Diffuse Neurofibroma of the Thigh Mimicking a Cutaneous Hemangioma: a case report

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Diffuse neurofibroma is a rare variant of neurofibroma occurring mainly in children and young adults. The most common sites of involvement are head, neck and trunk. Diffuse neurofibroma is an ill-defined infiltrative lesion and typically involves the subcutaneous tissue down to the level of fascia [1, 2]. We report a case of diffuse neurofibroma at the thigh. The patient presented with a slowly growing mass at right thigh since childhood. The clinical features and MR findings mimic a cutaneous hemangioma. The characteristic MR imaging patterns are discussed and literatures are reviewed.

CASE REPORT

A 14-year-old girl presented with a slowly growing soft tissue mass at right thigh since early childhood. No significant past medical history or family history of neurocutaneous disease was noted. On physical examination, a soft brown-erythematous mass with curling skin about 7 × 4 cm was seen at the medial aspect of right thigh. (Fig. 1) There was no other clinical abnormality such as “café au lait” macules, freckling in the axillary or inguinal regions, or classic neurofibromas. The laboratory data, including hematology and blood chemistry tests, were normal.

Magnetic resonance (MR) examination revealed a bulging tumor at the medial aspect of right thigh. The mass involved the skin and subcutaneous layer. MR images of the lesion demonstrated linear strands of intermediate signal intensity in the subcutaneous fat with thickened soft tissue in the overlying skin on T1-weighted images. On T2-weighted images, the tumor showed high signal intensity of the thickened soft tissue and the linear strands in the subcutaneous fat. Strong enhancement of the tumor was noted after IV gadopentetate dimeglumine (Gd-DTPA) administration. (Fig. 2a-2c) According to the clinical features and MR findings, our preoperative diagnosis of the lesion was subcutaneous venous hemangioma or angiolipoma.

The patient underwent surgical resection of
the tumor. Histopathological examination of the specimen showed the features of skin and fibroadipose tissue with collagens and spindle neoplastic cells which were bearing wavy nuclei. Some nerve bundles were found in the tumor. (Fig. 3) The final pathological diagnosis was subcutaneous diffuse neurofibroma.

**DISCUSSION**

Diffuse neurofibroma, also called paraneurofibroma, is an uncommon but distinctive form of neurofibroma which occurs primarily in children and young adults. The most often sites of involvement are the head, neck and trunk regions. Diffuse neurofibromas are poorly circumscribed non-encapsulated tumors and typically involve the subcutaneous tissue down to the level of fascia [1-4]. They cause focal skin thickening and spread extensively along connective tissue septa and in-between adipose tissue. About 10% of patients with diffuse neurofibromas also have associated neurofibromatosis type 1 [1, 2]. The ultrasound features of diffuse neurofi-

**Figure 1.** A soft brown-erythematous mass about 7 × 4 cm with curling skin was seen on the medial aspect of the right thigh.

**Figure 2.** a. Coronal and b. axial precontrast T1-weighted (TR/TE/excitations: 575/13/2) MR images reveal a cutaneous mass on the medial aspect of the right thigh. There are linear strands of intermediate signal intensity in the subcutaneous fat with thickened soft tissue in the overlying skin (arrows). c. Postcontrast axial T1-weighted (484/12/2) image with fat suppression, marked linear enhancement of the mass are showed (arrows).
MRI of diffuse neurofibroma of thigh

The majority of soft tissue hemangiomas occur in young adults, with 80-90% presenting by 30 years of age. The characteristic MR appearances of hemangiomas are iso-signal intensity to skeletal muscle with high signal intensity fatty elements on T1-weighted images, and high signal intensity of well-demarcated tumors on T2-weighted images. There is usually marked enhancement of hemangioma [1, 8]. These MR findings are also seen in our case. In addition, the clinical features of erythematous discoloration of involved skin made it difficult to make a correct diagnosis before surgery.

In summary, the clinical features and MR findings of diffuse neurofibromas are similar to those of cutaneous hemangiomas. Two important MR features including ill-defined margin of the lesion, and linear or reticular enhancement on post-contrast T1-weighted images may provide information to differentiate diffuse neurofibroma from cutaneous hemangioma [1, 2, 8].

REFERENCES

大腿瀰漫性神經纖維瘤之磁振造影影像：病例報告

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瀰漫性神經纖維瘤是一種相當少見的神經纖維瘤。好發於小孩與年輕人，較常侵犯的部位是頭頸部與體幹部位。我們報告一14歲女孩病例，從小發現於右邊大腿內側緩慢生長的軟組織腫瘤。經手術切除證實為瀰漫性神經纖維瘤，其磁振造影影像及臨床外觀與皮下血管瘤非常相似。故提出報告並作其文獻回顧。