Teaching NeuroImages:

Hemiatrophy as a clinical presentation in facioscapulohumeral muscular dystrophy

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

(A) The patient shows remarkable atrophy of the right facial, shoulder girdle, pectoral, humeral, and calf muscles with a scapular wing (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. Replacement of muscle tissue by fat is asymmetrically pronounced in the right biceps and the triceps brachii, forearm, femoral, tibialis anterior, and calf muscles on MRI.

A 62-year-old, right-handed man with no family history presented with hemiatrophy (atrophy on one side of the body). He noticed right arm weakness in adolescence, and subsequently right foot drop. Neurologic examinations, including electromyography and imaging, showed right side predominant weakness/atrophy (figure). No asymmetry was apparent in systemic organs, including the brain. Southern blotting analysis for facioscapulohumeral muscular dystrophy (FSHD) revealed a 20-kb *Eco*RI fragment on 4q35 (normal >35 kb).

Asymmetric muscle involvement is a characteristic feature of FSHD¹; however, hemiatrophy with

minimal contralateral involvement is rare in elderly patients. Asymmetry might depend on mechanical factors and genetic predisposition.² This report may expand clinical variability of FSHD.

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