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# MRI in an unusual case of congenital spinal mesenchymal proliferation

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**Abstract** We report a child aged 2 years presenting with delayed motor development. A thoracolumbar subcutaneous mass was noticed in the first months of life. MRI showed a low conus medullaris, confirmed the presence of the mass and detected a second solid lesion in the intradural space. Surgery confirmed that the two lesions were distinct, as on MRI. The histopathological features were in common with fibrous hamartoma of infancy, giant cell angioblastoma and the "diffuse type" of infantile fibromatosis. The presence of a low conus medullaris associated with a congenital clinical presentation suggested a disontogenetic aetiology.

**Key words** Angioblastoma · Fibromatosis · Hamartoma · Magnetic resonance imaging · Spinal tumours · Children

## Introduction

Fibroblastic proliferations in children include a group of rare disorders peculiar to this age which have no clinical or morphological counterpart in adult life. They often have unusual microscopic features that pose special diagnostic problems; high cellularity and rapid growth can mimic sarcomas but they usually are low-grade lesions and surgical excision is curative [1, 2].

Fibrous proliferations peculiar to childhood include fibrous hamartoma, infantile desmoid fibromatosis, fibromatosis colli and digital fibromatosis, which have distinctive morphological and clinical features [2].

We describe the MRI and pathological features of a case with a composite fibroblastic proliferation not previously described.

## Case report

A 2-year-old boy was admitted because of delayed motor development: at 18 months of life he was unable to stand or walk. At 6 months the parents had noticed a lumbar mass, which apparently had grown little.

The family history was unremarkable except for cutaneous angiomas in the father. A painless subcutaneous mass measuring $3 \times 4.5$ cm was visible and palpable in the left thoracolumbar paravertebral region. It was fixed to the deep muscular tissues but the overlying skin was normal. The leg muscles were poorly developed and a severe paraparesis was present without any sensory impairment. Multiple cutaneous angiomas were observed on the fingers, arms and legs.

Spinal and trunk MRI (Fig. 1) was performed at 0.5 T in three planes. T1-weighted spin-echo images before and after intravenous contrast medium and unenhanced T2-weighted images were obtained. They showed two distinct lesions: an intradural mass extending from T5 to L3 and a posterolateral extraspinal paraverte-

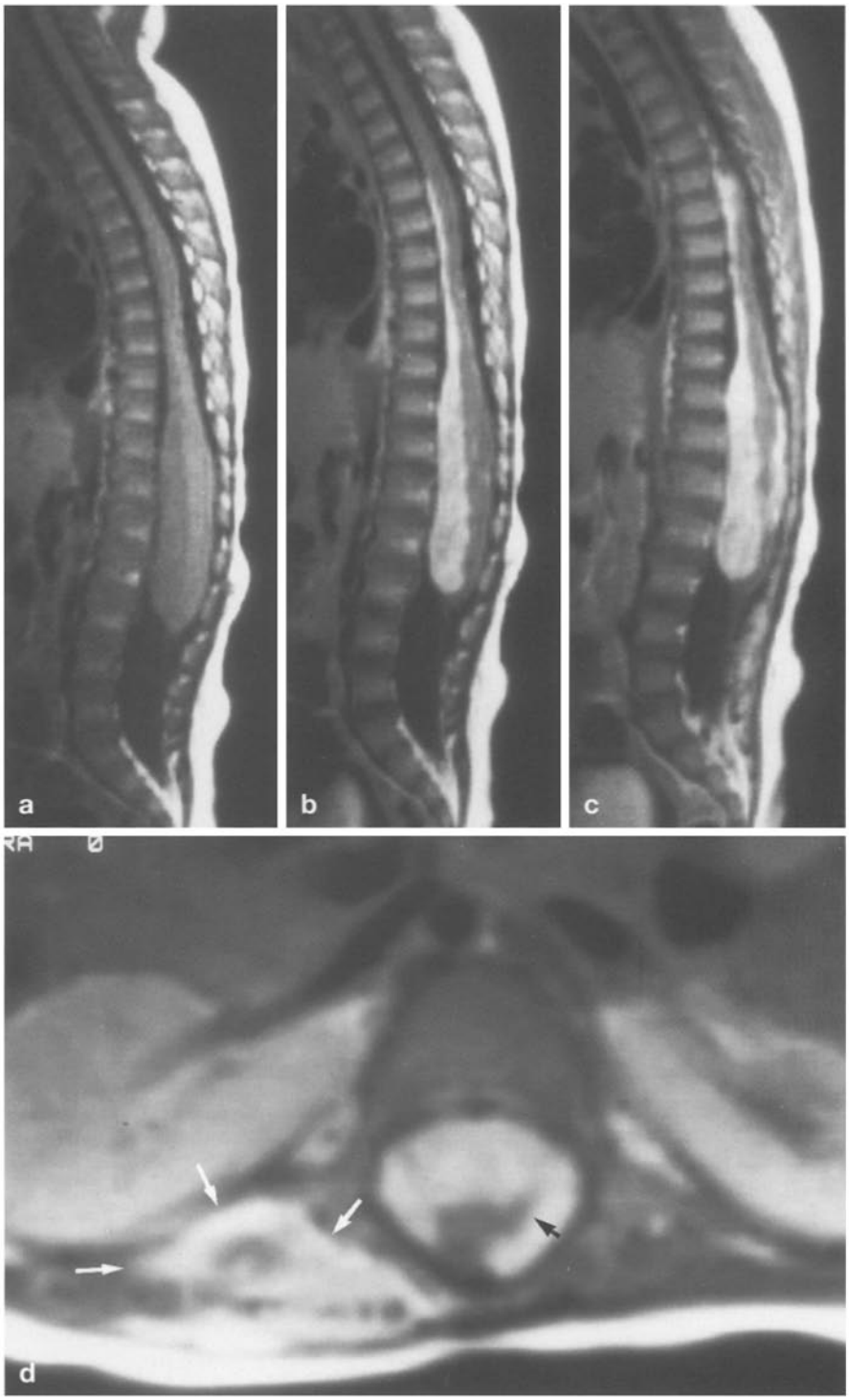


**Fig. 1** **a** Sagittal T1-weighted spin-echo (SE), **b, c** contrast-enhanced sagittal T1-weighted, **d** contrast-enhanced axial T1-weighted SE images. The spinal cord is "club-shaped", swollen from T10 to L3 in the midsagittal plane **(a)**. On contrast-enhanced images it appears to be coated by a thick sleeve of markedly enhancing tissue from T5 to the level of a low conus medullaris, where a short, thick filum terminale arises. The spinal cord is irregular and displaced posteriorly, while the spinal canal is widened and deformed with erosion of the posterior arches. **c** A subcutaneous mass in the left posterior paravertebral region *(white arrows)* shows contrast enhancement similar to that of the intraspinal lesion. A thick nerve root *(black arrow)* originates from the right anterolateral margin of the spinal cord, completely enclosed by the sleeve of tumour

bral mass, with no demonstrable connection with the first. No intraspinal extradural component was observed.

On sagittal T1-weighted images the spinal cord appeared "club-shaped", swollen from T10 to L3 (Fig. 1a). Contrast-enhanced images (Fig. 1b) showed an extremely deformed and irregular spinal cord, displaced posteriorly and surrounded by a sleeve of markedly but inhomogeneously enhancing tissue. This was detectable from T5 to the level of a low-lying conus medullaris, where a short, thick filum terminalis arose. A slight tail of intramedullary contrast enhancement was detectable (Fig. 1b).

Some thickened roots originated from the anterolateral border of the cord at the L1 level (Fig. 1c).

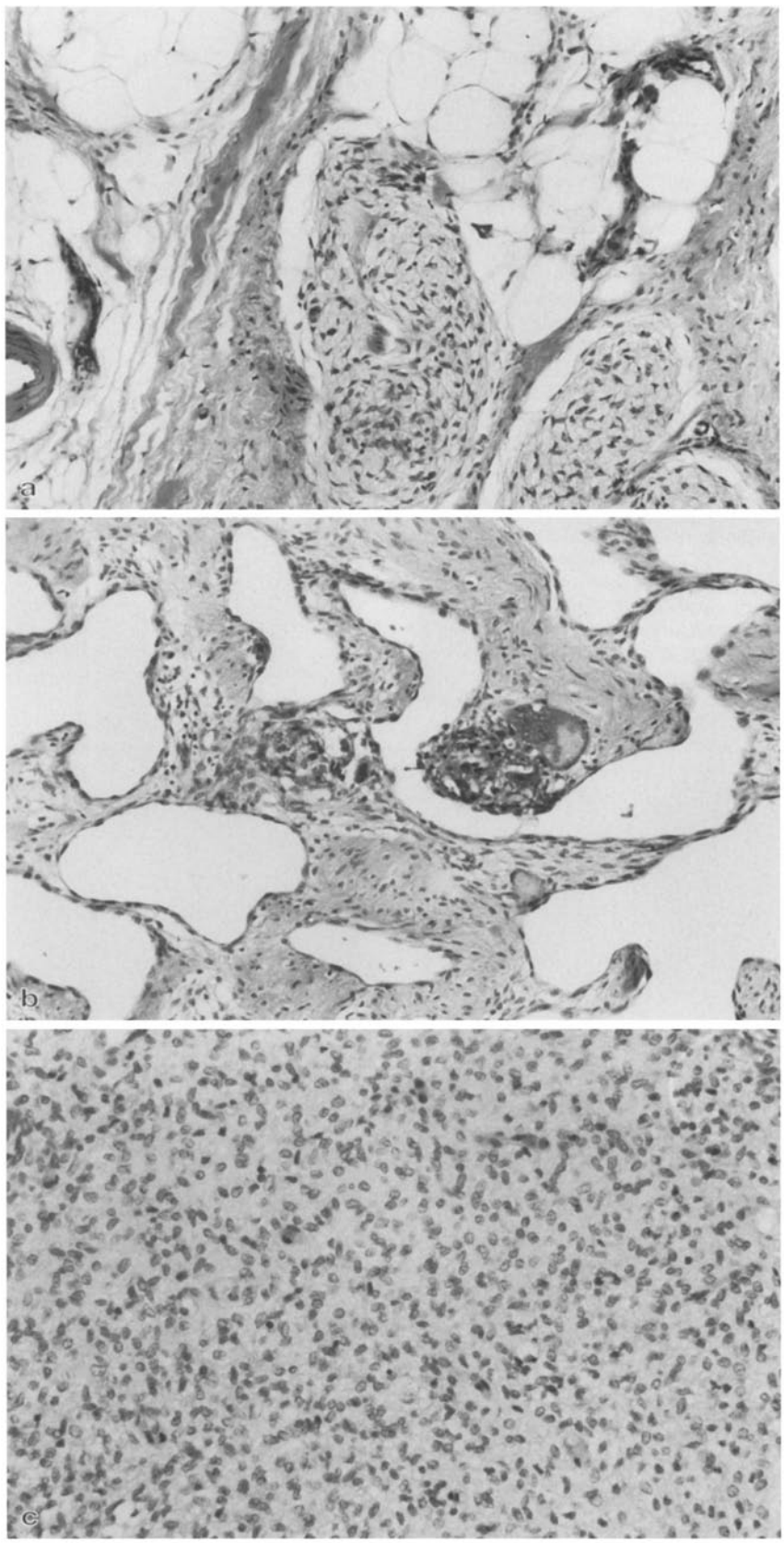


**Fig. 2** **a** Bands of connective tissue, fat and myxoid foci (original magnification × 114). **b** Dilated, communicating vascular channels, cellular foci and a few giant cells (original magnification × 114) **c** Highly cellular area composed of immature fibroblasts (original magnification × 163)

The thoracolumbar spinal canal was widened, particularly in its sagittal diameter.

A paravertebral mass was detected on the left at the T12–L1 level; the 5 × 2.5 cm lesion was heterogeneous and infiltrating, and replaced the dorsal musculature with no evident connection with the intraspinal component. However, the pattern of contrast enhancement was similar.

At surgery the subcutaneous mass, whitish-yellow and with a lipomatous appearance, was firmly connected to the underlining muscle, which showed abnormal bleeding. The dura mater was intact but thin; the intraspinal mass was entirely intradural, and extensively attached to the spinal cord, nerve roots and blood vessels. The cord was displaced posteriorly and laterally. The paravertebral mass measured 4.5 × 3.5 × 1.7 cm and was composed of yellowish-grey tissue with myxoid and haemorrhagic areas. The small fragments from the intraspinal component of the lesion were composed of greyish-white tissue.

On histology, three different patterns were observed from the subcutaneous tissue to the deeper paravertebral and intraspinal region. The more superficial part displayed an organoid pattern due to trabeculae of fibrous tissue mixed with abundant fat and multiple foci of small, immature spindle cells embedded in a myxoid matrix (Fig. 2a). The deeper component was mainly composed of a prominent vascular network; the vessels were irregular and communicated with each other, creating an anastomosing pattern. Endothelial cells had hyperchromatic nuclei. Multiple cellular foci composed of histiocytic cells and multinucleate giant cells were observed in the lumen or within the wall of the vessels (Fig. 2b). Spindle-shaped cells embedded in mature collagen were observed between the vessels. The intraspinal lesion was characterised by a proliferation of immature fibroblasts with rounded nuclei and scant cytoplasm. Mitotic figures were rare and necrosis absent. Foci of similar cells were also observed within the paravertebral muscles, with infiltration of the thoracolumbar fascia and spinal nerves (Fig. 2c).

## Discussion

The peculiar histological features of these lesions render diagnosis and treatment difficult [1, 2]. The interpretation of imaging is also difficult because of the paucity of MRI and CT reports [3–5].

Age, location, number of lesions and tendency to recur after excision or spontaneous regression differ in the numerous distinct entities which represent the fibrous proliferations peculiar to infancy and childhood [2].

From the histological point of view, our case seems to combine features of three different entities: fibrous hamartoma of infancy [6], giant cell angioblastoma [7] and "diffuse type" infantile fibromatosis [2].

The central nervous system is an uncommon site for fibromatosis in infants and all reports are of infantile myofibromatosis, the only type of fibrous proliferation in children which affects multiple sites and involves soft tissues and internal organs [1, 2, 8, 9].

The unusual histological features and clinical characteristics in this case did not fit any of the clinicopathological entities previously described.

To our knowledge, this is the first report of such a composite fibroblastic proliferation in an infant.

The association with malformations has not been reported previously. A low conus medullaris and the first recognition of the lesion at 6 months of age suggest a disontogenetic aetiology.

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