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Image Correspondence

Ehler Danlos syndrome

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A 37-year-old male presented to skin department with complaint of multiple scars and lesion on chest. On dermatological examination, multiple well-defined and atrophic scars were present on forehead and cheeks. Epicanthal folds were also prominent [Figure 1a]. Multiple cigarette paper scars were present over both wrists, forehands, and knees [Figure 1b]. Multiple skin colored to white colored well-defined 1 cm \times 2 cm papules identified as spheroids were present on the upper chest [Figure 1c]. Skin stretching revealed hyperextensible skin with normal recoil [Figure 1d]. Skin was soft and there was history of skin fragility. Joint mobility was normal. Systemic

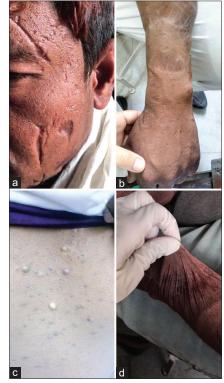


Figure 1: (a) Scars on face and epicanthal folds. (b) Cigarette paper scars on wrist and forearm. (c) Subcutaneous spheroids. (d) Hyperextensible skin.

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examination was normal. There was no history of bleeding tendency. Family history was also not significant.

We considered a diagnosis of Ehler Danlos system and reviewed the crtieria for the same as mentioned below:

- Major criteria
 - 1. Skin hyperextensibilty and atrophic scarring
 - 2. Generalized joint hypermobility.
- Minor criteria
 - 1. Easy bruising
 - Soft and doughy skin
 - Skin fragility 3.
 - 4. Molluscoid pseudotumors
 - Subcutaneous spheroids
 - Hernia (or history thereof) 6.
 - 7. Epicanthal folds
 - Complications of joint hypermobility 8.
 - Family history of first degree relative who meets clinical criteria.

Our patient fulfilled one major and four minor criteria. Confirmatory molecular testing for mutations in gene encoding type V collagen (COL5A1 and COL5A2) is necessary for the final diagnosis but it could not be done due to lack of availability of resources.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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