CASE REPORT

Ancient schwannoma of the thigh mimicking a malignant tumour: a report of two cases, with emphasis on MRI findings

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ABSTRACT. Ancient schwannomas are rare, encapsulated tumours of long duration and are benign in nature. The tumour is solitary and may grow to a large size before detection of notable degenerative changes. The term “ancient schwannoma” is used to describe a tumour that has undergone such changes, typified by relative loss of Antoni type A tissue, perivascular hyalinisation, calcification, cystic necrosis, haemorrhage and the presence of degenerative nuclei that may be misinterpreted as sarcomatous pleomorphism. We report two cases of ancient schwannoma in the thigh mimicking malignancies. Identifying the fibrous capsule of the mass and a split fat sign using MRI is important for differentiating ancient schwannoma from other malignant tumours.

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Ancient schwannoma is an uncommon schwannoma variant with a course typical of a slow-growing neoplasm [1]. The term “ancient” was proposed to describe a group of neural tumours showing degenerative changes and diffuse hypocellular areas [2]. These lesions are often erroneously diagnosed as malignant tumours, because they present with atypical features [1, 3]. MRI findings in patients with ancient schwannomas of the thigh have been rarely reported in the literature. This case report describes two patients with ancient schwannoma of the thigh mimicking malignant tumours, focusing particularly on MRI findings that are useful in pre-operative differential diagnosis.

Case report

Case 1

A 53-year-old woman presented with a rapidly growing palpable mass in the left thigh of one year's duration. She showed no tenderness and had no history of trauma. On physical examination a 15 × 10 cm non-tender, soft, movable mass in the anterolateral aspect of the thigh was found. Plain radiography showed an unmineralised soft-tissue mass with no bony involvement. Sagittal fast-spin echo, T_1 and T_2 weighted, and coronal and axial T_1 and T_2 weighted MR images were obtained using a 1.5 T instrument (Intera; Philips Medical Systems, Best, the Netherlands). The images showed a well-defined heterogeneous mass of low T_1 but high T_2 signal intensity in the intermuscular area between the rectus femoris, vastus intermedius and lateralis muscles

(Figure 1). The mass showed slightly increased signal intensity focus on T_1 weighted imaging (T1WI), and a low-signal intensity focus on T2WI. On post-contrast T1WI, the mass showed slight heterogeneous enhancement with central non-enhanced foci. A rim of fat surrounding the mass, suggesting the “split fat sign”, was also seen. An excisional biopsy was performed. The mass was located in the anterior aspect of the proximal thigh between the rectus femoris and the vastus lateralis muscles and measured 12 × 6 × 5 cm; it was firm, movable and yellow/dark greenish in colour. There was no definite adhesion to adjacent tissue. Inside the mass, a haemorrhagic, discoloured area was noted. On gross examination, the specimen, received in formalin, was an encapsulated solid mass attached to some adipose tissue, measuring 10.5 × 5.5 × 4.0 cm. The external and cut surfaces were focally haemorrhagic. On sectioning, the mass was tan in colour, myxoid and soft in consistency, and showed multiple foci of haemorrhage and necrosis. Microscopic study revealed low cellularity, wavy spindle cells with ropy collagen fibres, multifocal infiltration by mononuclear inflammatory cells and some haemosiderin-laden macrophages. Multiple microcyst formation, haemorrhage and necrosis were seen. There were scattered ectatic irregular vessels with surrounding hyalinisation. Differentiated Schwann cells immunohistochemically expressed the S100 protein. A diagnosis of ancient schwannoma was made (Figure 2).

Case 2

A 48-year-old woman presented with a slowly growing palpable mass in the left thigh of 20 years' duration. She showed no tenderness and had no history of trauma. MRI showed a well-defined soft-tissue mass at the posterolateral aspect of the mid-to-distal thigh. The mass

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Figure 1. Patient 1. MR images of a 53-year-old woman with ancient schwannoma in the intermuscular area between vastus intermedius, rectus femoris and lateralis muscles. (a) Coronal T_1 weighted MR image shows a large, well-defined mass with slightly increased signal intensity including a high-signal intensity area (arrow). A rim of fat surrounding the mass (arrowhead) is seen, known as the split fat sign. (b) Axial T_2 weighted MR image reveals heterogeneous, mainly hyperintense, signal, including dark signal area in the mass and a peripheral low-signal rim of tumour (arrowhead), suggesting fibrous capsule. (c) Gadolinium-enhanced T_1 fat-suppressed axial image shows heterogeneous enhancement, including non-enhanced foci and peripheral rim enhancement (arrow).

showed intermediate T_1 signal intensity and heterogeneously high T_2 signal intensity, with multiple internal low-signal foci. The mass showed good enhancement but contained several non-enhanced necrotic foci. The lesion had a low-signal peripheral rim with the split fat sign (Figure 3a). The mass involved the long head of the biceps femoris muscle. An excisional biopsy was performed. An encapsulated mass about $6 \times 5 \times 4$ cm was seen in the biceps femoris muscle, and included fibrotic, multifocal, necrotic foci without neurovascular invasion. The mass, received in formalin, was

encapsulated, measuring $5.5 \times 4.0 \times 3.0$ cm. The cut surface was yellowish-grey, of variegated appearance, with scattered haemorrhagic foci. Microscopic findings included Antoni A cells, showing nuclear palisading alternating with Verocay bodies and an Antoni B area. Tumour cells were wavy and spindle shaped, with vesicular to hyperchromatic nuclei. The cellular area showed marked fibrinoid deposition, hyalinisation, cystic changes, haemorrhages and microcalcification (Figure 3b). Some vessels were surrounded by hyalinisation. The final diagnosis was ancient schwannoma.

Figure 2. Patient 1. Gross (a) and histological findings (b, c). (a) This encapsulated solid-to-cystic mass shows extensive foci of degenerative change, including a myxoid area, necrosis and haemorrhage. (b) The mass is surrounded by a thick fibrous capsule attached by some peripheral nerve bundles. The inner area of the capsule is a marked degenerative myxoid change (haematoxylin and eosin $40\times$). (c) This tumour is diffusely positive immunostaining for S100 protein.

Figure 3. Patient 2. MRI (a) and histological (b) findings in a 48-year-old woman with ancient schwannoma in the intramuscular area of the long head of the biceps femoris muscle. (a) Coronal T_2 weighted MR image reveals heterogeneous, mainly high-signal intensity areas, including dark-signal foci and peripheral low-signal rim (arrow). (b) Histological finding of this tumour show focally palisading nuclei of spindle cells represented by a Verocay body, marked myxoid degeneration and hyalinisation (haematoxylin and eosin 40 $\times$).

Discussion

Schwannoma, also termed neurilemmoma or neurinoma, is a benign nerve sheath tumour consisting of two components. Antoni A areas are more organised and are hypercellular, and are composed of spindle cells arranged in short bundles or interlacing fascicles. Antoni B regions are hypocellular, less organised and contain more myxoid, loosely arranged tissue, with a high water content. These components are intermixed within schwannomas and occur in varying amounts [4]. Schwannoma is a slow-growing benign tumour and may be a large tumour with degeneration, especially when the tumour is situated in the deep regions such as the mediastinum and retroperitoneum [5]. Schwannoma with pronounced degenerative changes is known as ancient schwannoma, which is a rare variant of schwannoma and is usually a deeply situated large mass of long duration [5, 6], representing 0.8% of all soft-tissue tumours [7]. The term “ancient neurilemmoma” was first suggested by Ackerman and Taylor [8] in a review of 48 neurogenic tumours of the thorax. The cited authors reported 10 patients with tumours showing features similar to those of typical neurilemmomas, but differing in that significant tumour portions contained only a few cells within hyalinised matrices. They found that these features occurred in schwannomas of long duration, and hence coined the term “ancient schwannoma”. Such tumours are characterised by diffuse hypocellular areas, relative loss of Antoni type A tissue,

focal accumulations of hyaline material, calcification, cystic necrosis, haemorrhage and fatty degeneration [2, 9]. In addition, as the tumours are usually infiltrated by large numbers of siderophages and histiocytes, and display cellular degenerative changes, including nuclear atypia and pleomorphism, along with a tendency to nuclear palisading [5, 10], malignancy may be erroneously diagnosed [8]. On histological tumour examination, intense immunostaining for S100 protein suggests a neural origin and is helpful in diagnosis, especially of a totally cystic degenerated mass [5, 6].

Because the tumour contains cystic areas, ancient schwannoma has been radiologically misdiagnosed as other tumour types, such as malignant fibrous histiocytoma, malignant peripheral nerve sheath tumour, liposarcoma, synovial sarcoma or haemangiopericytoma. However, only a few reports have dealt with the radiological features of ancient schwannoma in the thigh because the tumour is so rarely encountered [2]. On MRI, the peripheries of ordinary schwannomas yield low-signal intensities on T_1 weighted images and high-signal intensities on T_2 weighted images, corresponding to the Antoni B region, and low-to-intermediate signal intensities in other areas on T_1 and T_2 weighted images with strong enhancement by gadolinium contrast media, a characteristic of the Antoni A regions [2]. The ancient schwannoma, however, shows a well-circumscribed inhomogeneous signal intensity and a different enhancement pattern. The relative amounts of Antoni A and B areas vary, and these regions are randomly blended,

reflecting such MRI findings [6]. Isobe and colleagues [2] reported imaging findings of seven ancient schwannomas situated mainly in the extremities as well-circumscribed complex cystic masses. The histological findings showed that the Antoni type B area occupied most of the tumours; the Antoni type A area had become smaller and was visible in the area around the degenerative lesions. The cited authors suspected that Antoni type A areas had degenerated to haematomas or cysts. In our cases, haemorrhagic changes were demonstrated on MRI. The first case showed a well-defined cystic mass with enhancement of the fibrous capsule on MRI, and histology showed an Antoni type B area in most of the tumours. The second case revealed a well-defined inhomogeneous signal intensity solid mass with random cystic areas.

The target sign of a peripherally hyperintense rim and central low intensity on T_2 weighted imaging has been reported in peripheral nerve sheath tumours [4, 11]. This MRI finding corresponds histologically to peripheral myxomatous tissue and central fibrocollagenous tissue [4, 11] and is absent in lesions with cystic, haemorrhagic or necrotic degeneration. In our cases, the target sign was not seen.

The fibrous capsule, consisting of epineurium and residual nerve fibres, appears as a hypointense rim on T_1 and T_2 weighted images and shows contrast enhancement on post-contrast T_1 weighted images [6]. According to the findings of Isobe and colleagues [2], the circumferences of the degenerated area and fibrous tumour capsule enhanced on MRI are important for discriminating an ancient schwannoma from other tumours, even though there are few reports of the radiological features of retroperitoneal ancient schwannomas [12, 13]. Our cases showed hypointense fibrous capsules and enhancements.

A rim of fat (split fat sign) is often present around deep-seated neurogenic neoplasms and has been previously described on CT scans [4]. Because each neurovascular bundle is surrounded by fat, masses arising in such bundles maintain a rim of fat as they slowly enlarge. Although not a specific sign for peripheral nerve sheath tumours, such a finding suggests that the tumour originated in the intermuscular space around the neurovascular bundle; neurogenic neoplasms are the most common such tumours. The split fat sign is more common in benign nerve sheath tumours and lesions of large nerves, whereas malignant nerve sheath tumours less frequently demonstrate complete fat rims, reflecting the more infiltrative growth patterns of such tumours [4]. Both our cases showed a split fat sign, which was considered as a characteristic finding reflecting benign neurogenic tumours.

A pre-operative differential diagnosis of a comparatively large soft-tissue tumour in a thigh, appearing well enhanced and with inhomogeneous signal intensity on MRI, tends to identify the tumour as malignant. Differentiation of ancient schwannoma from a malignant tumour can be achieved by considering the characteristic

clinical findings and radiological features of ancient schwannoma, as described by Isobe and colleagues [2]. First, there should be a mass with a long-term clinical course presenting with symptoms of a neurogenic tumour. Second, the mass should be palpable with local pain and characteristics similar to Tinel's sign, and should have distinct radiological features including a circumscribed degenerative area and an enhanced fibrous tumour capsule. However, one of our patients had a tumour history of only 1 year, and neither case showed tenderness. If the mass is deep in an extremity, the patient tends to ignore it until it finally reaches an excessive size, but a long-term clinical history is not essential for ancient schwannoma diagnosis. In our patients, the most characteristic MRI findings were a well-defined mass with a fibrous capsule, showing the split fat sign.

In conclusion, two cases of ancient schwannoma in the thigh are described. Two MRI features are important for differentiating ancient schwannoma from other malignant tumours: the mass should have a fibrous capsule and the split fat sign should be evident.

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