Sweet's syndrome - rare associations

Síndrome de Sweet - raras associações

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To Editor

I read a recent paper prepared by Kaur et al., describing a 33-year-old man affected by Sweet's syndrome associated with chronic neutrophilic leukaemia.[1] They highlighted the rarity of the association based on the finding of a unique previous report.[1] The patient presented with fever, visceromegaly, scattered violaceous papules and plaques, in addition to edema and pseudovesiculation; the diagnosis of the syndrome was based on upper dermis neutrophilic infiltrate and leukocytosis.[1-5] The patient underwent a course of hydroxyurea and a systemic corticosteroid and improved rapidly.[1-5] Sweet's syndrome coexisting with chronic neutrophilic leukemia is an exceeding rare condition, and case reports can contribute to enhance the knowledge about factors playing a role in associations.

Although the report is well documented; some additional comments should be addressed in this setting. The authors emphasized the hematological conditions and solid tumours among the etiologic factors of the Sweet's syndrome;[1] notwithstanding, this rare entity may be idiopathic or drug-induced as well.[2-5] The main related hematological malignancies are acute myeloblastic leukaemia, Hodgkin's lymphoma, large B-cell lymphoma, myelofibrosis, myelodysplastic syndrome, and myeloproliferative neoplasms.[4] More frequently related solid tumours include breast, and genitourinary and gastrointestinal tracts.[4] Anticancer drugs may also be involved, as the example of all-trans retinoic acid, azacitidine, bortezomid, decitibine, granulocytic colony stimulating factor, imatimib mesylate, and lenalinomide.[4] Additional concern is about possible side-effect of hydroxyurea, as discussed in the case of a 79-year-old woman with chronic myeloid leukaemia and unique bullous lesion in the site of the venepuncture.[2] As occurred with the man reported by Kaur et al., the lesion of the old woman cured with prednisone.[1,2] A possible initial diagnostic challenge may be about similar skin changes found in leukemia cutis; nevertheless, this hypothesis can be ruled out by the absence of the characteristic histopathology

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data.[2,4] Worthy of note, the female also presented with an ulcerated lesion in the region of the vaginal introitus, and both changes are manifestations commonly observed in women with diagnosis of Behçet disease.[3]

This uncommon disease, which involves pathergy phenomena is rarely related to Sweet's syndrome.[3] Concurrence of these conditions was described in a 49-year-old woman with Behçet disease and presenting an isolated bullous lesion consistent with Sweet's syndrome in the site of venepuncture.[3] Although without current clarification, some authors hypothesize about common pathogenic factors.[3]

Sweet's syndrome is a rare condition which may be idiopathic, paraneoplastic or drug-induced. The exact pathogenesis is not well-known, and clinical diagnosis may be challenging because the main features can be mistaken by other disorders. Thence, well-documented case studies as commented herein can contribute to clear both pathophysiology and associations of this rarely reported syndrome.

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