Teaching NeuroImages: Unilateral arm and contralateral leg amyotrophy in FSHD
Unusual presentation

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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. Asymmetry might depend not only on handedness, but also on genetic predisposition.

REFERENCES
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