Case report 509

Mark I. Burnstein, M.D.1, Sambasiva R. Kottamasu, M.D.2, Lester Weiss, M.D.3, and Michael E. Katz, M.D.2

1 Department of Radiology, University of Wisconsin Hospital and Clinics, Madison, Wisconsin, 2 Diagnostic Radiology and Medical Imaging, and 3 Pediatrics, Henry Ford Hospital, Detroit, Michigan, USA

Clinical information

This 16-year-old white male was referred to our institution with a chief complaint of pain in the right inguinal region. The patient was the product of a normal term pregnancy with uncomplicated labor and delivery. At birth he had been noted to have a lump on his right upper back which was subsequently surgically removed. Histopathology had revealed a lipoma. By 18 months of age he had developed a protuberant abdomen and slender extremities. Exploratory abdominal surgery had demonstrated extensive intra-abdominal lipomatosi. A recent physical examination showed a boy of normal intelligence with short stature and scoliosis. A large mobile mass involved the right chest wall and back. The abdomen was markedly dis- tended with enlarged veins on the abdomen and chest. He weighed 131 pounds and was 56 inches tall. Sexual development was Tanner I. The upper and lower extremities were devoid of subcutaneous fat. The left calf was longer and larger than the right and overgrowth of the left index and middle fingers was present. A verrucous palmar rash was the only significant skin abnormality. All laboratory studies were normal.

Skeletal survey demonstrated a hypoplastic right fifth rib and a soft tissue mass of decreased density corresponding to a large lipoma involving the right side of the upper portions of the chest and back (Fig. 1). Severe rotoscoliosis of the lower thoracic and lumbar spine was associated with multiple enlarged and dysplastic vertebral bodies (Fig. 2). Radiographs of the hands demonstrated marked overgrowth of the left index finger with radial deviation (Fig. 3). The left tibia and fibula...
were elongated with associated overgrowth of soft tissue of the left calf (Fig. 4). The skull was normal. Computed tomography of the chest and abdomen demonstrated large masses of fat density in the right axillary region, posterior chest wall, and right paraspinal region, extending from the mid thoracic to the lower lumbar regions (Fig. 5). Diffuse fatty infiltration of the mesentery and posterior abdominal wall was present (Fig. 6). The lungs, heart, liver, spleen, and kidneys were normal.