

Case report

Magnetic resonance imaging of subcutaneous diffuse neurofibroma

¹W C G PEH, FRCR, ²T W H SHEK, FRCPA and ³D K H YIP, FRCS

Departments of ¹Diagnostic 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.

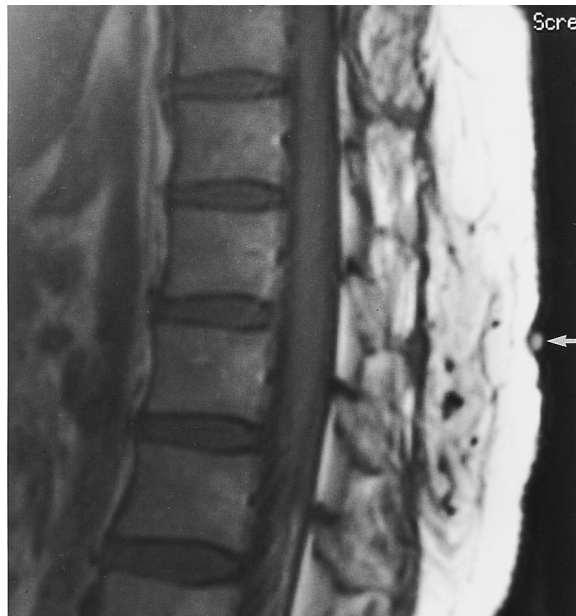
Received 15 April 1997 and in revised form 16 June 1997, accepted 26 June 1997.

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.

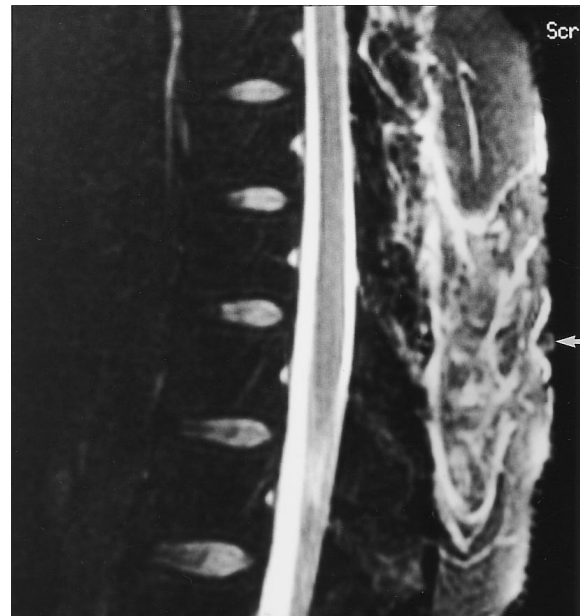
Spin echo (SE) T_1 , fat-suppressed fast SE (FSE) T_2 and post-contrast fat-suppressed SE T_1 weighted MRI scans of the thoracic and lumbar spine were obtained in sagittal and axial planes. MRI showed multiple interconnecting tubular areas within the subcutaneous fat overlying the T11–L2 vertebrae. These areas were T_1 isointense and T_2 hyperintense relative to skeletal muscle, and displayed marked enhancement following iv gadopentetate dimeglumine (Gd-DTPA). The underlying paraspinal muscles, bone and spinal cord were normal (Figure 1).

The diagnosis at this time was subcutaneous venous haemangioma. Fine needle aspiration for cytology was inconclusive. Digital subtraction angiography, with selective cannulation of the T9–11 intercostal, subcostal and L1–3 lumbar branches bilaterally, and red blood cell scintiscan, were negative. At surgery, a large fleshy mass was found within the subcutaneous fat, superficial to the fascia of the paraspinal muscles. Part of this fascia was removed together with the mass. The mass was discrete at its superior, inferior and lateral margins, and could be enucleated. The excised specimen measured 17 cm × 9 cm × 1 cm.

Histological examination showed a diffuse neurofibroma which was characterized by a diffuse proliferation of spindle cells within mature adipose tissue. These spindle cells possessed small comma-shaped nuclei and were situated within a fibrillary matrix in which occasional Wagner–Meissner bodies were found. The tumour cells were immunoreactive for S100 protein, confirming their peripheral nerve sheath nature. The tumour also contained numerous ectatic blood vessels (Figure 2). The patient made an uneventful



(a)



(b)



(c)

Figure 1. Sagittal MRI (1.5 T) scan of the thoracolumbar spine. (a) SE T_1 weighted (540/10) image shows an isointense network of interconnecting tubular structures within the subcutaneous fat. (b) Fat-suppressed FSE T_2 weighted (3480/104) image shows hyperintensity of these tubular areas. (c) Post-Gd-DTPA fat-suppressed SE T_1 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.

recovery and remains well at 10 months post-operation.

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

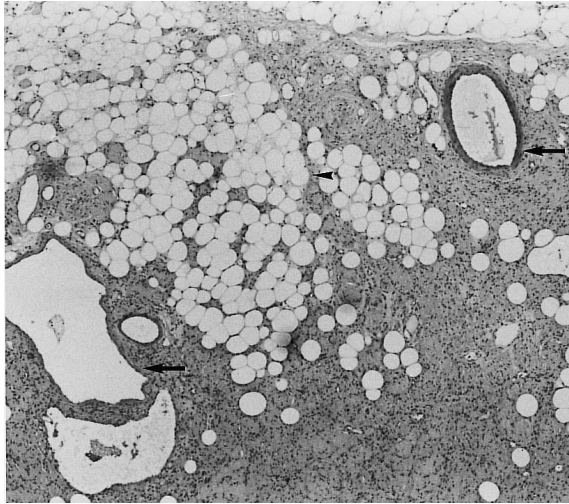


Figure 2. There were ectatic blood vessels (arrows) and adipose tissue (arrowhead) interspersed between the tumour tissue. (Haematoxylin and eosin stain, original magnification, $\times 50$.)

principally in children and young adults. Unlike the commoner focal neurofibroma, diffuse neurofibroma is often ill-defined and spreads extensively along connective tissue septa and in-between adipose tissue. Diffuse neurofibroma typically involves the subcutaneous tissue down to the level of fascia. It is not clear exactly how often diffuse neurofibroma is associated with neurofibromatosis, although it has been suggested that about 10% of patients with this lesion also have neurofibromatosis type I [15]. Neurofibromatous tissue would be expected to produce a T_1 isointense or slightly hyperintense and T_2 isointense to hyperintense signal relative to muscle, and enhance following Gd-DTPA administration. The presence of ectatic vessels within the tumour in our case contributes not only to the interconnecting tubular appearance, but also to T_2 hyperintensity of the tumour. These vessels remain T_1 isointense and enhance markedly.

The histological demonstration of ectatic blood vessels and mature adipose tissue correlated well with the MRI appearances of the lesion in our patient. It is of interest to note that both these elements are characteristically found in diffuse neurofibroma. Sometimes, the large ectatic vessels may be striking enough so as to eclipse the neural element and result in the erroneous impression of exuberant granulation, even on histological examination [15]. The MRI findings in the present case were similar to the case of diffuse neurofibroma of the neck described by de Varebeke et al in 1996 [14], although no mention was made of ectatic vessels on histological examination.

The vast majority of soft tissue haemangiomas occurs in young adults, with 80–90% presenting by 30 years of age. Plain radiographic findings are usually non-specific although the presence of

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.

Acknowledgment

We thank Ms Alice Lau for her secretarial assistance.

References

1. Berquist TH, Ehman RL, King BF, Hodgman CG, Illstrup DM. Value of MR imaging in differentiating benign from malignant soft tissue masses: study of 95 lesions. *AJR* 1990;155:1251–5.
2. Crim JR, Seeger LL, Yao L, Chandnani V, Eckardt JJ. Diagnosis of soft tissue masses with MR imaging: can benign masses be differentiated from malignant ones? *Radiology* 1992;185:581–6.
3. Moulton JS, Blebea JS, Dunco DM, Braley SE, Bisset GS III, Emery KH. MR imaging of soft-tissue masses: diagnostic efficacy and value of distinguishing between benign and malignant lesions. *AJR* 1995;164:1191–9.
4. Nelson MC, Stull MA, Teitelbaum GP, Patt RH, Lack EE, Bogumill GP, et al. Magnetic resonance imaging of peripheral soft tissue hemangiomas. *Skeletal Radiol* 1990;19:477–82.
5. Rak KM, Yakes WF, Ray RL, Dreisbach JN, Parker SH, Luethke JM, et al. MR imaging of symptomatic peripheral vascular malformations. *AJR* 1992;159:107–12.
6. Murphey MD, Fairbairn KJ, Parman LM, Baxter KG, Parsa MB, Smith WS. Musculoskeletal angiomatous lesions: radiologic-pathologic correlation. *RadioGraphics* 1995;15:893–917.

7. Memis A, Arkun R, Ustun EE, Kandiloglu G. Magnetic resonance imaging of intramuscular haemangiomas with emphasis on contrast enhancement patterns. *Clin Radiol* 1996;51:198–204.
8. Suh JS, Abenzoza P, Galloway HR, Everson LI, Griffiths HJ. Peripheral (extracranial) nerve tumors: correlation of MR imaging and histologic findings. *Radiology* 1992;183:341–6.
9. Sakai F, Sone S, Kiyono K, Maruyama A, Ueda H, Aoki J, et al. Intrathoracic neurogenic tumors: MR–pathologic correlation. *AJR* 1992;159:279–83.
10. Varma DGK, Moullopulos A, Sara AS, Leeds N, Kumar R, Kim EE, et al. MR imaging of extracranial nerve sheath tumors. *JCAT* 1992;16:448–53.
11. Kransdorf MJ, Berquist TH. Neurofibroma. In: Berquist TH, editor. *MRI of the musculoskeletal system* (3rd edn). Philadelphia: Lippincott-Raven, 1996:789–92.
12. Kumar AJ, Kuhajda FP, Martinez CR, Fishman EK, Jezic DV, Siegelman SS. Computed tomography of extracranial nerve sheath tumors with pathological correlation. *JCAT* 1983;7:857–65.
13. Cohen LM, Schwartz AM, Rockoff SD. Benign schwannomas: pathologic basis for CT inhomogeneities. *AJR* 1986;147:141–3.
14. de Varebeke SJ, de Schepper A, Hauben E, Declau F, Marck EV, de Heyning PHV. Subcutaneous diffuse neurofibroma of the neck: a case report. *J Laryngol Otol* 1996;110:182–4.
15. Enzinger FM, Weiss SW. *Soft tissue tumors* (3rd edn). New York: Mosby, 1995:889–928.