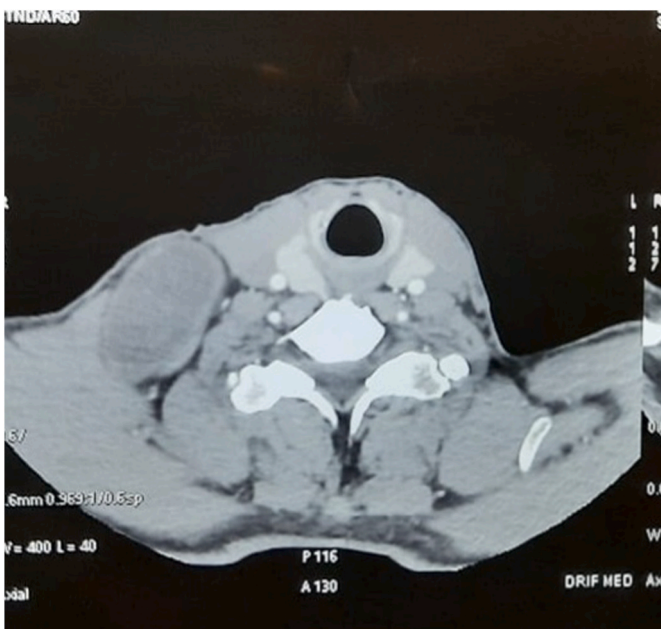


Fig 2. CT scan revealing a large well-defined mass located in the right supraclavicular region.



Fig 4. Excision of the mass.

unencapsulated, containing numerous neofomed with Cystic tumor proliferation, with elongated spindle-shaped cells with corrugated nuclei, without atypia, on a fibrous background. (FIG: 5).In immunohistochemistry, the Tumor cells express pS100 intensely and diffusely. Tumor cells do not express AML, EMA, or CD99. (FIG: 6). In the light of these findings (clinical evolution, histology, immunohistochemistry (IHC) and the imaging data), the lesion was diagnosed as solitary plexiform neurofibroma.

Regarding the increase in the volume of the mass and the esthetic problem, the patient was planned for a resection churgical. The patient was followed up carefully, without evidence for local recurrence 1 year after surgery.

3. Discussion

Neck nerve tumors are rare primary tumors, but have been known since 1742 when Haller discovered the carotid corpuscle; then in 1803 Rodier introduced the term “neuroma” for tumors of the peripheral

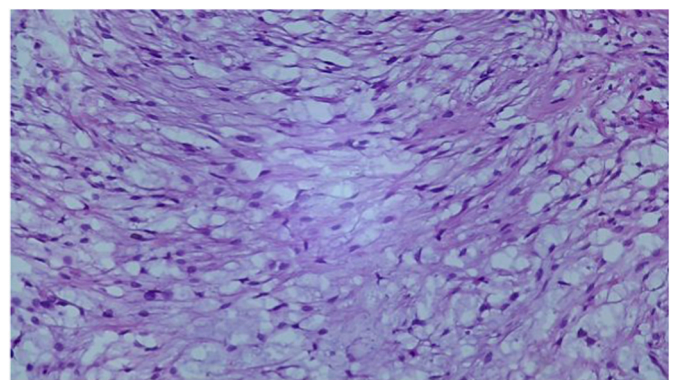


Fig 5. Pathological study.

nerves [5].

PNF, an uncommon benign tumour, usually presents at birth or in the first years of life [6]. A neck mass is the usual clinical presentation of neck nerve tumor. The mass is often isolated and painless although it may be an esthetic blemish, but can also be associated with clinical signs related to the function of the affected nerve. Clinical diagnosis is

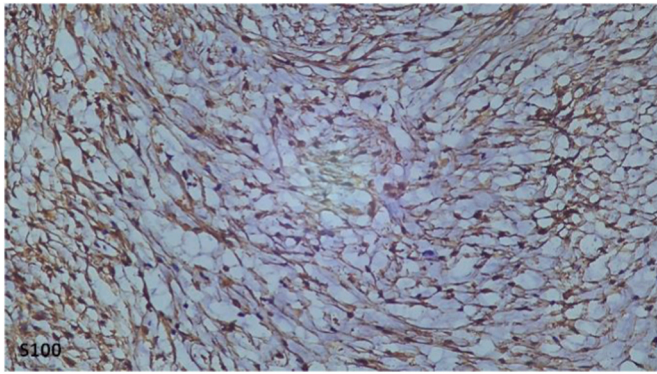


Fig 6. Immunohistochemistry.

hampered by the numerous differential possibilities in this location, but may be suggested by slow progression, by relative absence of symptoms, and on imaging [7].

Recent advances in immunohistochemistry and imaging allow increasingly precise diagnosis [8,9]. The majority of such peripheral nerve tumors are benign, but malignant transformation may occur [10].

Plexiform neurofibroma is strongly associated with neurofibromatosis, especially Von's disease Recklinghausen (NF1), where it occurs in 24–32% of the patients [11,12]. The location of the screen-facial display exists within 3–7% of the In the case of a neurofibromatosis type I case [13].

Plexiform neurofibroma belongs to the group of to the 4 types of neurofibromas encountered in the NF1 according to the classification of the consensus conference of 1988 from the National Institute of Health Development [14]:

- _ discrete cutaneous neurofibromas of the epidermis or dermis;
- _ discrete subcutaneous neurofibromas that are deeper;
- _ deep nodular neurofibromas;
- _ diffuse plexiform neurofibromas.

These masses can be quite disfiguring, and hemifacial hypertrophy can occur. These tumors are known to cause symptoms ranging from minor discomfort to extreme pain. The consistency of the lesion has been compared to that of a “bag of worms” because of the presence of soft areas interspersed with firm nodular areas. This consistency was well appreciable in the lesion seen in our patient. These lesions sometimes demonstrate a vascular nature.

On CT, the NFPs appear spontaneously isodense with respect to the muscles. They take up contrast only weakly, less than the muscle, which gives them a hypodense aspect (different aspect from schwannomas and neuro-fibrosarcomas) [15]. On MRI, plexiform neurofibromas are typically isointense with muscle on T1 WI, hyperintense on T2 WI and enhance markedly following gadolinium contrast injection [16]. Deep plexiform neurofibromas typically present with a target-like appearance on T2-weighted MR images, with central low-signal intensity and peripheral high-signal intensity. This architecture could account for the centrally T2 dark (nerve fibers) and peripherally T2 bright (myxoid) appearance of these target-like lesions. The absence of a target-like appearance does not rule out a lesion being a neurofibroma, particularly if the lesion has a superficial location [15].

At histological analysis, a localized, solitary neurofibroma is composed of interlacing fascicles of wavy, elongated cells that often contain abundant amounts of collagen. Rarely, myxoid areas and degenerative regions could be found in neurofibromas. Diffuse neurofibroma contains very uniform, prominent fibrillary collagen. Both localized and diffuse neurofibromas are positive for S-100 protein at immunohistochemical analysis but this is not a stable finding [17,18]. It needs to be differentiated from schwannoma which is encapsulated, while plexiform

neurofibroma is noncapsulated [19].

In anatomopathology, neurofibroma is poorly limited, not encapsulated, with the absence of thick-walled vessels; Immunohistochemistry is of limited value in this differential diagnosis [20].

Treatment of localized and diffuse neurofibromas (not associated with NF-1) is often surgical resection. Neurofibromas cannot be easily separated from normal nerve, and complete excision of the neoplasm may require sacrifice of the nerve

[17]. While this treatment may be acceptable in superficial lesions, deep lesions can only be managed from the patient's morbidity related to nerve damage and neurological deficits of the surgery [20].

In our case report, total removal of the lesion was performed because it was not related to a major cervical nerve. Transformation to malignant peripheral nerve-sheath tumours is the most feared complication of NF-1 but is less common in solitary neurofibroma and diffuse neurofibroma [21].

The estimated prevalence of malignant transformation varies from 2 to 29%. Sudden increase in size of a previously diagnosed neurofibroma well known to be clinically and radiologically stable, should be viewed with great suspicion of malignant transformation and must lead to immediate biopsy. Malignant transformation of these lesions, without association to NF-1, is rare and not well documented in the literature [22].

4. Conclusion

Solitary plexiform neurofibroma has to be considered in the differential diagnosis of cervical masses. They are rare tumors with low risk of malignant transformation. If local conditions and the general condition of the patient allow surgery, complete resection of the lesion can be proposed.

Surgery remains the gold standard in the treatment of these locally invasive tumors; however, guidelines need to be established regarding the selection of surgical candidates. It is essential that the surgeon keep in mind the possibility of these tumors as a differential diagnosis of a neck tumor.

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Declaration of competing interest

The authors declare having no conflicts of interest for this article

Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2021.102225>.

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Ethical approval

I declare on my honor that the ethical approval has been exempted by my establishment

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

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 Mennouni Mohamed Amine: writing the paper
 Youssef Oukessou: study concept
 Sami Rouadi: study concept
 Redallah Abada: study concept
 Mohamed Roubal: correction of the paper
 Mohamed Mahtar: correction of the paper
 Regragui Meriem: data analysis
 Karkouri Mehdi: data analysis

Registration of research studies

Researchregistry5198.

Guarantor

DR AHMED BRAHIM AHMEDOU

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