PREGUNTA 10

Pregunta vinculada a la imagen nº 10.
Mujer de 80 años con antecedentes de hipertensión arterial, dislipemia y artrosis cervical en tratamiento con enalapril/hidroclorotiazida y simvastatina desde hace años y esporádico con paracetamol. Consulta por lesiones cutáneas pruriginosas en tronco y extremidades de 2 semanas de evolución para las que no ha realizado tratamiento médico (Imagen). La paciente se encuentra hemodinámicamente estable y afebril cuando acude al Servicio de Urgencias. Señale la opción CORRECTA:

1. En la biopsia cutánea realizada de una lesión de reciente aparición esperaría encontrar espongiosis epidérmica, vesículas espongióticas y un infiltrado eosinofílico.
2. Las lesiones son compatibles con una pustulosis exantemática aguda generalizada por lo que iniciaría tratamiento con amoxicilina/clavulánico por vía oral.
3. Con la sospecha de una enfermedad ampollosa autoinmune se realiza una biopsia de las lesiones para inmunofluorescencia directa. Se cursa ingreso hospitalario y se inicia tratamiento con prednisona a dosis de 1mg/kg/día.
4. Con la sospecha de eccema diseminado se realiza biopsia de lesiones para hematoxilina-eosina y se inicia tratamiento con prednisona a dosis de 1mg/kg/día.

Pregunta controvertida sobre un caso clínico asociado a imagen. Nos presentan a una mujer anciana con factores de riesgo cardiovascular que comienza con clínica de lesiones cutáneas muy pruriginosas de reciente aparición sin otra clínica asociada. En la exploración física podemos observar múltiples erosiones y costras serohemáticas sobre una base de piel eritemato-edematosa. Dicho cuadro clínico en una paciente anciana debe orientar al diagnóstico clínico de un penfigoide ampolloso.

Bibliografía:
"McKee's Pathology of the Skin" Calonje E, Brenn T, Lazar A, McKee PH. 4th Edition. 2012 Capítulo 4, página 121

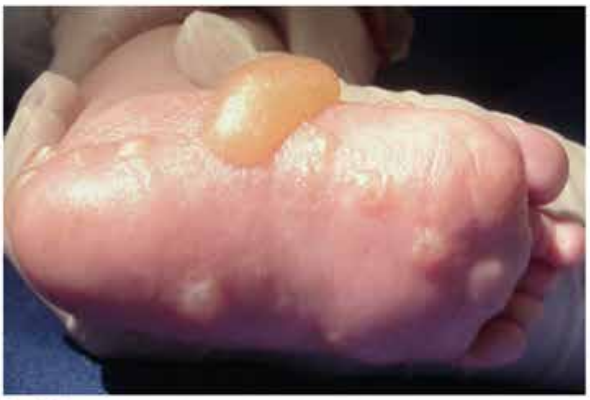

Fig. 4.61
Childhood BP: plantar involvement is sometimes the only site of disease. By courtesy of M. Liang, MD, The Children's Hospital, Boston, USA.

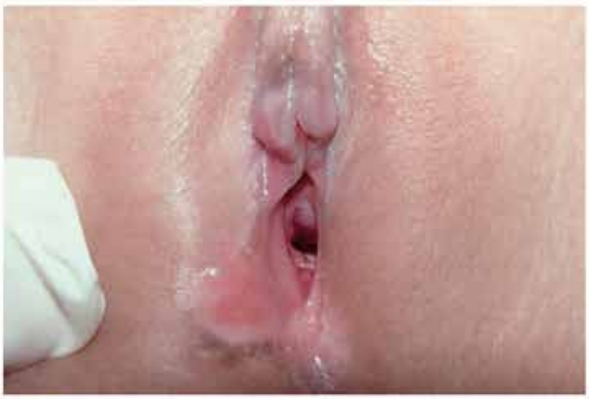

Fig. 4.62
Childhood BP: note the perineal scarring and isolated blister. By courtesy of M. Liang, MD, The Children's Hospital, Boston, USA.

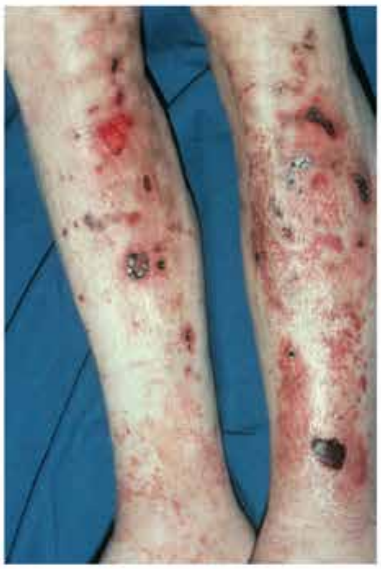

Fig. 4.63
Localized pemphigoid, nonscarring variant: lesions are found particularly on the lower legs of females. The prognosis is usually good, but occasionally the condition can become generalized. By courtesy of R.A. Marsden, MD, St George's Hospital, London, UK.

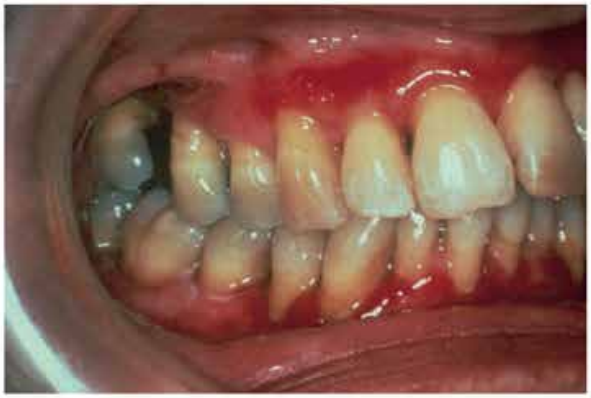

Fig. 4.64
Desquamative gingivitis: note the intense gingival erythema and retraction. Such features may also be seen in mucous membrane pemphigoid and pemphigus. By courtesy of P. Morgan, FRCPath, London, UK.

Pathogenesis and histological features

The histological features of bullous pemphigoid depend to some extent upon the age of the lesion biopsied. Early erythematous and urticarial lesions most often show upper dermal edema associated with a perivascular lymphohistiocytic infiltrate accompanied by usually conspicuous eosinophils (*Figs 4.65* and *4.66*). Eosinophilic spongiosis is sometimes evident and occasionally, if eosinophils are present in sufficient numbers, flame figures may be a feature. Mild interface changes characterized by basal cell hydropic degeneration can be seen in early or prodromal lesions.

If the biopsy is taken from an established blister, the changes are most often those of an inflammatory (cell-rich) variant.[73] The blister, which is subepidermal, is typically unilocular and covered by attenuated epithelium (*Fig. 4.67*). In early lesions the roof epidermis may appear unaffected or show occasional to even confluent necrotic basal keratinocytes. The blister contents include coagulated serum, fibrin strands, and large numbers of inflammatory cells including conspicuous eosinophils (*Fig. 4.68*). Variable numbers of neutrophils may be present.

A typical finding in bullous pemphigoid is retention of the dermal papillary outline (festooning) which project like sentries into the vesicle cavity (*Fig. 4.69*). The underlying dermis is inflamed and usually shows widespread severe edema. An infiltrate of eosinophils and mononuclears surrounds the blood vessels and extends between the adjacent collagen bundles. Leukocytoclasis is not seen and features of vasculitis are absent. The adjacent papillary dermis is often edematous and, very occasionally, eosinophil microabscesses are a feature (*Fig. 4.70*). Exceptionally rarely, neutrophil microabscesses may be seen (see vesicular pemphigoid), raising diagnostic confusion with dermatitis herpetiformis. Eosinophilic spongiosis is also sometimes evident in the adjacent epidermis (*Fig. 4.71*).[74]

Cell-poor (noninflammatory) features are occasionally seen if biopsies are taken from lesions arising on noninflamed skin (*Fig. 4.72*). Because inflammatory cells are sparse or, exceptionally, even absent in such cases, there may be considerable problems with the differential diagnosis, particularly if adequate clinical information and immunofluorescence findings are not available.