

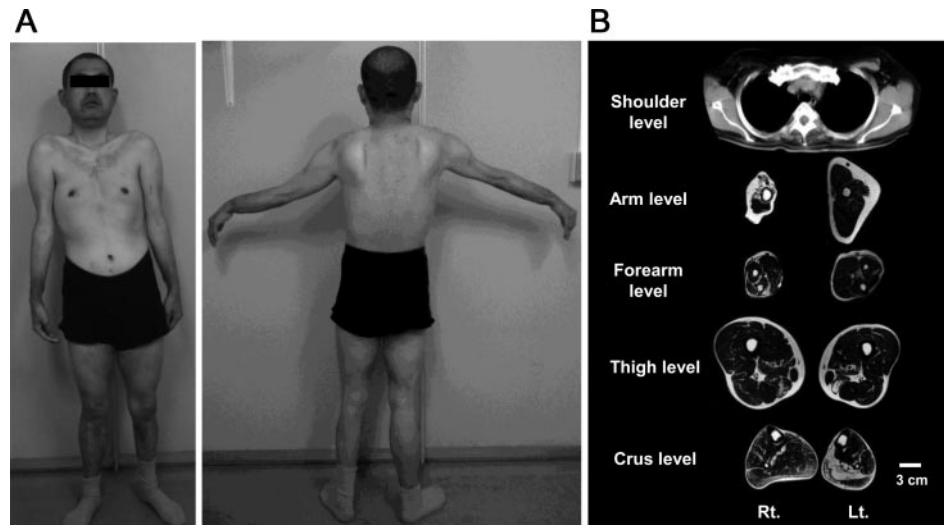
Teaching NeuroImages: Unilateral arm and contralateral leg amyotrophy in FSHD

Unusual presentation

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Figure Photographs and muscle imaging findings of the whole body in a patient with facioscapulohumeral muscular dystrophy (FSHD)



(A) The patient predominantly shows remarkable atrophy of the right facial, shoulder girdle, and arm muscles and of the left leg muscles. He put his weight on his right leg (with permission). (B) CT of the shoulder and T2-weighted MRI of the 4 limbs. CT shows atrophy of the right shoulder girdle and greater pectoral muscles. Amyotrophy and replacement of muscle tissue by fat is asymmetrically pronounced in the right sides of the biceps and triceps brachii and forearm, and the left sides of the femoral and calf muscles on MRI.

A 43-year-old, right-handed man noticed right arm weakness at age 23, followed by the development of left leg weakness and claudication. Although his deceased mother was considered to have had facioscapulohumeral muscular dystrophy (FSHD), her clinical symptoms were unclear. Neurologic examinations and imaging showed predominant weakness/atrophy in the right arm and left leg (figure). EMG demonstrated predominant myopathic changes in the right arm and left leg. No involvement of the CNS or peripheral nerves was apparent. Southern blotting analysis for FSHD revealed a 20-kb *EcoRI* fragment on 4q35 (normal >35 kb).

Asymmetric muscle involvement is a characteristic feature of FSHD.^{1,2} Asymmetry might depend not only on handedness, but also on genetic predisposition.

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