

## IMAGES IN CLINICAL MEDICINE

## Elephantiasis Neuromatosa



A 22-YEAR-OLD WOMAN PRESENTED WITH AN 8-YEAR HISTORY OF A SLOW-growing mass involving the buttocks and right leg. She had received a diagnosis of neurofibromatosis type 1 (NF-1) at the age of 18 years, when she had presented with axillary freckling, a family history of NF-1, café au lait macules, and scoliosis. A physical examination at the current presentation revealed hypertrophy of the right leg and an irregular pendulous mass with overlying hyperpigmentation and hypertrichosis involving the right trunk, right buttock, and right leg (Panels A and B). Lisch nodules were seen on ophthalmologic examination. Histopathological examination showed spindle cells arranged in short fascicles that infiltrated the dermis and underlying soft tissues — a finding that was suggestive of a massive soft-tissue neurofibroma. The patient underwent surgical debulking to alleviate the pain and improve the cosmetic appearance. Massive soft-tissue neurofibroma is an uncommon type of neurofibroma and is usually found only in patients with NF-1. Massive soft-tissue neurofibroma involving hypertrophy of the limb — termed elephantiasis neuromatosa — is a rare clinical presentation.

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