Unusual presentation of more common disease/injury

Solitary fibrous tumour of the lower leg: an uncommon site with atypical histopathological features

Rana K Sherwani, Amit Kumar

Department of Pathology, J N Medical College, AMU, Aligarh, Uttar Pradesh, India

Correspondence to Rana K Sherwani, prof.ranasherwani@gmail.com**Summary**

A 40-year-old man of low socioeconomic status presented with a slowly growing painless mass for 1 year at the antero-medial aspect of the right leg. On local examination, the overlying skin was normal and the mass was soft to firm in consistency, non-tender and freely mobile with no fixity to underlying structures. MRI revealed a lobulated mass with central areas of necrosis and no involvement of underlying muscles and neurovascular bundles suggesting a benign soft tissue tumour. The tumour was totally excised and submitted for histopathological examination, which, along with immunopositivity for CD34, CD99 and focally for Bcl2, led to a confirmatory diagnosis of solitary fibrous tumour with atypical histopathological features of increased mitosis and necrosis. After 4 months of follow-up, the patient was disease free without any recurrence or metastasis.

BACKGROUND

Solitary fibrous tumours, originally recognised as lesions of the pleura, have been documented in many other sites including extremities. The numbers of cases involving distal extremities are few and the differential diagnosis includes various soft tissue tumours both benign as well as malignant ones. Solitary fibrous tumours correspond to the relatively benign end of the spectrum and respond well to surgical excision alone. On the other hand, the aggressive sarcomas are treated by surgery along with radiotherapy and/or chemotherapy. In WHO classification these tumours are included in a separate 'intermediate category—rarely metastasising' group and cases with features of malignancy have been reported for these tumours. We report this case because of the uncommon site, atypical histopathological features suggestive of its malignant potential and importance of immunohistochemical stains, which are important for diagnosis and precise definition of these tumours.

CASE PRESENTATION

A 40-year man presented with a progressively enlarging painless mass in the antero-medial aspect of the right leg for 1 year with no previous history of trauma. On local examination, the mass measured 10×8×4 cm, with no tenderness, freely mobile with no fixity to underlying structures and the overlying skin normal. A MRI revealed a well-defined soft tissue lobulated mass lesion with central areas of necrosis suggestive of a soft tissue tumour (figure 1A,B).

The mass was resected along with a 1 cm margin and subjected for histopathological examination.

Grossly, the specimen received was well-circumscribed, nodular, pale yellow in colour and measured 8.5×5.5×4.5 cm. On cut sections, the appearance was whitish with focal areas of central haemorrhage (figure 1C).

Microscopically, the tumour was well-circumscribed with both hypocellular and hypercellular areas, which at places were separated by keloid-type collagen deposition.

Random arrangement of cells was seen in most areas. Areas with thick walled hyalinised blood vessels and haemangiopericytoma like pattern were observed. Cells were spindly in most areas along with focal myxoid change, necrosis and haemorrhage. Mitotic count was 5 per high power field (HPF) (magnification×400). Immunohistochemically, the tumour was strongly positive for CD34 along with focal positivity for CD99 and BCL2. The microscopic features are presented in figure 2A–F.

The above findings were helpful for a final diagnosis of solitary fibrous tumour of the soft tissue of the right leg.

INVESTIGATIONS

Haemoglobin, total and differential leucocyte count and platelets were normal for age.

MRI showed a well-defined soft tissue mass in the subcutaneous plane of the antero-medial aspect of the lower part of the right leg, overlying tibia and adjacent tibialis anterior and gastrocnemius muscle. There was no evidence of origin, involvement or erosion of underlying bone. The mass measured 8.9×7.8×5.4 cm in circumferential, transverse and anteroposterior diameters, respectively, with prominent post-contrast enhancement and central areas of non-enhancement suggestive of necrosis. Few curvilinear fibrous septae were seen traversing the mass. Tibia, fibula, muscles around the mass and neurovascular bundles appeared normal and there was no evidence of any calcified foci within the mass. A presumptive diagnosis of soft tissue tumour was given.

DIFFERENTIAL DIAGNOSIS

The pattern of arrangement of cells in solitary fibrous is haphazard or pattern less or may be a combination of various patterns like storiform, neural or herring bone. The proliferation of branching blood vessels may be seen in other high-grade sarcomas. The important differential diagnosis includes haemangiopericytoma, fibrous histiocytoma,

Figure 1 MRI and gross findings of the solitary fibrous tumour involving the right leg. (A, B) MRI showing a well-defined soft tissue mass, with no involvement of underlying structures and bone, on the antero-medial aspect of right leg. (C) Cut section of the tumour showing a whitish soft tissue mass with focal central areas of haemorrhage and necrosis.

dermatofibrosarcoma protuberans, leiomyosarcoma and malignant peripheral nerve sheath tumour.

The exact diagnosis of these tumours is based on their immunoprofile where they are characteristically immunoreactive for CD34 and CD99 along with variable positivity for epithelial membrane antigen, BCL2 and smooth muscle actin. Focal and limited reactivity for S100 protein, cytokeratins and/or desmin has also occasionally been reported.

TREATMENT

Total surgical excision of the tumour.

OUTCOME AND FOLLOW-UP

After 4 months of follow-up the patient was disease free and without any recurrence.

DISCUSSION

Solitary fibrous originally described in pleura have been reported from various extra pleural locations including extremities. The sites in extremities from which the tumour has been reported include thigh, popliteal fossa, flank, neck, shoulder, deep groin and gluteal region.^{1 2} The number of

reported cases in the extremities are few and according to Fukui *et al* there were only 28 cases reported, including their two cases, involving shoulder and thigh.³ The histopathological features and immunological profile include the widely accepted characteristic features of solitary fibrous tumour.⁴ CD 34 has been reported to be the most sensitive immune marker for solitary fibrous tumour.^{4 5} Malignant change and atypical features suggestive of malignant potential have been previously described. Atypical histological features for these tumours include nuclear atypia, areas of increased cellularity, necrosis and four or more mitoses per 10 HPF.⁶ Later on, tumour size greater than 10 cm and positive surgical margins were also included as unfavourable prognostic factors.⁷ The tumour in our case exhibited areas of necrosis and high mitotic rate of 5 per HPF; two of the atypical histological features, sufficient to classify it as atypical fibrous tumour with malignant potential. WHO has placed solitary fibrous tumour in the 'intermediate (rarely metastasising)' category and above mentioned atypical features are contributory to the malignant potential of these tumours.⁴

The treatment protocol for solitary fibrous tumour is total surgical excision of the mass and was followed in our case. Previous studies have documented malignant change

Figure 2 Photomicrographs of solitary fibrous tumour with atypical features. (A) Hyalinised blood vessels with haemangiopericytoma-like arrangement of spindle cells (H&E ×40). (B) Keloid-like thick bands of collagen traversing the tumour (H&E ×400). (C) Hypocellular areas with random arrangement of cells and hyalinised blood vessels (H&E ×100). (D) Hypercellular areas with marked cytological atypia and mitotic figures (H&E ×400). (E) Focal areas of haemorrhage and necrosis (H&E ×40). (F) Immunopositivity for CD 34 (magnification ×400).

in solitary fibrous tumours and the atypical histopathological features are associated with such malignant transformations;^{1 6 7} hence, continuous follow-up has been recommended for solitary fibrous tumours. The patient in our case is on continuous follow-up and after 4 months there is no evidence of recurrence or metastasis.

Learning points

- ▶ Atypical solitary fibrous tumours are ubiquitous neoplasms, can occur at any site and have undetermined malignant potential.
- ▶ The diagnosis of atypical solitary fibrous tumour rests on histopathological examination and immunohistochemical markers, notably CD34 and CD99, and it is very important to differentiate them from other high-grade mesenchymal neoplasms especially sarcomas.
- ▶ The atypical histopathological features in these predominantly benign tumours include nuclear atypia, areas of increased cellularity, necrosis and four or more mitoses per 10 HPF, tumour size greater than 10 cm and positive surgical margins.
- ▶ The treatment protocol for these tumours is total resection of the mass and long-term follow-up of the patient especially when atypical histopathological features are present.

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Competing interests None.

Patient consent Obtained.

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