Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com/journal/sjmcr/home

OPEN ACCESS

Isolated Neurofibroma: A Rare Case and an Unusual Localisation

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DOI: 10.36347/sjmcr.2019.v07i06.009 | **Received:** 15.06.2019 | **Accepted:** 26.06.2019 | **Published:** 30.06.2019

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Abstract **Case Report**

Isolated neurofibroma is a rare entity, with only few cases reported in the literature so far. Magnetic resonance imaging (MRI) is the key to the diagnosis. We report a 37 year-old man with an antero medial thigh mass diagnosed as neurofibroma on the basis of the imaging findings on US and MRI examinations. The differential diagnosis can be excluded by the fine imaging analysis always associated with the clinical context.

Key words: Neurofibroma – isolated - femoral nerve – uncommon - MRI.

INTRODUCTION

Neurofibroma is one of the benign peripheral nerve sheath tumors that's commonly being a part of the various signs of the multisystem genetic disorder called Von Recklinghausen's disease or neurofibromatosis type 1 (NF-1).

Isolated neurofibroma is a rare and uncommon condition, based on our literature review, only few cases have been reported. The world Health organisation (WHO) had subdivided this entity into two wide categories : dermal and plexiform. The first one arise from a single peripheral nerve, while the second is associated with multiple nerve blundles [1].

The diagnosis is based on imaging and in some cases, histological examination is necessary to rule out the differential diagnosis. Ultrasound (US) and magnetic resonance imaging (MRI) are the reference modalities. Computed tomography (CT) is rarely performed because of its low resolution for soft tissues.

For illustration, à rare case of isolated neurofibroma of a cutaneous branch of the femoral nerve is presented.

CASE REPORT

A 37 year-old patient was complaining chronical right mechanical gonalgia with no history of fever or trauma. The physical examination found a small mobile mass in the right tigh root wich generated an exquisite pain radiating down the antero medial side of the thigh until the knee. Therefore, the patient were send to our department to conduct an US examination of soft tissue. It showed a fusiform and well defined mass, hypoechoic homogeneous with sharp borders that seem to continue a linear nervous structure on the both sides. This swollen area measures : 18 x 15 mm (Figure 1) and it's moderately vascular on doppler color flow.

For a better characterization and locoregional extension assesment of the mass, an MRI was performed. The protocol consisted of axial T2 weighted, fat-suppressed T2 weighted, T1-weighted scan and contrast-enhanced fat-suppressed, axial and coronal T1-weighted images using a 1.5-Tesla system.

It objectified a small elongated antero medial thigh mass centered to the nerve root (a cutaneous branch of the right femoral nerve) wich appears in isosignal on T1 weighted sequence with moderate enhancement after intravenous gadolinium injection. (Figure 2).

On the basis of these imaging findings, and remains clinical data, specially no « cafe-au-lait » spots or similar family cases were noted, the diagnosis of a solitary neurofibroma was strongly suggested, and finally confirmed by histological examination (Figure 3).

DISCUSSION

A rare case of a solitary neurofibroma of the lower extremity was presented. According to literature data, this entity without any other evidence of von Recklinghausen's disease is uncommon. It represents only 10% [2] of all isolated nerve tumors [3].

Borchardt first reported, in 1927, a solitary tumor not related to neurofibomatosis [4]. Since 1962, isolated neurofibroma have become recognised as a distinct entity following Heard's report on 46 such peripheral tumors wich included 35 superficial and 11 deep lesions [5]. Harkin and Reed suggested the existence of a forme fruste of multiple neurofibromatosis.

It usually affects young adults between 20 and 30 years old with a sex ratio of 8.7 (M/F), rarely painful and most frequently located proximally in the upper limbs [7]. In our case, it has actually affected a young man without any spontaneous pain but localisated in the lower limb, a cutaneous branch of the femoral nerve.

US imaging aspects are typically a fusiform swelling mingled with the nerve tissue. A well defined and hypoechoic centered mass with sharp borders and nerve fibers entering and exiting the ends of the swollen area. No anechoic areas but rare internal calcifications could be found [8,9]. It was the case of our patient.

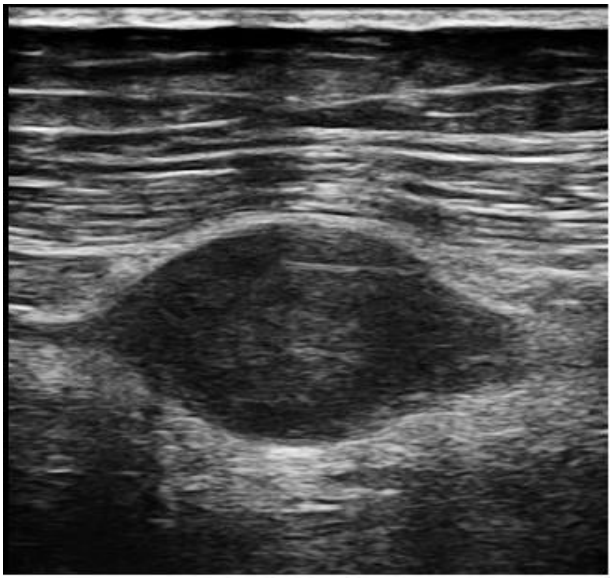

Fig-1 : An US image on longitudinal section shows the hypoechoic fusiform and well defined mass with sharp borders that seem to continue a linear nervous structure on the both sides

On MRI, neurofibromas appear elongated and centered to the nerve root, usually have an identical signal intensity to that of muscle on T1 sequences. After contrast medium injection, the enhancement is variable and usually moderate [10], while the target sign in T2 (low signal intensity on the central area and high signal intensity on the surroundings), initially described as pathognomonic, is often non-specific [9-11].

It's usually seen in neurofibromas because of their histological composition, the surrounding area corresponds to the presence of myxomatous tissue while the component of the central zone is fibrocollageous tissue. So how can we exclude the other differential diagnosis ?

Schwannomas are fusiform masses that arise eccentrically along the involved nerve unlike neurofibroma, and are encapsulated by an epineurium [12] while the plexiform neurofibroma is exclusively seen in Von Recklinghausen disease. That's led us to confirm the diagnosis of isolated neurofibroma.

CONCLUSION

In the summary, despite its rarity, the diagnostic of isolated neurofibroma should be kept in mind especially in young people with an evocative appearence on MRI.

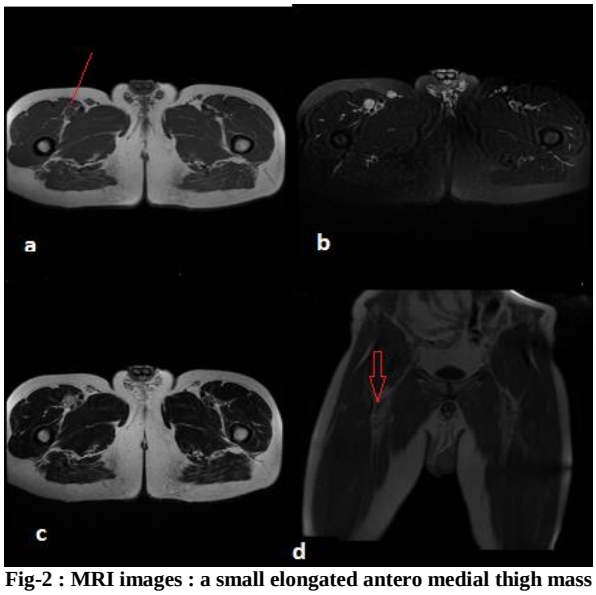

Fig-2 : MRI images : a small elongated antero medial thigh mass centered to the nerve root, a : Isosignal on axial T1 weighted sequence, b : Axial T1 FS WS : moderate enhancement after intravenous gadolinium injection, Moderate hypersignal on axial (c) and coronal (d) T2 weighted sequence

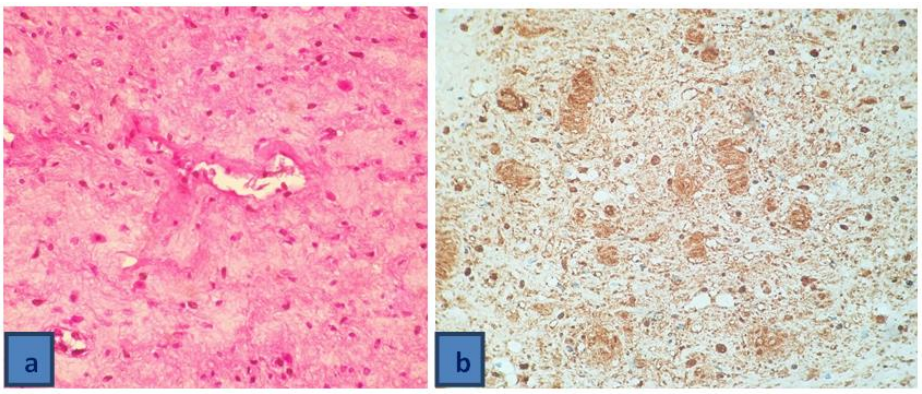

Fig-3: Histological examination, a: Neurofibroma, high grossissement (x40), b: immunohistochemical analysis showing a positive Ps 100 staining

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