Pyoderma vegetans and ulcerative colitis

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A 35-year-old man presented with erythematous and crusted lesions over the face, forehead and chest of two weeks duration. It initially began as small pustular lesions over the face, which gradually evolved to form large erythematous, edematous, painful and crusted lesions. He also developed similar lesions over the forehead and chest. He had history of intermittent diarrhea with blood and mucus for the last two months. He had no other systemic symptoms. He was diagnosed to have ulcerative colitis two years previously for which he was initially on medications (mesalamine, a derivative of salicylic acid, which acts as an anti-inflammatory agent) that were later stopped due to symptomatic improvement. Dermatological examination showed multiple, large, erythematous, exudative and crusted cutaneous plaques over the face, forehead and chest [Figure 1]. There was no involvement of mucosal surfaces, groin or any other areas. The abdominal examination was normal and there was red blood on the examining finger on rectal examination.

The laboratory examinations disclosed an elevated white cell count of 16.2 x 10^9/L (Normal 4.0 - 10.0 x 10^9/L), anemia (Hb 9.6 gm/dL) and raised ESR of 60 mm/1^st hour. Bacterial and fungal cultures from the cutaneous lesion were negative. The skin biopsy revealed subepidermal micro-abscess containing neutrophils and eosinophils with a few lymphocytes and a superficial and deep dermal inflammatory infiltrate composed mainly of neutrophils. The edge of the lesion showed hyperkeratosis and irregular acanthosis [Figure 2].

There was no acantholysis and intra-epidermal micro-abscesses. Stains for fungi were negative. Colonoscopy showed erythematous granular friable mucosa with macro-ulcerations extending up to the transverse colon and histopathology was characteristic of ulcerative colitis.

Treatment with topical corticosteroids (betamethasone dipropionate 0.1% cream) led to improvement of the skin lesions. Total resolution of all skin lesions was achieved with oral prednisolone at 40 mg daily for four weeks [Figure 3]. He was also started on mesalamine; metronidazole and a systemic antibiotic for ulcerative colitis. The corticosteroids were gradually tapered and stopped over two months and he was continued on mesalamine. There was prompt relief of colitis and no recurrence of skin lesions. The patient is on oral mesalamine and asymptomatic on follow-up for the last one year.

Discussion

The association of pyoderma vegetans (PV) with inflammatory bowel disease is well known but rarely reported in literature.[1,2]

Cutaneous manifestations are common in inflammatory bowel disease. PV is a rare benign cutaneous, chronic, inflammatory pustular and vegetating condition characterized clinically by erythematous vesiculo-pustular, exudative, vegetating plaques. The condition was first described by Hallopeau in 1898 as pyoderma vegetante and later in 1949 McCarthy described similar findings limited to the oral mucosa and coined the term pyostomatitis vegetans and considered it to be the oral counterpart of Hallopeau’s pyodermites vegetans.[3]

The etiology of pyoderma vegetans is not known. It has been described in patients with an immunosuppressive state or a dysfunction of the immune system.[4] Other factors also have been speculated such as bacterial infections, foreign materials, tattoos, ingestion of halogens or pre-existing cutaneous neutrophilic dermatoses particularly in the setting of compromised immune function. The association of pyoderma vegetans and ulcerative colitis has rarely been reported in literature.[1,2,5] Pyodermitis-pyostomatitis vegetans, a related mucocutaneous pustular vegetating entity is strongly associated with inflammatory bowel diseases[6] although in a number of reported cases investigations for inflammatory bowel disease have revealed no disease; suggesting that a solely mucocutaneous variant exists.[7] It is still uncertain whether pyodermitis-pyostomatitis vegetans, is a separate disease or simply a variant of pyoderma vegetans.[8] The cutaneous lesions and histopathological findings of both pyoderma vegetans and pyodermitis-pyostomatitis vegetans show similar features.

Pyoderma vegetans also has been described in patients with immunological anomalies due to malignant tumors (T-cell lymphoma, chronic myeloid leukemia, paraproteinemias, colon carcinoma, seminal carcinoma) and primary...
immunodeficiency. These conditions must be excluded as the treatment is mostly directed at the underlying systemic condition. Pemphigus vegetans can be difficult to distinguish from pyoderma vegetans clinically but can be distinguished by the prominent acantholysis on histology and a positive immunofluorescence.

Usual therapeutic measures are topical antibiotics and corticosteroids, parenteral antibiotics and systemic corticosteroids. In our patient both the inflammatory bowel disease and cutaneous disease showed a parallel activity. Parallel course is also seen with pyostomatitis vegetans. There was dramatic response of both bowel symptoms and skin lesions to systemic corticosteroids and mesalamine in our case.

References


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