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Dermatology

PYODERMA GANGRENOSUM IN PATIENT OF MYASTHENIA GRAVIS AND THYMOLIPOMA

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ABSTRACT Pyoderma gangrenosum is a rare ulcerating disorder of unknown aetiology and autoimmune pathogenesis . This sterile neutrophilic dermatosis is known to occur in association with malignancy, infection, autoimmune disorders and drugs. Here we present a case of 48 year old male who was a known case of myasthenia gravis , in association with a rare benign tumour thymolipoma .Despite being on immunosuppressive agents for myasthenia gravis , he developed typical lesions of pyoderma gangrenosum.

KEYWORDS : Myasthenia gravis, Pyoderma gangrenosum, Thymolipoma

INTRODUCTION

Pyoderma gangrenosum (PG) is a rare non-infectious neutrophilic dermatosis characterised by cutaneous ulcers. Although it was first described by Brocq, the name PG was given by Brunsting, Goeckerman and O'Leary. [1]It is seen to be associated with haematological malignancies, infections, autoimmune diseases, inflammatory bowel disease and drugs.

Thymolipoma is a rare ,benign, slow-growing, encapsulated mediastinal mass constituting less than 10% of thymic tumours which does not recur after complete surgical resection. The pathogenesis of thymolipoma is still unclear, but two theories, the hyperplasia theory and the mixed tumour theory have been proposed. Various studies have suggested a relationship between thymolipoma and myasthenia gravis (an autoimmune neuromuscular disease) and it is also associated with other paraneoplastic syndromes.[2]\

We hereby report a case of myasthenia gravis with an uncommon benign tumor thymolipoma in a patient who later manifested lesions of PG while being on chronic immunosuppressive therapy.

CASE REPORT

A 48 year old male patient came with complaints of recurrent painful raw areas over the body for the last 6-7 months . About 6-7 months ago he began developing pea sized brown to black colored raised lesion first over chin , ears followed by scalp , trunk , both upper and lower limbs.These lesions gradually increased in size and ruptured leaving behind non discharging painful raw areas which healed gradually over period of 20-30 days with treatment, leaving behind brownish discoloration and wrinkling of skin . Since the last 20 days he had developed similar raw areas over right leg.There was no history of trauma , abdominal pain , altered bowel habits , low grade fever with night sweats , blood in urine or stool and recent weight loss.

Cutaneous examination revealed 4-5 discrete well demarcated, tender ulcers of varying sizes and shapes (largest being approx 7*6 cm in size and smallest being 1*1 cm size) with erythematous base , black necrotic crusting , peripheral violaceous pigmentation present over the right leg. (Figure A)

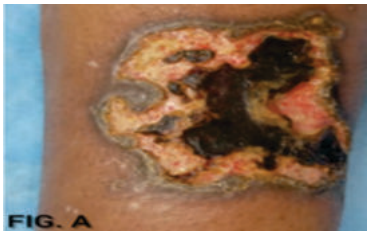

FIG. A

Peripheral pulsations were intact. Multiple hyperpigmented atrophic scars were present over the posterior aspect of the scalp, face, both ears, trunk, both upper and lower limbs and left buttock. The Pathergy test was negative.

Patient was a known case of refractory myasthenia gravis for which he was on tab.prednisolone 30 mg daily , tab methotrexate 7.5 mg/week and two gram of rituximab had been given 7-8 months ago. Open thymectomy had been done two years ago and histopathological examination of excised tissue revealed it to be thymolipoma.

Haematological investigations revealed low haemoglobin , peripheral neutrophilia with lymphopenia and antinuclear antibody (ANA) levels by immunofluorescence was reported 1:100 with a homoge neous ANA pattern.

Other investigations like renal and liver function test, urine routine examination, random blood glucose, C and P antineutrophil cytoplasmic antibodies were normal. Ultrasonography of the whole abdomen, colour doppler (arterial and venous) of both legs also didn't reveal any abnormalities. Differential diagnosis of pyoderma gangrenosum and medium vessel vasculitis were considered and 5 mm punch biopsy was performed.

Histopathological examination showed—epidermal ulceration, necrotic debris, superficial vasculitis, inflammatory infiltrate of neutrophils and lymphocytes and subepidermal fibrosis findings consistent with diagnosis of pyoderma gangrenosum. (FIGURE B- H and E,10X)

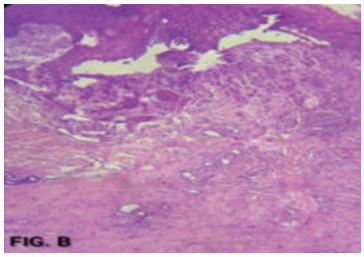

FIG. B

Keeping the same dose for methotrexate, we increased the dose of oral corticosteroids.The PG lesions remained refractory so intralesional injections of triamcinolone acetonide (40 mg/ml) were added along with platelet rich fibrin therapy. There was mild improvement in the lesions but the patient was lost to follow up.

DISCUSSION

Pyoderma gangrenosum is a neutrophilic dermatoses of unknown aetiology characterised by chronic, recurrent, cutaneous ulceration. It

commonly involves females, between 40-70 years of age. [1]

Both innate and cellular immunity play a role in pathogenesis of PG but what triggers immune dysregulation is not known. Immune dysregulation is complex involving neutrophils, T cells (overactivity of T cells with increased levels of T helper cells, IL-17, decreased T regulatory cells) and inflammatory mediators IL 1beta, IL-8, TNF alpha. [3]

Most commonly it is associated with inflammatory bowel disease (crohn's disease,ulcerative colitis), rheumatoid arthritis, haemato logical malignancies(acute myeloid leukaemia, monoclonal gammo pathies, myelodysplasia). Certain drugs like isotretinoin, gefitinib, propylthiouracil, tyrosine kinase inhibitors may trigger PG. Haim et al. suggested that immunosuppressive therapy (azathioprine, metho trexate, 6-mercaptopurine) may also play an aetiological role in the disease. [4]

Due to its association with malignancies it is also classified as a reactive dermatosis with a paraneoplastic feature. The pathogenesis of paraneoplastic autoimmunity differs from classic autoimmune diseases which are not related to cancer. Diseases associated with paraneoplastic autoimmunity often have more severe symptoms, their treatment is difficult and usually relies on treatment of the underlying malignancy. [5] Clinical variants of PG include ulcerative (most common), bullous/ atypical PG, pustular and vegetative/ superficial granulomatous pyoderma.

The lesions begin as a single or multiple painful erythematous papule, pustule, vesicle, nodule or furuncle which enlarges developing a surrounding zone of erythema. As it enlarges, the centre degenerates, erodes giving rise to ulcers . These ulcers have bluish/violaceous under-mined edges ,base is covered with purulent necrotic material and they heal with atrophic cribriform pigmented scars. Prodromal symptoms like fever, malaise, myalgia, arthralgia may be present. Extracutaneous sterile neutrophilic infiltrates may occur in the bones, lung, liver, pancreas, spleen, kidneys and central nervous system of patients with PG. [1,6]

Differential diagnosis include vasculitis, vaso-occlusive diseases, infections, other neutrophilic dermatoses like sweet's syndrome. Treatment approach involves therapeutic immunomodulation to reduce inflammation and promote wound healing. Agents like systemic glucocorticoids, cyclosporine, mycophenolate mofetil, azathioprine, methotrexate, dapsone, minocycline, Biologics (infliximab , etanercept,adalimumab,ustekinumab), thalidomide, intravenous immunoglobulin have been used in treatment of PG.[6]

Our patient had ulcerative type pyoderma gangrenosum which had developed while he was on immunosuppressants and lesions were responding to treatment very slowly. Thymolipoma is an uncommon benign thymic lesion which may also present with paraneoplastic syndromes such as myasthenia gravis, Graves' disease, pure red blood cell aplasia, aplastic anaemia, hypogammaglobulinemia, Hodgkin's disease and lichen planus. [2] This case is being reported for the rare association of thymolipoma , myasthenia gravis and pyoderma gangrenosum.

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