Case report

Magnetic resonance imaging of subcutaneous diffuse neurofibroma

W C G PEH, FRCR, TW H SHEK, FRCPA and D K H YIP, FRCS

Departments of 1Diagnositic Radiology, 2Pathology and 3Orthopaedic Surgery, The University of Hong Kong, Queen Mary Hospital, Hong Kong

Abstract. A 31-year-old woman presented with increasing pain and tenderness of a long-standing soft tissue mass on her back. MRI showed a network of interconnecting tubular areas, which were \( T_1 \) isointense and \( T_2 \) hyperintense relative to skeletal muscle, and displayed marked Gd-DTPA enhancement. The lesion was situated within the subcutaneous fat. Clinically and radiologically, this mass was considered to be a subcutaneous venous haemangioma. Histological examination of the excised mass showed a diffuse neurofibroma with ectatic vessels and entrapped adipose tissue. Similar MRI appearances of subcutaneous haemangioma and diffuse neurofibroma may result in failure to make the correct diagnosis and in inappropriate management.

Magnetic resonance imaging is now the imaging modality of choice for evaluating suspected soft tissue tumours of the musculoskeletal system, and should be obtained following plain radiography. A specific diagnosis can be made in a quarter to one-third of soft tissue tumours, particularly in benign lesions [1–3]. MRI is accurate in providing a specific diagnosis in haemangioma [1–7]. We describe a patient with histologically proven diffuse neurofibroma who had features suggestive of a subcutaneous venous haemangioma.

Case report

A 31-year-old Chinese woman presented with a painful soft tissue mass over the thoraco-lumbar junction of the spine. She had noticed this mass since early childhood. There was a 1 year history of progressive pain and tenderness over the mass, which had also increased in size. She had no significant past medical history. There was no family history of neurocutaneous disease. On examination, a soft fluctuant mass was palpable over the midline of the spine at T11–L1 level. The mass was tender, well-defined and did not transilluminate. The overlying skin had multiple, light blue, fine punctum-like markings. There was no other clinical abnormality. Thoracolumbar radiographs showed no bony abnormality or soft tissue calcification.

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Address correspondence to Dr Wilfred C G Peh, Department of Diagnostic Radiology, The University of Hong Kong, Room 415, Block K, Queen Mary Hospital, Hong Kong.
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Figure 1. Sagittal MRI (1.5 T) scan of the thoracolumbar spine. (a) SE T₁ weighted (540/10) image shows an isointense network of interconnecting tubular structures within the subcutaneous fat. (b) Fat-suppressed FSE T₂ weighted (3480/104) image shows hyperintensity of these tubular areas. (c) Post-Gd-DTPA fat-suppressed SE T₁ weighted (760/10) image shows marked enhancement of the tubular structures. The vertebrae and contents of the spinal canal are not involved. Surface marker (arrowed) indicates the T12/L1 level.

Discussion

Neurofibroma most commonly arises in association with cutaneous nerves, symptoms being related to the size of the lesion. Most of the descriptions of the MRI appearances of peripheral neurofibromas have been in regard to the focal discrete form, which may have a typical “target” pattern [8–10]. Kransdorf and Berquist have stated that plexiform neurofibroma may occasionally resemble intramuscular haemangioma, with entrapped fat simulating the lace-like pattern of fat interdigitating between the vascular elements of a haemangioma [11]. Two previous studies correlating CT with histological evaluation of peripheral neural tumours have concluded that these tumours may mimic fat-containing lesions due to the presence of adipocytes intermingling with neurofibroma cells and the entrapment of perineural adipose tissue by plexiform neurofibroma [12, 13]. The MRI findings in our patient with diffuse neurofibroma would support all these observations. To our knowledge, MRI features of diffuse neurofibroma has been previously reported in only one other case [14].

Diffuse neurofibroma is an uncommon but distinctive form of neurofibroma which occurs...
phleboliths may be a helpful clue to the diagnosis. Haemangiomas often contain a large amount of non-vascular tissue, especially adipose tissue, which contributes to their characteristic MRI appearances. On $T_1$ weighted images, the lesion is largely isointense to skeletal muscle, with the hyperintense fatty elements being depicted as fine lace-like to coarse band-like septations. On $T_2$ weighted images, the haemangioma is typically well marginated and hyperintense compared with the subcutaneous fat. The $T_2$ hyperintense areas represent slowly flowing blood within the vessels of the haemangioma, with the $T_1$ hyperintense areas reflecting fatty tissue interspersed in between the vessels. There is usually marked enhancement of the vascular elements following administration of Gd-DTPA [4–7]. These features were present in our patient, with the $T_2$ hyperintense and enhancing serpiginous areas suggesting a venous haemangioma.

Although the majority of neurofibromas are solitary, the possible association with neurofibromatosis type I must always be considered. Onset of pain or enlargement of a neurofibroma may herald malignant transformation. It is useful to be aware that diffuse neurofibroma may have a very similar MRI appearance to subcutaneous haemangioma.

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References

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