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Case report

Atypical schwannoma of the median nerve. A case report

Schwannome atypique du nerf médian. À propos d'un cas

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ABSTRACT

Schwannomas of the hand are very rare tumors and represent less than 3% of all soft tissue tumors in the hand. These tumors share clinical, epidemiological and imaging characteristics with the other soft tissue and peripheral nerve tumors; thus, it can be difficult to make a preoperative diagnosis. Here we report the case of a 48-year-old woman who presented with a schwannoma arising from the palmar branch of the median nerve. The tumor measured 54 × 41 x 52 mm and was located in the thenar eminence. The first hypothesis was a vascular tumor. After surgery and histological analysis, the final diagnosis of an atypical schwannoma was established. The presence of shared immunohistochemical characteristics with cellular histiocytoma and myoepithelial tumors forced us to adopt an aggressive follow-up protocol. As of the last follow-up at 9 years, the patient had good clinical outcomes and no recurrence. This case highlights the difficulties encountered in clinical practice to diagnose such tumors.

RÉSUMÉ

Les schwanommes de la main sont des tumeurs très rares et représentent moins de 3% de toutes les tumeurs des tissus mous de la main. Ces tumeurs ont des caractéristiques cliniques, épidémiologiques et d'imagerie partagées avec les autres tumeurs des tissus mous et des nerfs périphériques. Cela peut rendre difficile l'établissement d'un diagnostic préopératoire. Nous rapportons ici le cas d'une patiente âgée de quarante-huit ans qui présenta un schwannome provenant du rameau palmaire du nerf médian. La tumeur était localisée au sein de l'éminence thénar et ses dimensions étaient de 54 × 41 x 52 mm. La première hypothèse était qu'il s'agissait d'une tumeur vasculaire. Après son exérèse et son analyse histologique, le diagnostic final d'un schwannome atypique fut retenu. Mais la présence de caractéristiques immunohistochimiques communes aux histiocytomes cellulaires et aux tumeurs myoépithéliales nous força à adopter un protocole de suivi agressif. Au dernier recul, la patiente avait de bons résultats cliniques sans aucun signe de récidive. Ce cas clinique met en lumière les difficultés rencontrées en pratique clinique pour diagnostiquer de telles tumeurs.

Introduction

Schwannomas of the hand are rare in clinical practice [1,2], making up less than 5% of upper limb soft tissue tumors [1]. The clinical symptoms of schwannomas [1] and imaging findings on echography and magnetic resonance imaging (MRI) [3,4] are shared with other soft tissue tumors. Thus diagnosing schwannomas preoperatively can be challenging with a risk of erroneous diagnosis. The final diagnosis requires microscopic analysis using immunohistochemistry (IHC) [5]. While the IHC features of schwannomas have been well described [1], some atypical variants exist that can make it difficult to differentiate it from malignant tumors. We report the case of a rare hand schwannoma in a 48-year-old female patient with several atypical ICH findings.

Case report

A 48-year-old, right-handed woman working as a nursery assistant with a history of multiple benign lipomas presented with a large tumor in her right thenar eminence. The lesion had been

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present for 1 year and increased in volume during the past weeks. There was no family history of neurofibromatosis and no associated clinical features. Clinical examination of the hand revealed a soft, painless tumor of the thenar eminence with no sensory or motor deficits. No Tinel sign was present. Vascularization of the hand was preserved.

The patient's general practitioner had prescribed radiographs and a sonography, which showed no bone invasion but revealed vascularized elements within the tumor. To clarify these findings, arteriography was performed to look for any sign of arteriovenous malformation (AVM) (Fig. 1A). It showed a well-vascularized tumor taking its origin from the right deep palmar arch and numerous vascular collections within the tumor but without evidence supporting an AVM. The radiologist concluded on the potential presence of a hemangioma. MRI angiography revealed the presence of a large tumor, measuring 54 × 41 x 52 mm, close to the volar side of the thenar muscles, in contact with the median nerve but not including it, which was lobulated, well-delimited, with heterogeneous signal and hematic collections that were enhanced after gadolinium injection (Fig. 1B). The diagnosis proposed was a cavernous angioma.

Given the heterogeneous and atypical appearance of the lesion, a surgical biopsy was performed. Histopathological examination showed fusiform cells with multiple cellular rearrangement and multinuclear atypism. IHC analysis found an atypical profile with positive reaction for smooth muscle actin; however very few cells were positive for S-100 protein. There was no mitotic activity and very few cells were Ki67 positive. Two independent pathologists read the histology slides. Their conclusion was a possible atypical and cellular histiocytofibroma with a risk of sarcoma.

Based on these findings, complete resection was suggested and performed 3 weeks later by a senior surgeon under 4.3× magnification. The shape of the incision captured the previous biopsy site and was extended proximally with a "Z" incision to the proximal wrist crease (Fig. 1C). The carpal tunnel was released to access to the median nerve and expose all distal sensory branches as well as the recurrent branch (Fig. 1D). Exploration showed a tumor arising from the palmar branch of the median nerve. Intraneural fascicular dissection was impossible and the branch had to be sacrificed since it was included in the tumor. The distal nerve bundle was implanted into the thenar muscle to prevent neuroma formation.

Histopathological examination showed a fusocellular lesion that was well-delimited, well-encapsulated, with rich vascularization and hemorrhagic alteration. The mitotic index was low [with a maximum of 4 mitoses/10 high-power field (HPF)] but with heterogeneous expression of anti-Ki67 (maximum 10% of cells). IHC examination showed the tumor was widely positive for S-100 protein and GFAP but still diffusely positive for smooth muscle actin, with a few nuclei positive for anti-P63 and vessel wall positive for desmin. Two diagnoses had to be considered: cellular schwannoma with atypical features or myoepithelial tumor. After a multidisciplinary discussion including our local pathology team and the French National Sarcoma Group, the final diagnosis retained was an atypical cellular schwannoma.

The early follow-up was uneventful. Satisfactory healing and full functional recovery were obtained except for loss of sensation in the median nerve's palmar branch territory. The patient did not develop neuropathic pain. She was able to get back to work at 6 months postoperative. Her grip strength and key pinch were similar to those of the contralateral side (28 kg and 7 kg, respectively) and her QuickDASH Score was at 0/100. Clinical follow-up and MRI were done every 6 months for 5 years and then once every year. At 9 years' follow-up, there was no local recurrence or functional deficit.

Discussion

Schwannomas [representing 90% of all peripheral neural tumors (PNT)] [5] are rare and represent less than 8% of all benign soft tissue tumors [1]. Schwannomas of the median nerve therefore make up 0.1–0.3% of all hand tumors, and among them very few cases arising from the palmar branch of the median nerve have been described. The most frequent location of schwannomas seems to be in the upper limb [2], mostly on the volar side of the forearm and hand [1,6].

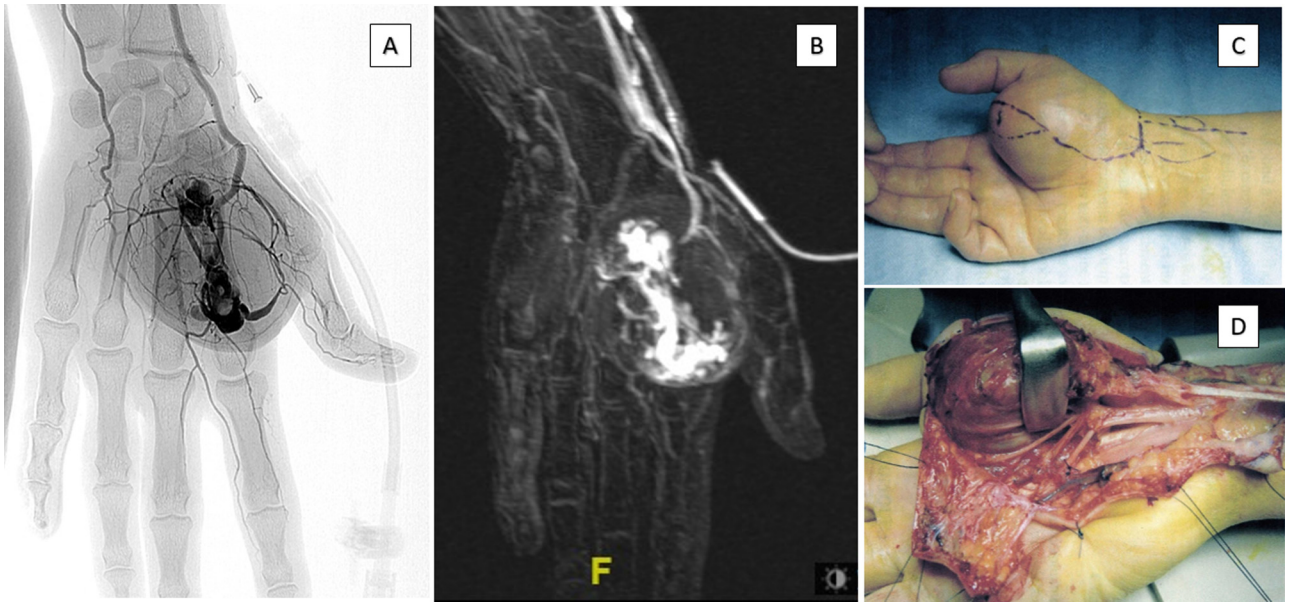

Fig. 1. Arteriography of the hand in the late arterial phase showing a well-vascularized tumor originating from the right deep palmar arch with numerous vascular collections within the tumor but without AVM, having the characteristics of a hemangioma (A). MRI in the coronal plane with signal enhanced after gadolinium injection showing a hematic collection within the tumor, with a heterogeneous and lobulated aspect and surrounded by a pseudocapsule (B). "Z" shaped incision that captures the biopsy site (C). Tumor dissection with median nerve sensory branches (D).

Despite their different histologic origin, benign soft tissue tumors such as PNT, vascular tumors, connective tissue tumors or muscle tumors have common epidemiological and clinical characteristics such as the occurrence in young and middle-aged adults [2,6], most common location in the extremities and trunk, size which is seldom more than 2 cm and presentation which is often a palpable painless mass but with possible compression symptoms. For this reason, identifying them in routine clinical practice can be challenging [1,2]. Furthermore, the Tinel sign classically described in schwannoma is absent most of the time [7].

Moreover schwannoma imaging descriptions are variable [4]. On ultrasonography, the smallest tumors are usually a homogenous mass with cystic content but in larger tumors, as the one described in our case, the tumor becomes heterogeneous with calcifications, hemorrhage and fibrosis [4]. This can lead to an erroneous diagnosis of ganglion, lipoma or fibroma in the smallest lesions and angioma-like lesions in the biggest lesions, as in our case. The best argument in favor of schwannoma is nerve continuity within the tumor but this can be missed in thin nerves. Another possibility is the ultrasonographic Tinel's sign, but it is rarely found in practice [4,7]. MRI angiography is considered the gold standard for PNT imaging [3], with a correct preoperative diagnosis up to 91% of cases [7]. Its high definition allows for better identification of nerve continuity in the small nerves [7]. The signal of the mass is hypo-intense or iso-intense on T1-weighted images and hyperintense, heterogeneous or not, on T2-weighted images, depending on intrinsic changes in the tumor [3]. In our case, MRI angiography showed an heterogeneous and hypervascularized tumor (which can also be related to the schwannoma's capsule often surrounded by a rich vascular network [2]); however the nerve branch could not be identified within it. This contributed to the misdiagnosis of the lesion as a vascular tumor.

In this tumor, the presence of unusual IHC markers made the diagnosis challenging. Indeed, the first biopsy showed very few cells expressing S-100 protein, whereas they are usually strongly expressed by schwannomas [5].There was a large response to smooth muscle actin and no expression of CD-34, which was consistent with the microscopic appearance of a cellular histiocytoma. The IHC profile of the resected tumor was compatible with an atypical schwannoma based on the positive S-100 protein and GFAP staining. On the other hand, the tumor was still diffusely positive for smooth muscle actin, a few nuclei were positive for anti-P63 and the vessel wall was positive for desmin, with a mitotic activity index of up to 4 mitoses/10 HPF, which is consistent with a myoephithelial tumor [8]. Up to 10% of cells also expressed anti-Ki67, which is correlated to cellular proliferation, and is often low ($<$1%) in schwannomas [5]. Macroscopically, the tumor was encapsulated and polycyclic. Along with its location inside the median nerve, these findings were taken into consideration to arrive at the final diagnosis of an atypical schwannoma.

Schwannoma atypical variants have been studied in their epithelioid form [9] and are defined by their high mitotic rate ($\geq$ 3 mitoses/10 HPF) and nuclear size variation as observed in our case. The issue concerning this sub-type is not the malignant transformation (very low possibility) but the erroneous diagnosis with an epithelioid malignant peripheral nerve sheath tumor or any other malignant tumor. Indeed a misdiagnosed malignant tumor can lead to local and general recurrence whereas a misdiagnosed benign tumor can lead to inappropriate large tumor resection or irradiation. For this reason, an aggressive follow-up protocol was used with our patient.

The present case also highlights the risk of erroneous diagnosis after a biopsy and even more after needle aspiration because of the possible heterogeneity of these tumors [2].

Schwannomas are treated by surgical resection with intraneural dissection. In some difficult cases, there is no clear macroscopic separation between tumor and nerve fascicles with bundle infiltration [2,7,10]. In these cases, the fascicles must be excised during the procedure to ensure complete tumor resection [6] as in our case. Mizushima et al. [10] reported that 15 of 43 patients developed permanent, newly acquired neurological deficits after schwannoma resection. An average of 50% of patients develop neurological deficits after surgery [10]. This risk seems to be higher in plexiform subtypes [2], in the most proximal locations and in large tumors [10]. Thus this must be included in the preoperative patient information and consent discussion, especially in patients without preoperative sensory deficits.

Schwannomas are rare tumors and those located in the hand are even more infrequent. Their clinical presentation and histological properties are well described. However this tumor shares common clinical, epidemiological and imaging characteristics with the other soft tissue tumors. In some rare cases, it can present atypical histological and IHC content that should not be confused with a malignant tumor. Consequently, care must be taken to correctly identify the tumor with clinical, imaging and histological evidence in order to arrive at the proper treatment and follow-up. Additionally, clear information should be given to patients about the possible postoperative neurological deficit.

Disclosure of interest

The authors declare that they have no competing interest.

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