Amyloidoma of soft tissue is rare and there have been no previously published reports of limb involvement. We describe a case in which the tumour was present in the popliteal fossa. There was no evidence of systemic amyloid disease or of malignant neoplasm one year after the diagnosis.

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Case Report

A 36-year-old mentally subnormal woman was referred to our clinic with a painless diffuse swelling in the right popliteal fossa 12.5 × 15 cm in size, and centred over the fibular head (Fig. 1). It was firm, non-tender and appeared to be in the subcutaneous plane posteriorly. Knee function was normal and there was no regional lymphadenopathy. Plain radiographs were normal and CT confirmed the presence of a soft-tissue mass around the head of the fibula. The ESR, full blood count and bone biochemistry were normal.

MRI (Fig. 2) showed a large, irregular infiltrating mass in the popliteal fossa involving the lateral head of gastrocnemius and soleus. The lesion covered the biceps tendon and common peroneal nerve, and extended posteriorly into the subcutaneous fat.

The clinical signs and MRI strongly suggested a soft-tissue sarcoma and the patient was referred to a Regional Orthopaedic Oncology Unit. CT of the chest was clear and MRI of the pelvis revealed a few shotty nodes in the right inguinal area. Histological examination of an open biopsy of the popliteal mass performed under a tourniquet showed a whitish tumour containing fat-laden histiocytes and foreign-body-type giant cells, with hypocellular and hyalinised fibrous tissue with a patchy dense perivascular infiltrate of plasma cells and histiocytes. There were scattered multinucleate giant cells containing intracytoplasmic pale eosinophilic hyaline material which stained positive with Congo Red and was apple green birefringent under polarised light (Fig. 3).

Immunohistochemistry for AA amyloid was negative, although non-specific background staining was high. Pretreatment with potassium permanganate before staining with Congo Red indicated that this was not AA amyloid, but rather the AL type normally seen in primary amyloidosis.

We sought a second opinion from Dr A. E. Rosenberg at the Massachusetts General Hospital who confirmed the
diagnosis of a soft-tissue amyloidoma. There was no evidence of systemic amyloid disease or of malignant neoplasm one year after the diagnosis. The patient refused a rectal biopsy. Since the swelling had not increased further in size and caused only an occasional ache, we decided not to proceed with a radical excision. MRI one year after the diagnosis (Fig. 4) showed no change, with no invasion of the vessels or adjacent bone.

Discussion

Solitary amyloidoma involving the soft tissues of the lower limbs has not been reported previously. A painless, solid popliteal swelling is more suggestive of a synovioma than an amyloidoma. The mean age at diagnosis for amyloid tumours of soft tissues is 66 years, and by definition they start as solitary lesions. Histological examination shows extensive amyloid deposits with an intense giant-cell reaction and a variable amount of lymphoplasmacytic infiltrate. The pathological diagnosis is difficult. In most of the cases reported the initial diagnosis was of a foreign-body granulomatous reaction. Immunohistochemistry is often useful to distinguish the neoplastic plasma cells of myeloma from the plasma cells seen in amyloidoma.

The distinction between AL and AA fibrils in amyloidoma may be of importance. Krishnan et al found that ten of their 14 patients had AL fibrils which constituted a primary amyloidoma and of these 80% subsequently...
developed a lymphoplasmacellular malignant neoplasm. The remaining four had AA fibrils secondary to a chronic infection or inflammation and are alive without systemic disease.\(^5\)

In the AL fibril (primary) amyloidoma progression to disseminated disease is common and additional tumours develop after an interval of three months to nine years.\(^1\) The prognosis of these patients is generally poor, with most dying of local or general complications of the disease in eight months to three years.\(^1\) There is now compelling evidence that most AL fibril (primary) amyloidomas represent plasmacytomas with massive amyloid deposition.\(^1\)

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References