INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

CUTANEOUS ROSAI-DORFMAN DISEASE WITH UNIQUE SKIN LESIONS AND LIVER CIRRHOSIS

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Background: Rosai–Dorfman disease (RDD) is a rare histiocytic disorder characterized by a benign proliferation of CD68–positive and CD1a-negative histiocytes within the sinus of lymph nodes. Extranodal involvement has been documented in 43% of cases with the most frequent sites being skin, soft tissue, upper respiratory tract, and multifocal bone. RDD confined to the skin has been recognized as cutaneous RDD (cRDD). Skin lesions typically begin with papules or plaques and form nodules and satellite lesions. However, yellowish plaques with severe swelling spread in our patient. Histopathological findings typical of RDD were observed in liver and skin biopsy specimens. We present a rare case of cRDD associated with severe liver cirrhosis.

Observation: A 62-year-old Japanese female who had been suffered from non-viral liver cirrhosis for 10 years noticed erythematous nodules and plaques with pigmentation 3 years ago. The lesions exacerbated with severe swelling on the bilateral eye lids, cheeks, forehead and neck, and she eventually couldn't open her eyelids. A wide range of yellowish plaques were found from the forehead to anterior neck except for the nose, but without lymphadenopathy. Skin biopsies revealed dense dermal and subcutaneous infiltrates essentially composed of foamy mononucleated histiocytes, Touton giant cells, and lymphocytes. Immunohistopathology showed that histiocytes were positive for CD163 and CD68, negative for CD1a. Emperipolesis was observed in some parts. The BRAF-V600E mutation was not found. The liver biopsy specimen showed pathological findings similar to the skin lesions. The bone scintigraphy did not show any involvement in the bones. From these findings, we made a diagnosis of cRDD with liver cirrhosis as extranodal sites. Oral corticosteroids for 3 weeks showed no response to both skin lesions and liver dysfunction.

Key message: We report a case of cRDD with unique skin lesions and liver cirrhosis.