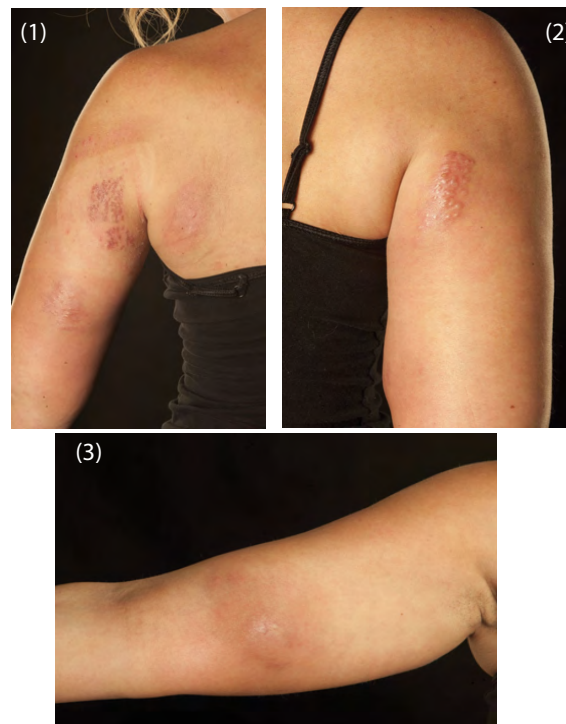


Case Blog

Alpha-1-Antitrypsin Deficiency Panniculitis

Rohi Shah*, Puja Channiyara and Mehreen Ahmad

Department of Plastic Surgery, Leicester Royal Infirmary, Infirmary Square, Leicester LE1 5WW, United Kingdom



Figures 1-3: Erythematous plaques and nodules distributed along her upper arms, back, abdomen and thighs.

Case Presentation

A 27-year-old woman presented with multiple indurated, ill-defined, erythematous plaques and nodules distributed along her upper arms, back, abdomen and thighs (Figures 1-3). Cutaneous biopsies revealed inflammatory infiltrates with fibrotic changes within the dermis and septa. Serum α 1-antitrypsin (A1AT) level was low at 0.30 g/l (RR: 1.10-2.40 g/l).

The clinical, hematological and histological findings were in keeping with a diagnosis of A1AT deficiency panniculitis. Further genetic phenotyping revealed a homozygous ZZ variant.

Panniculitis is a rare, but recognised cutaneous manifestation of A1AT deficiency. Histopathological examination usually reveals a lobular panniculitis, but may demonstrate septal involvement. It is often refractory to many medical treatments including corticosteroids and immunosuppressant. However, treatments with dapsone, tetracyclines and A1AT repositioning have been used with varying efficacy [1,2].

Although surgical debridement should be avoided in the acute stage, patients may seek surgery for cosmetic purposes.

References

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*Corresponding author: Rohi Shah, Department of Plastic Surgery, Leicester Royal Infirmary, Infirmary Square, Leicester LE1 5WW, United Kingdom, Tel: 07878889654; E-mail: shah.rohi@gmail.com

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