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Case Report

Fibrolipomatous Hamartoma – Magnetic Resonance Imaging Diagnosis

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Abstract

The fibrolipomatous hamartoma is a benign process that causes fusiform nerve enlargement as a result of the proliferation of mature fibrous perineural tissue. It is a rare disease, diagnosed mainly in children and young adults, presenting few early manifestations, eventually leading to symptoms related to nerve compression - pain, paresis and paresthesia (eg carpal tunnel syndrome) - or tumor.

The gold standard method for your diagnosis is the magnetic resonance imaging - there is no need for biopsy. Thickened and low signal nerve bundles are observed, surrounded by fibrolipomatous tissue with a high T1 sign, and the pathognomonic findings of this entity. Its appearance is compared to that of a coaxial cable in the axial plane, or "spaghetti-like" in the coronal section.

Keywords: Hypesthesia; Wrist; Median Nerve; Hamartoma; Magnetic Resonance Imaging

Abbreviations:

MRI: Magnetic Resonance Imaging

Introduction

Fibrolipomatous hamartoma was first described by Mason in 1953 [1]. It is a benign lesion that causes fusiform enlargement of the nerve as a result of the proliferation of mature perineural fat and fibrous tissue [2].

It is a rare disease, diagnosed mainly in children and young adults, with few early manifestations, eventually leading to symptoms related to nerve compression - pain, paresis, and paraesthesia (eg, carpal tunnel syndrome) - or tumor [1,3,4].

The most frequently damaged is the nerve median nerve - 80% of the cases, almost always in the carpal tunnel [3,5]. However, it eventually occurs in the radial, ulnar, sciatic, plantar, or even lung nerves [4]. Usually, it is unilateral, with lipomatous macular dystrophy occurring in 27 - 66% [1,4].

Case Report

A 45-year-old woman with right numbness for a long time in her right hand. Has a positive Tinel sign on physical examination and no visible deformities or skin changes. She denies previous surgeries and trauma. Magnetic resonance imaging (MRI) of the right wrist demonstrates marked diffuse thickening of the median nerve, with the appearance of a "coaxial cable" in the axial plane with hyperintense signal comparing to muscles, being compatible with fibrolipomatous hamartoma (Figure 1). The patient was referred to the hand surgeon for treatment.

Figure 1: MRI T1-weighted sequence in the axial section demonstrating marked diffuse thickening of the median nerve, with the appearance of a "coaxial cable" (white arrow).

Discussion

There is no consensus about fibrolipomatous hamartoma etiology, presenting two main theories [1,3]:

- Congenital (non-genetic).
- Inflammatory: this one would be caused by nerve irritation or trauma, causing it´s grown.

The gold-standard method for its diagnosis is MRI - no biopsy is required [4]. Thick, low-signal nerve bundles surrounded by high-signal T1 fibrolipomatous tissue are observed, and the pathognomonic findings of this entity [4]. Its appearance is compared to a coaxial cable in the axial plane or a “spaghetti” in the coronal section [3].

Differential diagnoses include ganglionic cyst, intraneural lipoma, vascular malformation, hereditary hypertrophic neuritis of Dejerine-Sottas syndrome, neurofibroma, and traumatic neuroma [3].

There is no definitive treatment for fibrolipomatous hamartoma, but open carpal tunnel decompression associated with hamartoma’s tissue removal seems to be related to the improvement of the symptoms [1]. In 33 - 60%, of cases, there may be a recurrence of nerve damage after surgical treatment [1].

Conclusion

We described the case of a patient with carpal tunnel syndrome caused by median nerve fibrolipomatous hamartoma, pointing out the typical findings of this pathology on MRI.

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