Schwannoma of the Median Nerve at the Wrist-A Case Report and Review of Literature

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Abstract

Benign tumours involving peripheral nerves of the upper extremity are uncommon. Schwannomas also known as neurolemmas are usually originated from Schwann cells located in the peripheral nerve sheaths. It is generally presented as an asymptomatic mass and its slow evolution remain an essential factor in diagnosis delays. Tumors with a long evolution and relatively large dimensions can undergo degenerative changes such as cyst formation, calcification, hemorrhage and fibrosis and are described as ancient schwannomas which can be misdiagnosed as sarcomas due to specific imaging and histologic findings.

We report a rare case diagnosed as ancient schwannoma of the median nerve in a 70-year-old male. We describe the clinical presentation, the specific imaging, histology, surgical findings and functional outcome. This tumor has a good prognosis with a low recurrence rate and potential for malignant transformation. Surgical removal is usually curative.

Keywords: Ancient schwannoma; Median nerve; peripheral nerve sheath tumors; Neurilemoma

Introduction

Peripheral nerve tumors are uncommon lesions categorized into primary neuronal, nerve sheath, and non-neuronal neoplasms. Nerve sheath tumors are the most common. Schwannoma, also known as neurilemoma, is a benign soft tissue tumour arising from the Schwann cells of the nerve sheath derived from the neural crest [1,2] and usually affects patients aged 20-50 years without race or sex predilection and accounts for approximately 5% of all benign soft tissue tumors [3].

Solitary schwannoma is a slow-growing tumor and it is often found incidentally as a painless mass for years before the onset of pain and neurological symptoms caused by compressing surrounding tissues and peripheral nerves. Tumors with a long evolution and relatively large dimensions can undergo degenerative changes such as cyst formation, calcification, hemorrhage and fibrosis and are described as ancient schwannomas [4] which can be misdiagnosed as sarcomas due to specific imaging and histologic findings. We report on a very rare case diagnosed as ancient schwannoma of the median nerve. We describe the clinical presentation, the specific imaging, histology, surgical findings and functional outcome.

Case Report

Figure 1: Swelling on the volar side of the right wrist with paresthesia in the corresponding innervation of the median nerve.

A 70 years old man presented with a painless swelling in the volar side of the right wrist of 18 years duration with complaints of increase in size of the swelling associated with pain for the
past two years. He complained of pain and paraesthesia in the median nerve distribution area of the right hand. The swelling (Figure 1) was mobile on the transverse axis and not on the longitudinal axis. There was loss of sensation in the distribution of the median nerve in the hand with weakness in performing her daily activities. Percussion over the nerve produced positive Tinel’s sign. An ultrasound (US) scan revealed an encapsulated, heterogeneous, solid mass, mainly hypoechoogenic, with regions of increased echogenicity.

A 1.5-T superconductive Magnetic Resonance Image (MRI) unit (GE Healthcare) was used to produce spin-echo images and a gadolinium-enhanced scan to confirm the presence of a well-circumscribed and encapsulated soft tissue mass, which showed low signal intensity on the T1-weighted sequences (Figure 2a) and high signal intensity in T2 images (Figure 2b). On T1 fat suppressed images, increased gadolinium enhancement was noticed at the periphery of the mass with a non-enhancing low signal in areas of hemorrhage or degeneration. Based on the long history of the patient, and the clinical, US, and MRI features, the diagnosis of a nerve sheath tumor arising from the median nerve was supported and the patient was scheduled for excisional biopsy.

The patient was taken up for surgery under general anesthesia and a longitudinal incision centered over the tumor bulk was performed. On exploration was found a 3 cm long well encapsulated tumor arising from the median nerve in an eccentric position to the axis of the nerve (Figure 3a). A marginal tumor excision was performed under magnification in order to preserve the median nerve (Figure 3b) and the specimen was sent for histopathological examination.

**Histologic examination**

Histology confirmed the presence of a well-defined neoplasm surrounded by a fibrous capsule focally connected to a displaced parent nerve. The tumor composed of fusiform cells with spindle-shaped, tapered nuclei with inconspicuous nucleoli and eosinophilic cytoplasm (Schwann cells) showed the typical biphasic pattern of Antoni A (Figure 4a) and Antoni B (Figure 4b) areas. The first one, referred to as Antoni A, was more densely cellular and consisted of Schwann cells compactly disposed in broad bundles and interlacing fascicles. In these areas some nuclei were grouped into palisaded clusters. The second one, named Antoni B, was loose-textured due to the presence of a myxoid matrix. Areas of haemorrhage and perivascular hemosiderin deposition were seen, but coagulative necrosis was not observed (Figure 4c) the vessels showed typically hyalinized walls (Figure 4d). Although remarkable nuclear pleomorphism was not a feature, some cells showed larger, hyperchromatic nuclei, with occasional pseudoinclusions. Mitotic figures were not found.

**Surgical procedure**

The patient was taken up for surgery under general anesthesia and a longitudinal incision centered over the tumor bulk was performed. On exploration was found a 3 cm long well encapsulated tumor arising from the median nerve in an eccentric position to the axis of the nerve (Figure 3a). A marginal tumor excision was performed under magnification in order to preserve the median nerve (Figure 3b) and the specimen was sent for histopathological examination.
resumed her previous activities 4 weeks after surgery. Five months after surgery, the patient was asymptomatic and without any local signs of recurrence.

Discussion

Schwannomas are common, slowly growing, and encapsulated benign nerve sheath neoplasms separated from the surrounding tissues. These tumors are soft in consistency, mobile in nature, and sometimes painless so they may be misdiagnosed as lipoma, fibroma, ganglion, or xanthoma but the present case report suggests that the combination of ultrasound and MRI findings provides features that narrow the differential diagnosis.

On pathologic analysis generally displays a biphasic pattern, with areas of highly ordered cellularity (Antoni type A) and less cellular areas in which a highly myxoid matrix predominates (Antoni type B). Although schwannomas of the hand and wrist are well reported [5-8], few reports of ancient schwannomas exist [9-12].

Ackerman and Taylor in 1951 [4] described ancient schwannoma with clear areas of hypo cellular tissues due to the long standing degenerative changes attributed to the growth and “aging” of the tumor hence called ancient schwannoma. An ancient schwannoma is described [13] as a benign schwannoma having severe degenerative changes that include cyst formation, calcification, hemorrhage, and marked hyalinization. Schwannomas can be asymptomatic or can produce pain, a positive Tinel’s sign or a Tinel’s like sensation and sensory alterations [14-16]. The slow growth pattern of benign nerve tumors, allows for adaptation of the nerve function to the pressure effects.

The mainstay treatment is complete excision, although local recurrence can occur in large and incompletely excised lesions. Malignant transformation of benign schwannomas is unusual [17,18].

Conclusion

Ancient schwannoma of the median nerve is a rare peripheral nerve tumor where the combination of ultrasound and MRI findings provides features to diagnosis. Surgical resection must be approached with caution to protect nerve function and continuity and it is associated with good outcomes. The recurrence rate is low.

Author’s disclosure statement

The authors report no actual or potential conflict of interest in relation to this article.

References
