CASE REPORT

Ehlers-Danlos Syndrome as a Cause of Temporomandibular Joint Disorders

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Ehlers-Danlos syndrome (EDS), a genetic disorder of collagen formation, creates unique restrictions in the treatment of patients with temporomandibular joint (TMJ) involvement. Although it is not a common cause of TMJ disorders, EDS can produce serious complications in patients receiving conventional TMJ therapy, and thus should be recognized in diagnostic procedures so that treatment planning can be modified accordingly. Three cases have been seen by the author and the most recent and severe is present here in detail.

Case Report

A 21-year-old woman presented to the Facial Pain Center at the Univeristy of Maryland Dental School complaining of pain and intermittent clicking in her right TMJ. The problem started approximately 6 months previously and was associated with eating a sandwich. Past medical history was not significant. The routine question "Do you have problems in any other joints in your body?" evoked the response "I'm double jointed." She interrupted the following question and said "I have Ehlers-Danlos syndrome." She then related that she had been under treatment by a dentist for TMJ problems but did not mention her EDS to him.

A Panorex film revealed no abnormalities. Extraoral head and neck examination revealed a maximum opening of 40 mm and pain on palpation of the right TMJ. No clicks, gait deviation, or muscle pain were found on examination. The remainder of the physical examination was concerned with the EDS. The elbow skin was hyperextendable, there were "cigarette paper" scars, the finger joints were hyperextendable, and the teeth showed abnormalities of enamel calcification (Figs. 1-3).

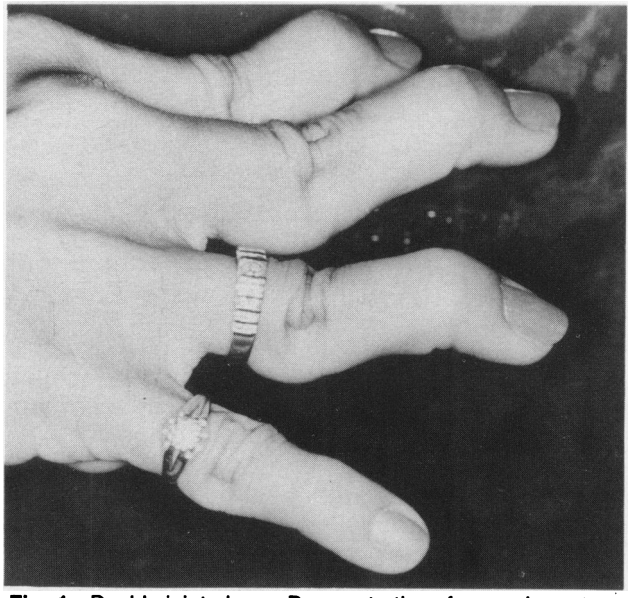

Fig. 1—Double-jointedness. Demonstration of excessive extension of digits voluntarily by patient. Other joints show similiar hypermobility.

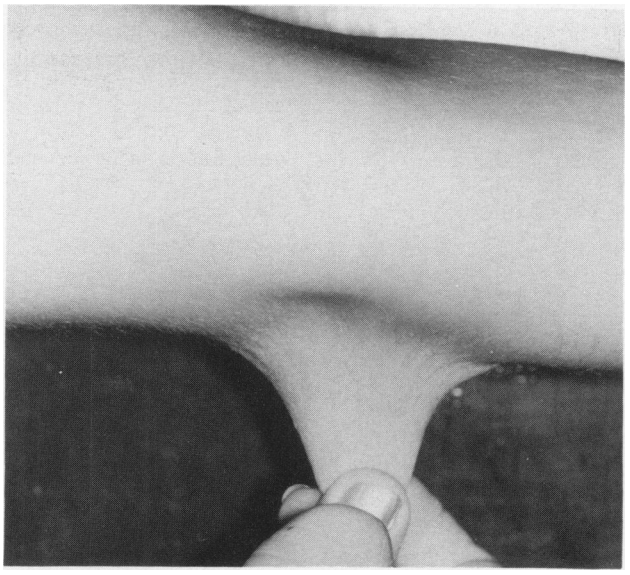

Fig. 2—Hyperelasticity of skin at elbow due to defect in collagen synthesis.

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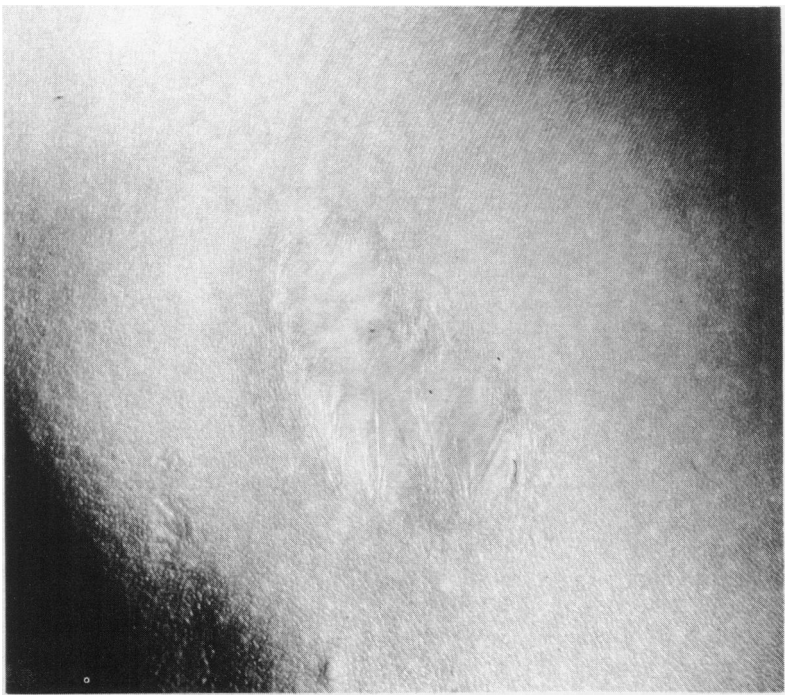

Fig. 3—"Cigarette paper" scar of knee area. Such scarring makes maxillofacial surgery difficult in Ehlers-Danlos syndrome patients.

The symptoms of TMJ problems in EDS patients are similar to those in others, but certain features of treatment are different. First, the range of mandibular opening must be restricted by prescription even more so than in other patients. That is, activities such as eating large sandwiches, opera singing, and lengthy dental appointments requiring extreme opening must be strictly avoided. Yawning must be controlled by gentle pressure on the chin to avoid excessive opening. Mandibular exercises must be moderate to avoid excessive stress on abnormal retrodiscal collagen fibers. Splint therapy may require more time than is usual to achieve results. Finally, oral surgical procedures, especially to repair retrodiscal tissues, are contraindicated because of the poor healing of tissues and the unusual scar formation. In many ways EDS serves as a model of internal derangements of the TMJ. The fragility of the retrodiscal collagen fibers in EDS underscores the vulnerability of these fibers in nonafflicted patients. The importance of reducing stress on the retrodiscal fibers as a major goal of treatment then becomes apparent. The necessity of a thorough medical history, physical examination of limb joints, and knowledge of collagen disorders and arthritis in treating TMJ patients cannot be overemphasized. Patients suspected of having EDS should be referred to a rheumatologist and geneticist.