Chronic Pain, Subjective Stiffness, and Hypermobility-Consider Ehlers-Danlos Syndrome

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Introduction

Ehlers-Danlos Syndrome (EDS) is a collection of heritable connective tissue disorders characterized by joint hypermobility, skin hyper-extensibility, and tissue fragility. The incidence of EDS is estimated to be 1 in 5000. There are 6 subtypes of EDS with varying features and some variants do not include the visually striking skin hyper-extensibility. Due to subtle clinical features and the rarity of this disorder, many patients remain undiagnosed. Chronic pain is common among this population with an incidence as high as 92%.

Case Series

We report a case series of seven patients with diffuse musculoskeletal pain who underwent years of failed treatments prior to a diagnosis of EDS. All patients reported subjective stiffness coincident with their hypermobility. Assuming stiffness was a function of soft tissue, we assessed muscle tenderness using an electrical stimulator to replicate motion in discreet muscles, rather than through palpation. All patients were found to have from 12 to more than a hundred specific tender muscles. The diagnosis of Hypermobile EDS is made clinically using the Beighton scale, and in inherited cases (~50%) also utilizing family history and the Brighton scale. All of these patients underwent muscle/tendon injections and low-level laser therapy (LLLT) to the identified muscles with significant relief of their pain.

Design and Outcomes

After a thorough history was obtained patients underwent a physical examination to identify specific muscles as a potential source of their pain and stiffness. Muscles were identified using electrical stimulation to produce a subclinical contraction of each muscle tested rather than using palpation as a means of identifying a painful muscle trigger point. This concept is thoroughly presented in prior publications. The identified muscles are then treated with a combination of LLLT and muscle/tendon injections. Outcome Assessment: Brief Pain Inventory (BPI) and Visual Analog Scores, self-assessment validated test instruments that determines pain intensity and interference in activities of living, were given to each patient prior to and following completion of LLLT and muscle/tendon injections by the office manager.

Conclusion

Diffuse musculoskeletal pain with subjective stiffness merits consideration of EDS. We suggest subjective stiffness coexisting with hyperflexibility be included as an additional criterion to describe and diagnose Hypermobile EDS. Treatments to reduce muscle stiffness, including LLLT and muscle/tendon injections may result in effective pain reduction. Earlier diagnosis of EDS may obviate unnecessary interventions and investigations whilst facilitating efficacious and timely treatment.

References