Russell Body Dermatitis Due to Syphilis

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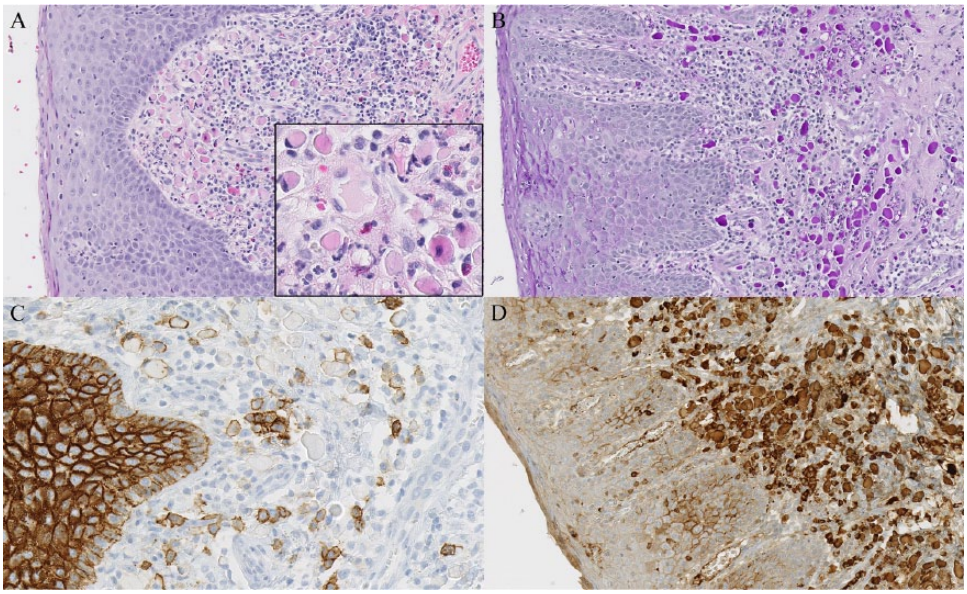

Figure 1. Preputial skin specimen demonstrating a mixed type inflammatory infiltrate of the upper dermis, along with round homogeneous eosinophilic structures, corresponding to Russell bodies (A, hematoxylin-eosin, original magnification ×200 and ×750 for the inset). These structures show strong periodic acid-Schiff (PAS) positivity (B, PAS, original magnification ×200). CD138 staining underlines plasma cells and shows a weak linear staining around some of the Russell bodies, confirming their localization in plasma cells (C, CD138, original magnification ×400). Kappa immunohistochemical stain highlights the numerous Russell bodies in the dermis (D, kappa, original magnification ×200).

A 38-year-old man with a history of HIV, hepatitis B, Hodgkin lymphoma, Guillain-Barré syndrome, and syphilis was referred to the urologist due to phimosis. The patient presented with increasing difficulty to pull back his foreskin associated with pain and redness of the skin. After several failed attempts of local treatment including corticoids and antifungal therapy, the patient underwent circumcision.

Macroscopically, the skin specimen presented a slightly heterogeneous color but with no obvious lesion. The histological examination revealed a mixed-type inflammatory infiltrate in the dermis associated with epidermal hyperplasia and parakeratosis (Figure 1A and B). Within this inflammatory infiltrate, round homogeneous eosinophilic structures were observed, and both the hematoxylin-eosin and the CD138 staining revealed these structures corresponded to inclusions located in the cytoplasm of plasma cells (Figure 1C). In some areas, it was difficult to recognize on the hematoxylin-eosin staining that the cells in which these structures were located were plasma cells. Hence, these structures corresponded to Russell bodies. The plasma cells had a polyclonal immunoreactive pattern with positive staining for both kappa and lambda (Figure 1D). In view of these findings, the diagnosis was Russell body dermatitis in a context of chronic syphilis.

Russell bodies are considered to be nondegradable, condensed, unreleased immunoglobulin components, as a result of an overstimulation of plasma cells and a block in the normal secretion pathway of immunoglobulins. These

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unreleased immunoglobulins accumulate in the rough endoplasmic reticulum of these plasma cells and may totally fill up the cytoplasm.[1] They sometimes can be seen in multiple myeloma,[2] and in theory, they can occur in any chronic inflammation with a lot of plasma cells, but in practice they are mainly seen in chronic inflammation of the upper gastrointestinal tract.[3,4] With the exception of rhinoscleroma, Russell bodies are extremely rarely observed in the skin.[1] The occurrence of Russell bodies in the setting of syphilis seems logical, since chronic syphilis-related dermatitis is characterized by a chronic lymphoplasmacytic infiltrate,[5] yet there are no reports on this topic, suggesting this is an extremely rare event. Why Russell bodies seem to be much rarer in this setting compared with chronic inflammation of the upper gastrointestinal tract is not clear. Cases of Russell body dermatitis in the setting of chronic syphilis and other types of dermatitis with a lot of plasma cells might be overlooked due to the difficulty and the rarity of this entity, especially since it can be difficult at first sight to appreciate that the round homogeneous eosinophilic structures are located in the cytoplasm of plasma cells. The differential diagnosis of Russell bodies include microorganisms such as histoplasmosis or cryptococcosis, fungi, Michaelis-Gutmann bodies, deposits of amyloid, colloid bodies, or elastic bodies.[1] These diagnoses can be excluded using periodic acid-Schiff, Grocott, Congo red, Miller, and CD68 stains. When eosinophilic homogeneous round structures are encountered in an inflamed skin biopsy, Russell body dermatitis due to syphilis should be considered in the differential diagnosis and immunohistochemistry for CD138 and kappa and lambda can be used as confirmatory staining.

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