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Paraneoplasic Behçet's syndrome associated with chronic myeloid leukemia

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Conflict of interest: the authors declare no conflict of interest.

Ethical statement: the spouse of the subject has given her written informed consent to publish the case (including publication of images).

Author contributions: Marta Moreira was the patient's doctor and wrote the manuscript. Isabel Tarrio and Alda João Andrade helped during research and management while the patient was hospitalized. Luis Lopes was responsible for the revision of the manuscript.

CASE REPORT

A 49-year-old male presented with a three-week history of abdominal pain, bloody diarrhea and fever. The patient also reported oral ulcers, weight loss and asthenia, as well as papulo-pustular lesions on his limbs and a recurrent ulcer in the lip (Fig. 1) in the previous year. During hospitalization, he developed pathergy at venipuncture sites and painful scrotum ulcers (Fig. 1).

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Laboratory showed pancytopenia and elevated C-reactive protein (CRP). Viral and autoimmune tests were negative. Abdominal computed tomography (CT) revealed thickening of the ileocecal region with adenopathies (Fig. 2). Blood smear and myelogram were compatible with chronic myeloid leukemia (CML). Bone marrow culture and BK were negative. Karyotype revealed no changes, namely, no trisomy of the 8th chromosome.

Ileocolonoscopy revealed aphthoid erosions of the ileocecal mucosa and ovoid punched-out cecal ulcers. Biopsies showed intense chronic inflammation in the lamina propria and submucosa with erosions and ulcers (Fig. 3).

Thus, presenting five points in the International Criteria for Behçet's Disease, this diagnosis was assumed as a paraneoplastic manifestation of CML. Corticosteroids improved symptoms, but the patient died three weeks later due to a blastic crisis.

DISCUSSION

Behçet's syndrome has been reported in association with CML (1,3), some concurrent with or following treatment with interferon-α or hydroxyurea (2). Although the pathogenesis remains unclear, there is increasing awareness of its link to hematological malignancies and trisomy of the 8th chromosome (1,3).

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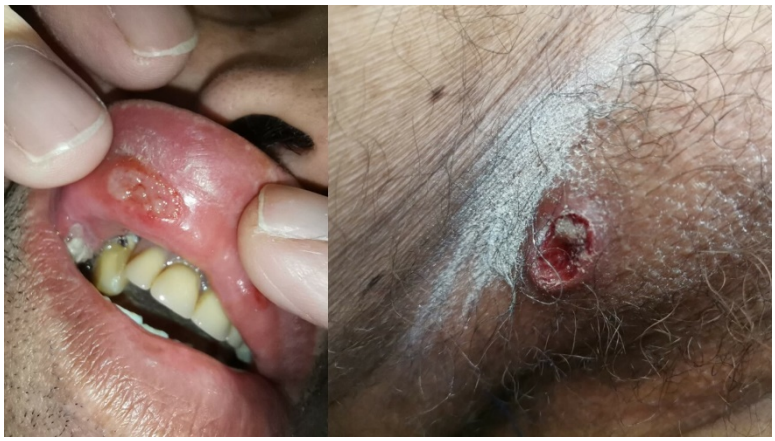

Fig. 1. Ulcerated chronic and recurrent lesion on the upper lip (on the right) and painful ulcer of the scrotum that the patient developed during hospitalization (on the left).

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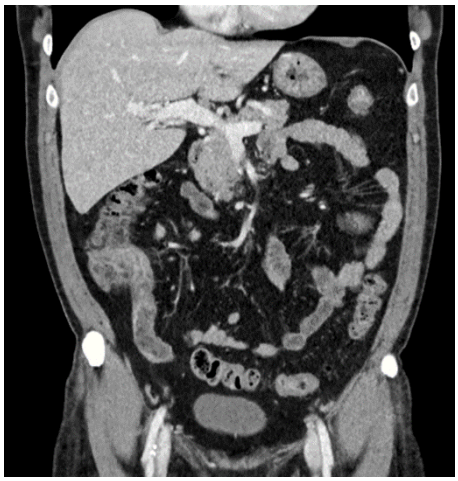

Fig. 2. Computed tomography (CT) scan showing thickening of the ileocecal region and hyperenhancement of the wall, with local adenopathies.

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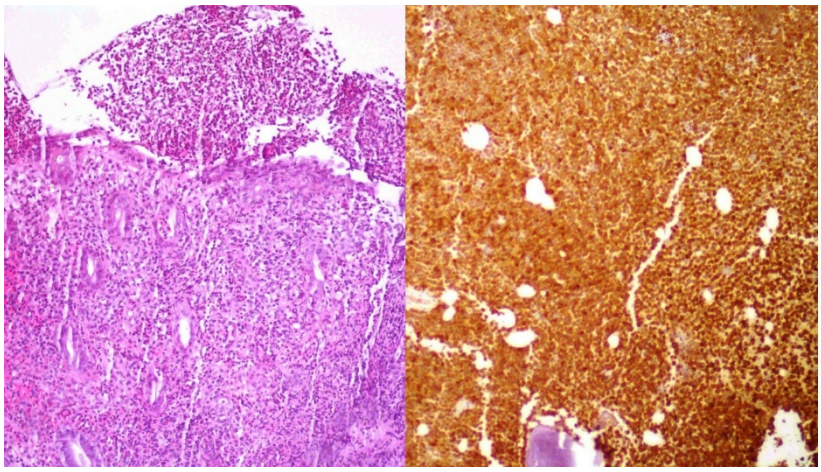

Fig. 3. Cecal region biopsies with intense chronic inflammation in the lamina propria and submucosa with erosions and ulcers (on the left) and bone marrow biopsy which were positive for the myeloperoxidase stain (on the right).