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IDIOPATHIC PYODERMA GANGRENOSUM ASSOCIATED WITH STERILE SPLENIC ABSCESS IN A PEDIATRIC PATIENT.

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BACKGROUND

Pyoderma gangrenosum (PG) is a rare condition in pediatric age group. It´s a neutrophilic dermatosis, characterized by inflammatory papules and pustules that rapidly progress to painful, deep and disseminated ulcers. The idiopathic form is less frequent, with most cases being secondary to systemic diseases, including inflammatory bowel disease, hematologic disorders and autoinflammatory syndromes. The diagnosis is suggested by clinical and histopathological data, and treatment is based on corticosteroids and immunosuppressive drugs. Systemic involvement is broad and can affect visceral organs. There are few literature reports about PG associated with splenic abscess in pediatrics.

CASE REPORT

R.N.L, 13-years-old teenager, started a 10-day cutaneous lesions characterized by painful erythematous papules in forehead, with dissemination to right shoulder, axillas and lower limbs, associated with unmeasured fever. Lesions rapidly became pustular, with ulceration and drainage of purulent and bloody secretion. She denied exanthema, oral ulcers, weight loss, urinary or gastrointestinal symptons. Physical examination: multiple ulcerated, painful, deep lesions with necrotic borders and granulomatous fundus, mainly in lower limbs, besides important arthritis in ankles and nodular lesion in right axilla with drainage of purulent secretion. Treatment with broad-spectrum antibiotic therapy showed no improvement. Initial complementary investigation revealed anemia and elevated inflammatory markers, as well as negative cultures and serologies. Colonoscopy and neoplasia screening were normal, besides ANA and ANCA negative. Thoracic computed tomography (CT) showed ulcerated heterogeneous lesion in the right axilla, suggestive of hidradenitis suppurative, and abdomen CT found splenomegaly associated with multiple rounded lesions, measuring between 1.5 and 2.9 cm, suggestive of splenic abscesses. Cutaneous biopsy revealed a marked neutrophilic infiltrate with epithelioid histiocytes and multinucleated giant cells, resembling non-caseous granulomas, suggestive of PG. Prednisone 1.0 mg/kg/day was started, evolving with progressive cutaneous improvement, receiving discharge with prednisone and dapsone 100 mg/day one month later. After two months the patient was asymptomatic, only with cicatricial lesions and disappearance of splenic abscesses.

CONCLUSION

Although uncommon, PG may present with extracutaneous manifestations. The finding of visceral abscesses associated with PG does not require additional antibiotic treatment, and its regression is expected with treatment of underlying disease. The presence of suppurative hidradenitis and pyogenic arthritis, in association with PG, suggests the possibility of PAPASH syndrome (pyogenic arthritis, PG, acne and suppurative hidradenitis), an autoinflammatory syndrome, which may be confirmed by genetic test.