INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

SCLEREDEMA ADULTORUM OF BUSCKHE: ATYPICAL PRESENTATION

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Background: Scleredema adultorum of Busckhe (SAB) is a rare sclerotic disorder usually involving the neck, shoulders, trunk and arms. Periorbital edema as the only manifestation of SAB is uncommon. We report two cases of SAB with prominent palpebral edema.

Observations: Observation 1: A 34-year old female patient presented to our department for a five months history of a marked bilateral periorbital edematous swelling with intense skin induration causing a palpebral occlusion. Histologic examination of a skin biopsy specimen showed thickening of the collagen bundles in the dermis, which were separated from one another by clear spaces. A diffuse pericapillary inflammatory infiltrate was noted. Alcian blue stains were positive. These histologic features in conjunction with the clinical picture confirmed the diagnosis of SAB. The results of laboratory investigations including blood glucose and serum and protein electrophoresis were normal.
Observation 2: A 36-year-old man consulted for a 10 days history of asymptomatic bilateral eyelid edema. On examination, he had an important bilateral periorbital edematous swelling with intense skin induration causing a palpebral occlusion. He had also erythematous induration of the skin around the neck, shoulders and back. Histologic examination revealed SAB features. The results of laboratory investigations were normal except for a high level of blood glucose.

Key message: The specific clinic and histologic features supported the diagnosis of SAB in both patients. SAB is a fibromucinous connective tissue disorder of unknown cause that belongs to a group of scleroderma-like diseases. Periorbital edematous swelling is exceptionally described in SAB. Only one previous case was reported in a 48-year-old patient with no underlying pathology, such as the case our first patient. Associated lesions of the neck, back, shoulders and hands was present in the second observation. In addition, an inaugural diabetes was discovered in this patient.