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Ehlers-Danlos syndrome, a very rare condition associated to hidradenitis suppurativa

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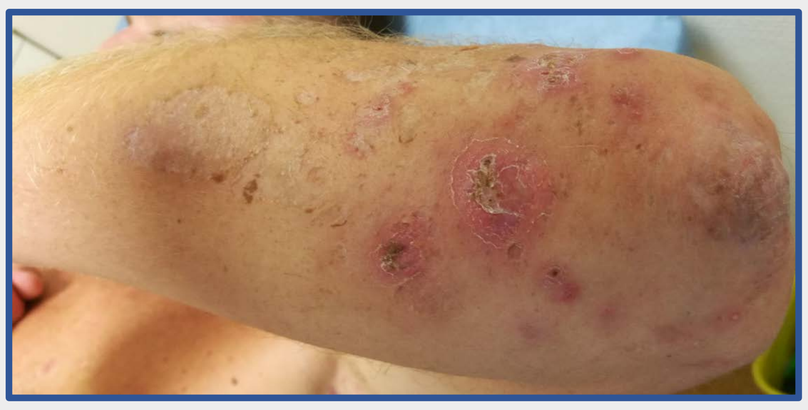
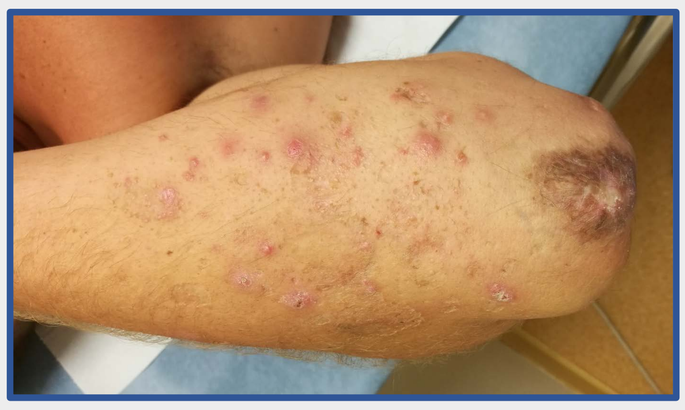
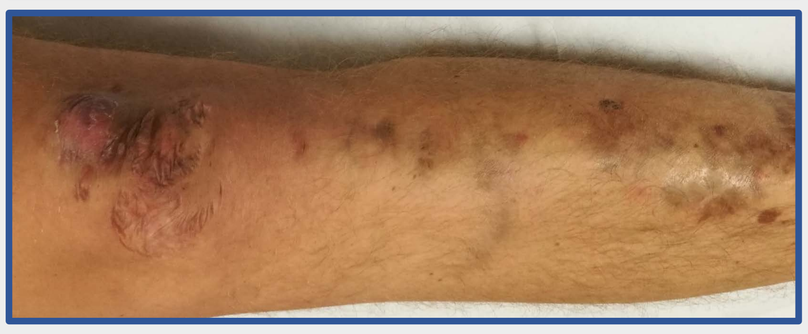
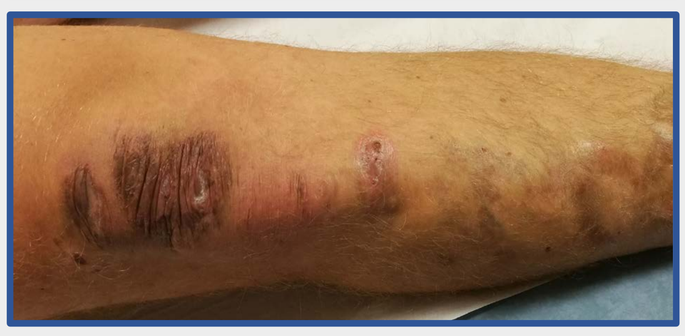
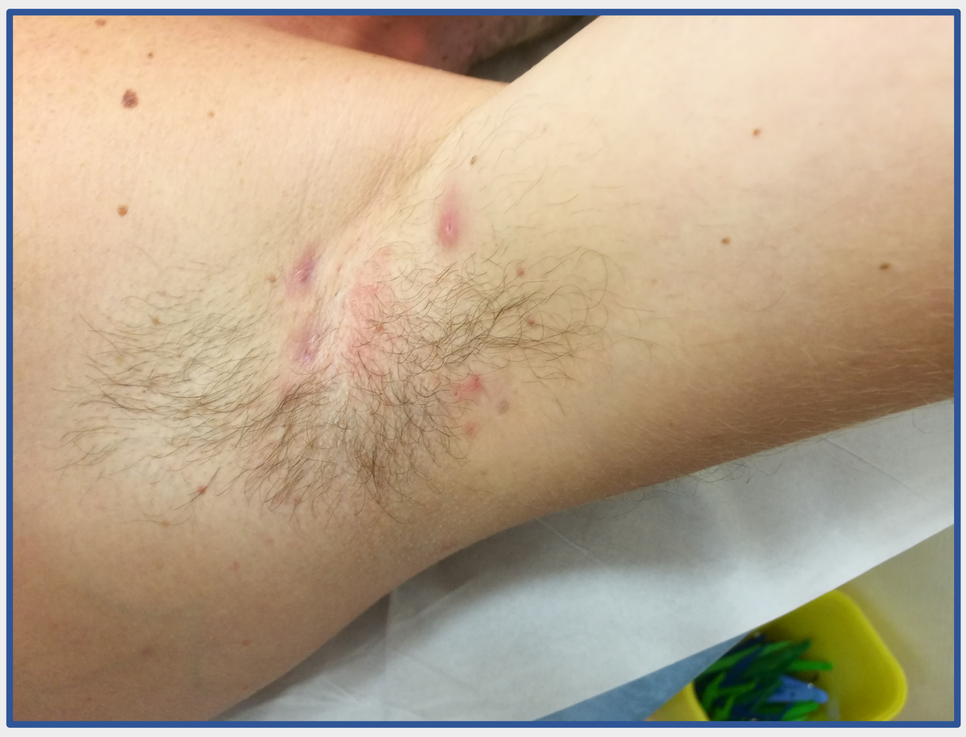
Introduction

- Ehlers-Danlos syndrome (EDS) is an heterogenous group of genetic disorders causing skin hyperextensibility and delayed wound healing.
- Hair follicles of the skin in patients with EDS are macroscopically more enlarged than those of healthy subjects. Enlarged hair follicles are also observed in the skin of patients suffering from hidradenitis suppurativa (HS).
- The association between EDS and HS has never been described previously.

Case report

- We report the case of a 24-year-old patient with both EDS and HS
- EDS evolved as a familial trait (father, two brothers).
- HS began at the age 16 in the elbows (olecranons) and secondarily affected armpits, groins, abdominal fold, neck and back.
- Other atypical affected sites were posterior part of the proximal third of the ulnas and tibial anterior crests.

Abcesses and inflammatory nodules in the elbows and tibias of both sides – Inflammatory nodules in the left axilla

Discussion

- The abnormal expression of the collagen-related genes in EDS is responsible for the increased extensibility and pliability of the skin.
- This may allow the hair follicles to become enlarged
- In parallel, HS lesions tend to develop in skin areas submitted to repeated friction which may also contribute to cause enlargment of hair follicle.
- Transcriptome analysis performed in HS skin revealed that collagen-related genes are among the most differentially expressed genes in comparaison to unaffected skin.

Conclusion

- The association between EDS and HS is very relevant
- The genetic basis of EDS may contribute to improve our understanding of HS pathophysiology.