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Clinical case

Lipofibromatous hamartoma of the radial nerve: An unusual location

Le neurofibrolipome du nerf radial : au sujet d'une atteinte rare

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Abstract

Lipofibromatous hamartoma is a congenital and ineradicable benign tumour of the peripheral nerve sheaths, affecting almost exclusively the median nerve and its branches. It corresponds to an infiltration of the nerve by lipofibramatous tissue that dissociates the fascicles. We report a highly unusual case of a lipofibromatous hamartoma of the radial nerve in the upper extremity in a 52-year-old female patient.

Keywords: Lipofibromatous hamartoma; Peripheral nerve sheath tumor; Surgery; Radial nerve

Résumé

Le neurofibrolipome est une tumeur bénigne, inextirpable et congénitale des gaines des nerfs périphériques, affectant presque exclusivement le nerf médian et ses branches de division. Elle correspond à un envahissement du nerf par un tissu fibroadipeux dissociant les fascicules. Nous rapportons le cas d'une atteinte exceptionnelle d'un neurofibrolipome, du nerf radial au bras, chez une patiente de 52 ans.

Mots clés : Neurofibrolipome ; Tumeur nerveuse périphérique des gaines ; Chirurgie ; Nerf radial

1. Introduction

Lipofibromatous hamartoma is a benign tumour of the peripheral nerve sheaths, affecting primarily the median nerve and its branches [1,2]. This ineradicable tumour is characterised by lipofibramatous infiltration of the nerve that dissociates the fascicles [2]. Radial nerve damage is rare, with only four cases reported in the literature [3–6]. The case presented below concerns a lipofibromatous hamartoma of the radial nerve in the upper extremity a 52-year-old female patient.

2. Case report

A right-handed 52-year-old healthcare assistant presented with a loss of strength in her left arm that had been progressively increasing over two years. The patient had no particular antecedents, other than having stopped smoking several years previously.

Clinical examination revealed a very voluminous palpable cord on the right side of the arm, in the distal third, along the course of the radial nerve. The patient suffered radial paralysis with neither sensory loss nor neurological pain. A complete motor deficit was observed for the extensor digitorum. The supinator, extensores carpi radialis and extensor carpi ulnaris enabled movement against gravity, but not against resistance. The extensor pollicis longus, extensor indicis proprius and extensor digiti minimi did not allow the patient to perform movement against resistance. The triceps brachii was not affected.

Initial electromyography (EMG) showed severe denervation of the radial nerve with an absence of motor response except for the triceps brachii. Sensory function was not explored at that point. Magnetic resonance imaging (MRI) revealed fatty

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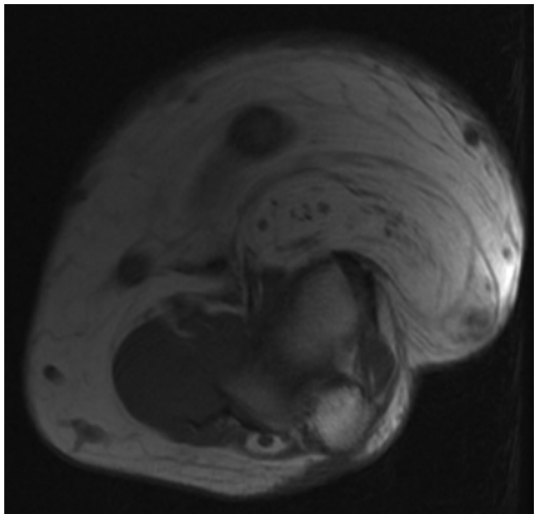

Fig. 1. MRI T1-Fatsat axial view with gadolinium injection. The radial nerve appears like a "spaghetti sack". In T1 sequences, fatty tumour tissue is hyperintense and the fascicles, hypointense.

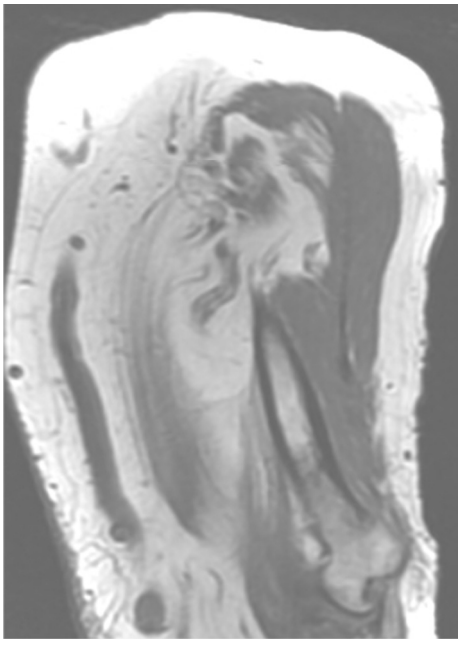

Fig. 2. MRI T1 weighted sagittal view. The radial nerve appears like "cables". In T1 sequences, fatty tumour tissue is hyperintense and the fascicles, hypointense.

tumour dissecting the fibres of the radial nerve and measuring 6 cm wide, 2 cm thick and 20 cm long with no suspicious contrast enhancement. The classic "spaghetti sack" image was seen in axial views (Fig. 1) and a "cable" form in views (Fig. 2). In the T1 weighted image sequences, the nerve fibres appeared as hyposignals, interspersed with hypersignal tumoral tissue.

Surgery was carried out under general anaesthesia with optical magnification (loupes). A lateral approach allowed us to detect a voluminous tumoral structure corresponding to the radial nerve. Nerve diameter became normal before bifurcation into superficial and deep branches. We performed neurolysis of the radial nerve along the entire length of the affected area, associated with decompression by epineurotomy following the axis of the tumour. A fusiform, yellowish tumour was identified with clear borders. The tumour infiltrated to the structure of the nerve, pushing back and sheathing the fascicules. The latter retained a healthy aspect, aside from one hollow bundle. Neurostimulation of the different fascicle bundles within the tumour made it possible to observe motor responses in all muscle groups, aside from the extensor digitorum. Five biopsies were taken along the length of the tumour, including tumoral tissue itself and the epineurium. Fascicles were spared, other than those considered to be hollow.

During the initial postoperative period, paresthesia was observed in the radial nerve territory, with no worsening of motor deficit. This sensation disappeared in less than two weeks.

The pathology report on the biopsies revealed a normal nerve structure, surrounded by mature adipose tissue without atypia (Fig. 3). The diagnosis of lipofibromatous hamartoma was thus confirmed.

As regards motor recovery, 19 months postoperatively the outcome of this radial nerve decompression at the level of the

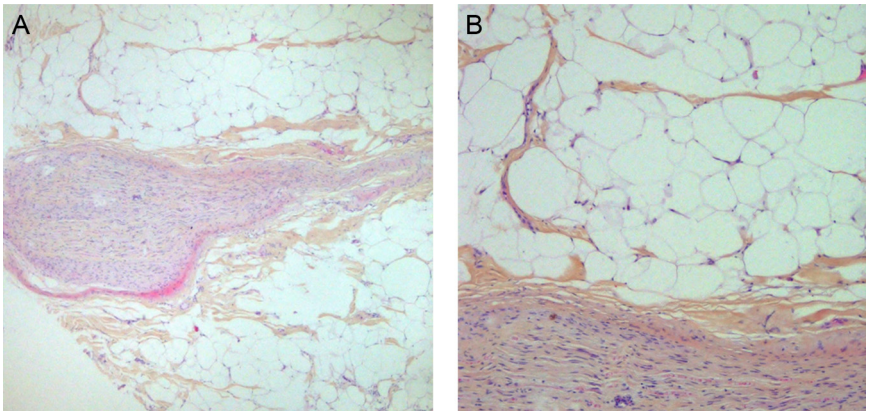

Fig. 3. Pathology slides of lipofibromatous hamartoma after HES staining. With minimal optical magnification (×40), the lesion consists of mature adipocytes of regular calibre, surrounding a central nerve trunk (A). With intermediate magnification (×200), adipocytes are mature, of regular calibre without atypia; a nerve element is visible at the bottom (B).

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