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Format: Abstract

Small fiber neuropathy is a common feature of Ehlers-Danlos syndromes.

Cazzato D[1], Castori M[1], Lombardi R[1], Caravello F[1], Bella ED[1], Petrucci A[1], Grammatico P[1], Dordoni C[1], Colombi M[1], Lauria G[2].

Author information

Abstract

OBJECTIVE: To investigate the involvement of small nerve fibers in Ehlers-Danlos syndrome (EDS).

METHODS: Patients diagnosed with EDS underwent clinical, neurophysiologic, and skin biopsy assessment. We recorded sensory symptoms and signs and evaluated presence and severity of neuropathic pain according to the Douleur Neuropathique 4 (DN4) and ID Pain questionnaires and the Numeric Rating Scale (NRS). Sensory action potential amplitude and conduction velocity of sural nerve was recorded. Skin biopsy was performed at distal leg and intraepidermal nerve fiber density (IENFD) obtained and referred to published sex- and age-adjusted normative reference values.

RESULTS: Our cohort included 20 adults with joint hypermobility syndrome/hypermobility EDS, 3 patients with vascular EDS, and 1 patient with classic EDS. All except one patient had neuropathic pain according to DN4 and ID Pain questionnaires and reported 7 or more symptoms at the Small Fiber Neuropathy Symptoms Inventory Questionnaire. Pain intensity was moderate (NRS ≥4 and <7) in 8 patients and severe (NRS ≥7) in 11 patients. Sural nerve conduction study was normal in all patients. All patients showed a decrease of IENFD consistent with the diagnosis of small fiber neuropathy (SFN), regardless of the EDS type.

CONCLUSIONS: SFN is a common feature in adults with EDS. Skin biopsy could be considered an additional diagnostic tool to investigate pain manifestations in EDS.

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