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 EcoRI fragment on 4q35 (normal >35 kb).

Asymmetric muscle involvement is a characteristic feature of FSHD; however, hemiatrophy with minimal contralateral involvement is rare in elderly patients. Asymmetry might depend on mechanical factors and genetic predisposition. This report may expand clinical variability of FSHD.

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