# Granuloma faciale in a patient with remitting seronegative symmetric synovitis with pitting edema

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## Abstract

Granuloma faciale (GF) is a rare benign chronic inflammatory dermatologic disease which is characterized by facial lesions. The diagnosis is mainly based on clinical and histopathology findings. It may be resistant to treatments and prone to relapse. Different treatment modalities include corticosteroid therapy, tacrolimus, cryotherapy and surgical methods. We report a case of GF in a patient with remitting seronegative symmetric synovitis with pitting edema (RS3PE). A male patient with RS3PE presented with reddish brown soft nodules on and over lateral aspects of his nose and adjacent areas on his face which were diagnosed histologically as GF. He was treated with prednisolone, methotrexate and clobetasol propionate cream successfully without recurrence. To the best of our knowledge this is the first case report of GF occurring in a patient with RS3PE.

## Introduction

Granuloma faciale (GF) is a rare, chronic benign inflammatory skin disease which presents as papules, plaques or nodules predominantly on face, lesions may be either asymptomatic or pruritic.[1,2] Diagnosis of GF is difficult mainly based on clinical and

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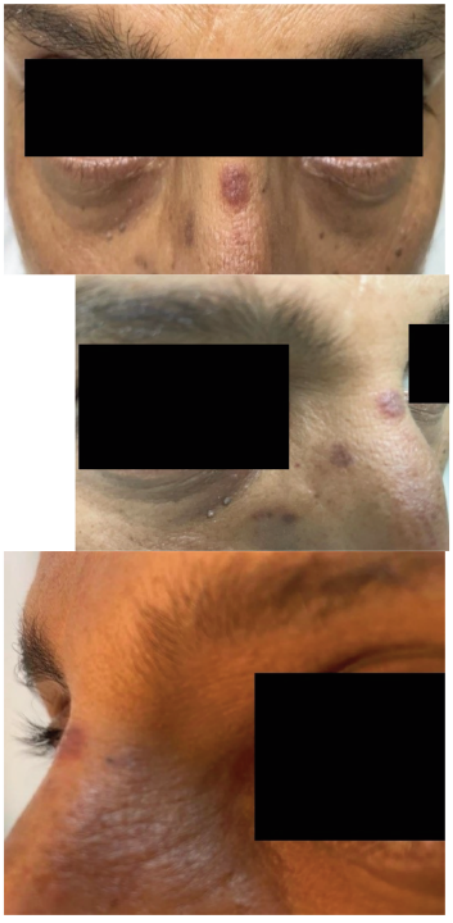

**Figure 1.** Reddish brown soft nodules on and over lateral aspects of nose and adjacent areas on face in a male patient with granuloma faciale.

histopathology findings.[2,3] It responds variably to different treatment modalities which include corticosteroid therapy, tacrolimus, cryotherapy and surgical methods.[4] We describe a case of GF in a patient with Remitting seronegative symmetric synovitis with pitting edema (RS3PE). To our knowledge, coexistence of GF and RS3PE has not been reported earlier.

## Case Report

A 46-year-old male initially presented to rheumatology clinic ten years ago with polyarthritis affecting the small and large joints of upper and lower limbs. These symptoms were accompanied by prolonged early morning stiffness (EMS) and edema of upper and

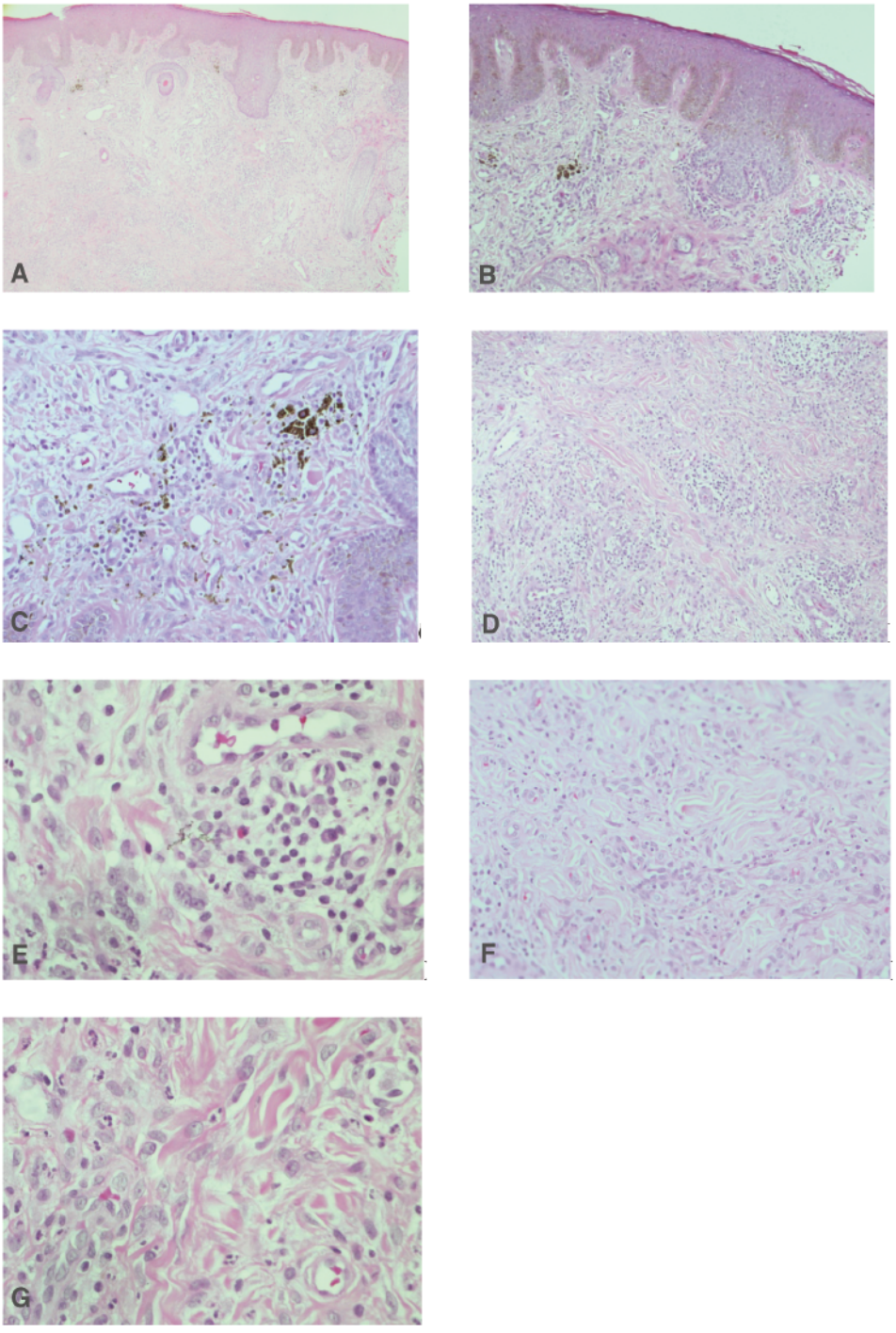


**Figure 2.** Histopathology of granuloma faciale lesion in our patient. **A**) Low power view shows diffuse inflammatory infiltrate involving the upper half of the dermis (H&E, 10x magnification); **B**) acanthosis and spongiosis (H&E, 20x magnfication); **C**) extravasated red blood cells and hemosiderin is noted (H&E, 40x); **D, E**) diffuse, polymorphous inflammatory infiltrate involving the upper half of the dermis including neutrophils, mast cells, eosinophil and lymphocytes (H&E, 40x and 100x); **F, G**) perivascular fibrosis with clefting between collagen bundles results in storiform fibrosis (H&E, 40x and 100x magnification).

lower extremities. He was unable to mobilize due to extreme pain and stiffness. His lab investigations at that time were all within normal limits; including CBC, erythrocyte sedimentation rate (ESR) was 14, rheumatoid factor (RF) was negative, C-reactive protein (CRP) was 4.930.

He was diagnosed to have remitted seronegative symmetrical synovitis with pitting edema (RS3PE). He was treated with methyl prednisolone 60 mg intravenously for three days, then started on methotrexate 15 mg orally weekly coupled with folic acid 10 mg weekly. He exhibited major improvement. He continued to take methotrexate for two years with remission of disease, then he stopped the medication on his own. He continued to feel well until two years later when he presented with reddish brownish soft nodules on and over lateral sides of his nose (Figure 1) for which he was referred to a plastic surgeon. A punch biopsy was taken from skin overlying right lateral nose for histopathology. The deep dermis showed dermal fibrosis and mixed dense inflammatory infiltrate consisting of lymphocytes, neutrophils, eosinophils, plasma cells, histiocytes, and mast cells. Blood vessels were dilated and red cell extravasation was noted (Figure 2). The histopathology results were consistent with granuloma faciale.

The plastic surgeon decided against the removal of the lesions for fear of leaving a scar. Over the succeeding weeks he developed further similar lesions over the adjacent areas. At that time his arthritis started to flare up. We started him on oral prednisolone (PSL) 10 mg daily, methotrexate (MTX) 15 mg weekly with folic acid 10 mg weekly. Once he had noticed improvement in skin and arthritis, he stopped all of his medication for one year. This was followed by relapse of the disease with joints pain, edema and stiffness and the appearance of fresh lesions on both sides of the nose. He was restarted on oral PSL 20 mg daily, MTX 15 mg oral weekly with folic acid and clobetasol propionate cream. After gaining full remission on this occasion, he once again tapered his medication. He is now 5 years after stopping all medications with no signs of new disease in skin or joint. He has no facial lesions, joint pain or EMS, no pitting edema at extremities and his laboratory findings including CBC, ESR, RF, CRP are all normal.

## Discussion and Conclusions

Granuloma faciale has an unknown etiology, but possible predisposing factors include actinic exposure, radiation, trauma, allergy, or an Arthus-like reaction. It occurs between the third and fifth decade, with chronic course and exacerbation on sun and heat exposure.[5] GF occurs as well-circumscribed papules, plaques or nodules, with varying color from red to violet on the face.[6] Our patient presented with reddish brown soft nodules on and over lateral aspects of his nose and adjacent areas on his face, which was diagnosed as granuloma faciale histologically. GF lesions are known to occur on sides of the nose (30%), tip of the nose (7%), preauricular area (22%), cheeks (22%), forehead (15%) and ear (4%).[6] Some reports have described burning and itching in the lesions, our patient did not have any of these.[4]

GF may be misdiagnosed as cutaneous sarcoidosis, discoid lupus erythematosus and lupus vulgaris. Histopathology of the lesion is required to confirm the diagnosis of GF, which is characterized by the intense infiltration of deep dermis by neutrophils, lymphocytes, histiocytes and eosinophils.[5] Erythrocyte extravasation and hemosiderin deposits are also reported. Histopathology result fulfilled the diagnosis of GF in our patient.

GF is known to be resistant to therapy and prone to relapse. Treatment includes topical, intralesional and systemic corticosteroids, cochicine, antimalarials, isoniazid, clofazimine, laser treatments, psoralen with ultraviolet A and topical tacrolimus. [4,7-12] Surgical procedures include excision with or without grafting, dermabrasion, laser argon, carbon dioxide laser, electrotherapy, pulsed dye laser and cryotherapy.[4,6] Our patient responded to prednisolone, methotrexate with folic acid and clobetasol propionate cream without recurrence during the last five years follow up. The co-existence of GF with autoimmune rheumatic diseases has only been reported in one rheumatoid (RA) patient and in another patient associated with RA and SS.[5,13] No study has reported co-existence of GF and RS3PE. To the best of our knowledge this is the first case report of GF occurring in a patient with RS3PE.

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