SKIN MANIFESTATIONS OF INTERNAL DISEASE

SINUS HISTIOCYTOSIS OF ROSAI-DORFMAN WITH CUTANEOUS REVELATION: ABOUT TWO CASES

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Background: Rosai-Dorfman disease (RDD),also named as sinus histiocytosis with massive lymphadenopathy,is a rare histiocytosis characterized by massive bilateral painless cervical lymphadenopathy and emperipolesis in tissue sections.We report the observation of 2 patients with a RDD,cutaneous and lymph node revealing purely cutaneous and cutaneous pure.

Observation:
Case 1: A 52-year-old patient,in good general condition had a progressive papulo-nodular eruption in the right flank and right gluteal region for 1 year. The lesions were erythemato-violin, infiltrated, with mammillated surfaces in places,mesuring 2cm and 9cm respectively.Histological examination with immunohistochemistry showed images of emperipolesis which was S100 positive and CD1a negative .
A PET scan,revealed polyadenopathies in the basicervical region, the mediastinum and the hiles with hypermetabolism.The ANA was positif at 640.The patient was treated with thalidomide 100 mg/day, in the absence of improvement,methotrexate at a dose of 25 mg/week was started with a slight improvement. The management was completed by an excision of the cutaneous lesions with a good evolution and the addition of a general corticosteroid 1 mg/kg/day.
Case 2: A 47-year-old woman,had for 1 year, 2 nodular lesions on an erythematous placard in her back. The clinical examination found two erythematous nodules, with presence of papular lesions satelites. The histological study with IHC confirmed the diagnosis.The extension assessment was negative. Due to the large size and unsightliness of the lesion, very strong class dermocorticoid treatment was undertaken, with partial regression.

Key message: The originality of these observations lies in the rarity of RDD revealed by pure cutaneous involvement in the first case and the purely cutaneous manifestation in the second.Cutaneous involvement is the most frequent extra-nodal involvement often associated with other localizations.The pure cutaneous form in the 2 patients made the diagnosis difficult.The treatment is not codified,the therapeutic abstention is rule in the absence of functional discomfort.