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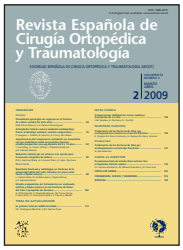

CASE REPORTS

Multiple median nerve schwannoma: a case report

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KEYWORDS
Multiple schwannomas;
Median nerve;
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Hand

Abstract
Introduction: The bulging masses and paresthesias caused by multiple schwannomas are often confused with the symptoms of other more common pathologies of the hand: lipoma, ganglion, carpal tunnel syndrome.
Clinical case: We present a rare case of multiple median nerve schwannomas in a 20-year-old male. These tumors are not accompanies by signi cant neurological impairment.
Conclusions: Magnetic resonance imaging (MRi) is very useful in preoperative diagnosis and surgical planning. Microsurgical intracapsular dissection was performed satisfactorily and the tumor has so far not recurred.

PALABRAS CLAVE
Múltiples schwannomas;
Nervio mediano;
Tumor;
Mano

Schwannomas múltiples de nervio mediano: descripción de un caso

Resumen
Introducción: el abultamiento y las parestesias que originan los schwannomas múltiples suelen confundirse con otras enfermedades más frecuentes en la mano: lipoma, ganglión, síndrome de túnel carpiano.
Caso clínico: presentamos un caso poco frecuente de schwannomas múltiples del nervio mediano en un varón de 20 años. Este tipo de tumores no se mani esta con dé cit neurológicos importantes.
Conclusiones: la resonancia magnética es muy útil en el diagnóstico preoperatorio y en la plani cación de la cirugía. La disección intracapsular microquirúrgica fue realizada de forma satisfactoria, y en la actualidad no hay recidiva del tumor.

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Introduction

Schwannoma (also known as neurilemoma), the most frequent benign tumor of the peripheral nerves, is an encapsulated neoplasm that originates from Schwann cells. Its growth is slow, intraneural and eccentric. Forty- ve percent of these tumors have been detected in the head and neck area and 19% in the upper limbs.[1,2] They commonly affect the volar rather than the dorsal aspect. In general, they are solitary neoplasms; very few cases of multiple schwannomas in the same limb have been reported in the literature.[1-8]

The clinical manifestations of schwannoma are scarce during the initial stages. Paresthesias and bulging along the course of the peripheral nerve are the most usual symptoms. Neurological impairment is rare. The infrequency of this tumor, together with its erratic presentation, has led to a misguided diagnosis in 75-80% of reported cases.[9] Magnetic resonance Imaging (MRi) is useful for diagnosis and preoperative planning.[9,10]

The purpose of this study is to report on a very unusual instance of a multiple median nerve schwannoma and alert readers to the multiplicity of this kind of tumor, which makes it advisable to perform an MRi before a nal diagnosis is made.

Case clinic

The patient is a 20-year-old male, with no relevant family or personal history. He presented with a 1-year history of pain in his left forearm, which increased with pressure on the anterior aspect and the proximal third. There were no concomitant paresthesias or loss of strength in the territory of the median nerve. Phyisical examination revealed a positive Tinel sign along the course of the median nerve at the middle-third and upper portion of the forearm. Radiology and electromyography (EMG) were normal. A coronal MRi shows formation of new spindle-shaped schwannomas in the area covered by the median nerve at the middle third of the forearm, and more proximally, at the level fo the elbow. (g. 1A). The sagittal view shows nodular formations adjacent to the nerve in that area (g. 1B y C), indicating the presence of a left median nerve schwannoma. Under ischemia and block anesthesia, the tumor is excised. Microdissection is carried out with the aid of x3,5 magnifying loupes. The affected nerve bundles are identi ed and a lobulated 2x1 cm tumor in the middle third of the forearm (g. 2A) and another 2 1x1 cm tumors proximal to the antecubital exure are resected (g. 2B). The specimens are subjected to a pathologic and immunohistochemical study that con rmed the diagnosis of benign schwannoma. During post-op the patient experienced paresthesia in the median nerve territory, especially intense in the third nger, which gradually disappeared. At 2 years, there has been no recurrence of the lesion, and the patient remains clinically asymptomatic, with normal MRi and EMG.

Discussion

Schwannomas are generally solitary neoplasms and account for less than 5% of upper limb tumors.[9] Multiple schwannomas affecting a single nerve have rarely been reported. The incidence of multiple schwannomas in the published series[3]

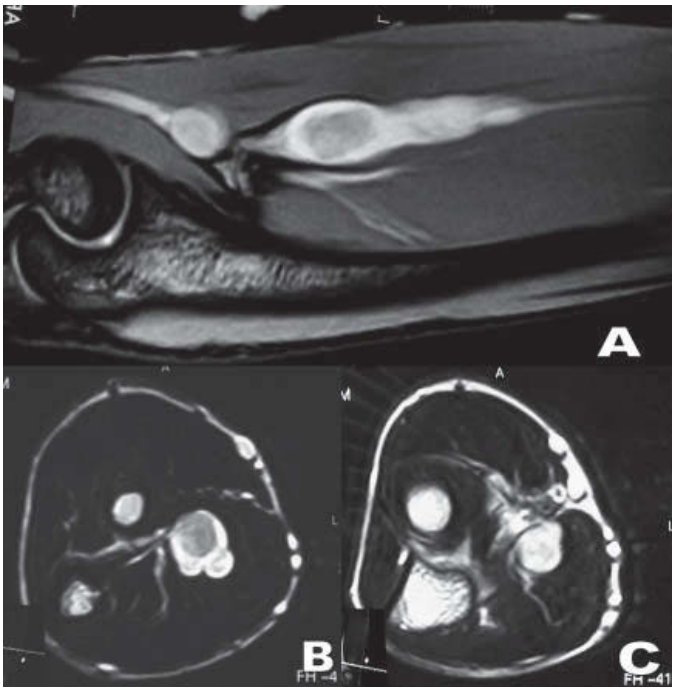

Figure 1 T2-weighted magnetic resonance scans. A: coronal view showing a spindle-shaped schwannoma encompassing the median nerve at the middle third of the forearm and a nodular schwannoma in the elbow. B: sagittal view showing a schwannoma developing eccentrically to the median nerve in the middle third. C: A schwannoma in the elbow.

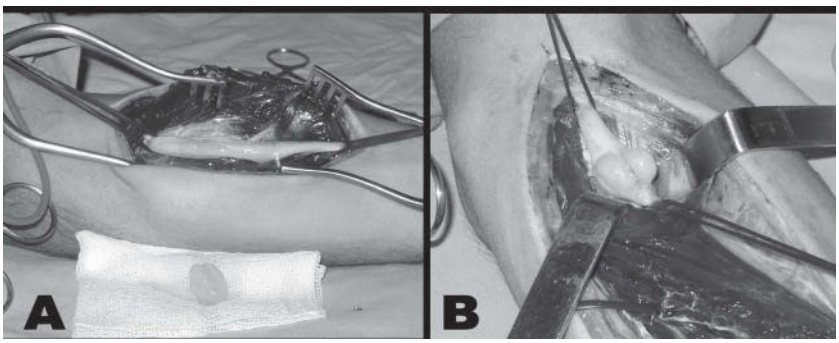

Figure 2 Intraoperative photograph. A: median nerve schwannoma dissected at the middle third of the forearm. B: 2 median nerve schwannomas on the anterior aspect of the elbow.

stands between 1 and 23% The ulnar and median nerves are more frequently affected than the radial nerve.[11] The most frequently affected age group is the one made up of individuals between 20 and 50 years of age and distribution among males and females is roughly even. The mean tumor size in the upper limb is 19 mm (range: 4–61 mm).[11]

Clinical diagnosis is dif cult because schwannomas share some symptoms with other more common hand and wrist pathologies. Paresthesias, pain, in ammation and Tinel's sign are not speci c to schwannoma and could lead the surgeon to make a diagnosis of schwannoma when the actual pathology is a lipoma, a ganglion, a neuro broma or carpal tunnel syndrome.[2,9-11] Neurological impairment is rarely present.

In the past, given their low incidence schwannomas were diagnosed on the basis of operative ndings. Since the advent of MRI diagnostic con rmation is much more certain. Even if MRI is not 100% speci c for the diagnosis of schwannoma, it is however useful to locate the neoplasm anatomically, it allows better preoperative planning and permits differentiation of solitary from multiple cases.[8] Neural tumors tend to be isointense to the muscle in T1-weighted sequences and hyperintense in T2-weighted ones. No doubt, the most speci c nding is a continuity of the mass with respect to the nerve. We can also nd other signs, such as the bull's eye sign, which refers to the hypersensitivity of the tumor's periphery with respect to the center in T2-weighted images, or the appearance of bundles within the tumor mass.[12] In schwannomas, the tumor may be located eccentrically in the nerve, but in neuro bromas it is always located in the center of the nerve. Lipomas, on the other hand, show an increased signal in T1 and T2-weighted images.

Histologically schwannomas are a benign proliferation of Schwann cells. There are 2 different histologic types, which may coincide in the same tumor: the Antoni A pattern, characterized by a high density of spìndled cells arranged in palisades, and the B pattern, where a smaller number of cells are separated by amorphous material. This type of tumor is strongly positive for S-100 protein.

The probability of damaging the nerve during surgery increases in proportion to the number of tumors detached and when tumors grow very close to one another in the same nerve.[7] In our case, treatment was based on a very careful microsurgical technique aimed at separating the nerve bundles from the tumor in a bloodless eld. In the case we are presenting, there was a portion of healthy nerve between the 2 schwannomas excised in the elbow and the one resected in the forearm. Reports in the literature recommend a longitudinal incision on the epineurium and the tumor capsule in order to perform an intracapsular excision and prevent a nerve lesion.

Prognosis is excellent following excision. Although recurrence is rare, some relapses have been reported in the literature.[8] In some cases, neurological sensitive impairment may persist in the form of paresthesias, which tend to disappear at 2 months from surgery. More severe motor de cits have also been reported.[4]

Conflict of interests

The authors have declared that they have no con ict of interests.

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