

CME MCQ – 28

CME MCQ – Influence of the Human Leukocyte Antigen Complex on the Development of Cutaneous Fibrosis: An Immunogenetic Perspective

The following questions are based on the Review article in Acta Derm Venereol 2010; 90: 563–574 by Sara M. McCarty; Serological Studies in Bullous Pemphigoid: Influence of the Human Leukocyte Antigen Complex on the Development of Cutaneous Fibrosis: An Immunogenetic Perspective.

- Cutaneous fibrosis covers a variety of human disorders with differing aetiology, but with a common dysregulation of connective tissue metabolism, particularly of dermal fibroblasts. Specific examples of cutaneous fibrosis include:
 - keloid disease
 - hypertrophic scars
 - scleroderma
 - morphea
 - eosinophilic fasciitis
- There appears to be a genetic predisposition to keloid development. Evidence suggests a predilection to keloid formation with:
 - autosomal dominant -
 - autosomal recessive -
 - X-linked recessive inheritance patterns
 - candidate genes of the highly polymorphic human leukocyte antigen loci
 - ethnic variation
- Research into the aetiology of the disease has been hampered by a number of factors. Indicate factors that may limit further research in the disease:
 - keloids only affect humans
 - extent of the polymorphism and linkage disequilibrium of specific human leukocyte antigen alleles
 - the time lapse between initial cutaneous injury and keloid formation is often several weeks to years
 - keloids mostly develop in areas lacking sebaceous glands
 - differentiation of keloid and hypertrophic scars still seems to be inconsistent in the clinical setting (despite the long-established guidelines)

Recommended answers:

1. A, B, C. Comment: Morphea (D) and eosinophilic fasciitis (E) has not been mentioned in the article, probably because the fibrosis in these conditions affect mainly dermis and subcutaneous fat as it can be seen very pronounced in pansclerotic morphea – a rare form of localised scleroderma that typically presents in childhood. Eosinophilic fasciitis mainly causes fascial fibrosis.

2. A, B, C, D, E.

3. A, B, C, E. Comment: Keloids very rarely develop in areas lacking sebaceous glands (D), such as the genitalia and the palms and soles.