Case Report

A Case Report of Limited Wegener's Granulomatosis Presenting With a Chronic Scalp Ulcer

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Wegener's granulomatosis (WG) is manifested by granulomatous necrotizing inflammatory lesions involving multiple organs. Limited WG is classification of WG with the absence of disease features that pose immediate threats to either a critical individual organ or to the patient's life. The most common skin lesions are palpable purpura, necrotic ulcerations, papules and nodules with many histological pattern, leukocytoclastic vasculitis, granulomatous vasculitis, and palisading granulomas. We report a patient with a limited form of WG who presented with a chronic large scalp ulcer that rapidly responded to an immunosuppressive therapy.

Keywords: *Wegener's granulomatosis, Leukocytoclastic vasculitis, Palisading granulomas, Granulomatous vasculitis*

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Wegener's granulomatosis (WG) is one of the ANCA-associated vasculitis. The disorder is manifested by granulomatous necrotizing inflammatory lesions involving multiple organs such as the upper or lowers respiratory tracts, genitourinary tract, gastrointestinal tract, eyes and central nervous system. Two forms of WG are recognized *i.e.* a systemic, generalized severe form and a localized, limited form[1].

We report a patient with a limited form of WG who presented with a chronic large scalp ulcer that rapidly responded to an immunosuppressive therapy.

Case Report

A 28 year old man presented with a chronic scalp ulcer for 7 months. The lesion firstly appeared as a solitary nodule, 2 cm in diameter, associated with alopecia at the occipital area of the scalp. Five months later, the lesion was ulcerated to a painless ulcer with exudative discharge. He was admitted for a treatment with parenteral antibiotics and wound care. Although the infection was clear, the ulcer was still progressively enlarged to 10 cm in diameter with the development of pain and low grade fever. He was then referred to this hospital.

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He was otherwise healthy and had no history of antecedent trauma to the affected area. Physical examination at the time of admission showed that the patient had low grade fever with multiple small cervical and inguinal lymphadenopathy. A large well-defined ulcer, 10 cm in diameter, with granulation tissue base, thick indurated border and exudative discharge was found at the occipital area of the scalp (Fig. 1).

There were two small painless skin colored nodules, fixed to the surrounding tissue at the extensor area of his left forearm and left shin. Ill-defined, non-blanchable, reticulated dark bluish patches were seen on both lower legs. Other physical examination was within normal limited.

Skin biopsy was done from the edge of the ulcer and revealed dense diffuse cellular infiltration that composed of lymphocytes, histiocytes, eosinophils, together with Langhans and foreign body giant cells. Polarized light examination was negative for any foreign body. Acid fast (AFB) and Periodic acid Schiff (PAS) stains were negative for micro-organisms.

Biopsy from a small nodule on the right shin revealed lobular panniculitis with eosinophilic granuloma with vasculitis and fat necrosis (Fig. 2-4). Complete blood count showed high eosinophils (absolute eosinophil count = 672/μL). Blood chemistry, thyroid function test, urine and stool examinations, antinuclear antibodies (ANA) tests, Electrocardiogram (EKG), Chest X-ray and abdominal ultrasonography

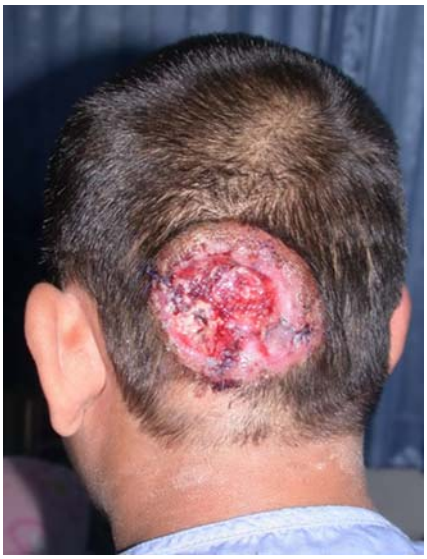

Fig. 1 a well-defined large ulcer with granulation tissue at the base with thick indurated border at the occipital area

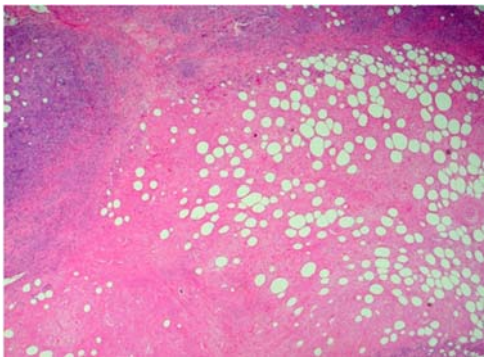

Fig. 2 revealed lobular panniculitis and necrosis of subcutaneous fat (Hematoxyline-eosin stain)

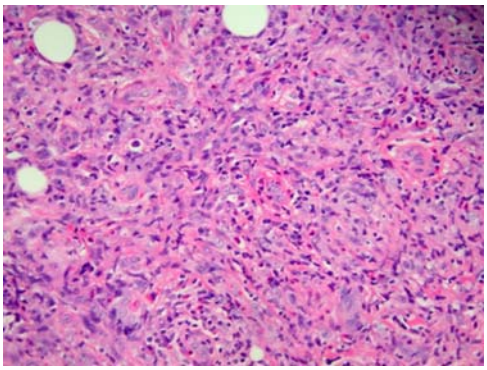

Fig. 3 numerous eosinophils infiltration (Hematoxyline-eosin stain)

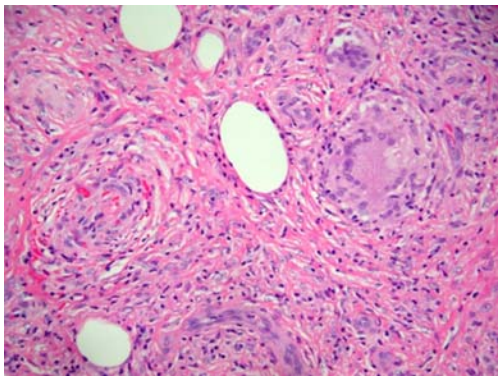

Fig. 4 Cellular infiltration in vessel walls with vessel wall destruction and fibrin deposit and giantcell (Hematoxyline-eosin stain)

were within normal limits. Pathergy test, antineutrophil cytoplasmic antibodies (c-ANCA and p-ANCA) were negative. Paranasal sinus roengenography showed a prominent right nasal turbinate. Fiberoptic laryngoscopy found mild generalized swelling of nasopharynx, turbinate and arytenoids but biopsies were all negative. Tissue fungal culture, AFB staining, mycobacterial culture, PCR for mycobacterial complex were all negative. He was diagnosed as limited form of Wegener's granulomatosis and treated with combination of prednisolone and methotrexate. The ulcer was healed with atrophic scar within 2 weeks without recurrence of new lesion within 6 months of follow-up.

Discussion

Limited WG is diagnosed by the fulfillment of the modified American College of Rheumatology criteria for the classification of WG together with the absence of disease features that pose immediate threats to either a critical individual organ or to the patient's life.[2] Criteria for the diagnosis of Wegener's granulomatosis consist of 1) nasal or oral inflammation, 2) chest x-ray showing nodules, infiltrates (fixed) or cavities, 3) microscopic hematuria or red cell casts in the urine and 4) granulomatous inflammation on biopsy (within vessel wall or perivascular)[(2)].

Patients with limited WG are more likely to be women and are significantly younger at the time that symptoms of WG began[(3)]. Skin lesions occur in 50% of diagnosed cases of WG and are a presenting sign in 10% of all patients[(3,4)]. The most common lesions are palpable purpura, necrotic ulcerations resembling pyoderma gangrenosum, papules and nodules[(4)]. The

extremities, especially legs and feet, are the most common sites. In the majority of cases, the histopathology is nonspecific and in 25 percent of cases, skin lesions demonstrate characteristic findings including leukocytoclastic vasculitis, palisading granulomas, and granulomatous vasculitis. Granulomatous inflammation is usually manifested as nodules with cutaneous and mucosal ulcerations[4,5]. Patients with extravascular granulomatous inflammation tend to have a more protracted course and more limited presentation. Moreover, these kind of lesions are more often occurred in patients with limited form[3,6].

Our patient presented with skin lesions that showed eosinophilic granuloma and vasculitis on skin biopsy with swelling of nasopharynx, turbinate and arytenoids that fulfilled two of the four criteria for the diagnosis of Wegener granulomatosis. Although the diagnosis of Wegener's granulomatosis is aided by the demonstration of antineutrophil antibodies which is a direct marker of the disease activity but negative ANCA test does not exclude the disease because they are found in only 78% of limited disease[7]. Furthermore, false-negative results are most likely occurred in limited cutaneous disease or disease remission.

The treatment of Wegener's granulomatosis consists of induction phase and maintenance phase. The first line therapy of the induction phase is systemic corticosteroid with methotexate (MTX). MTX with or without systemic corticosteroid are used in maintenance phase[1,8,9]. In a clinical trial using MTX for the treatment of fifty-two patients with limited WG, 46 of them (88.5%) achieved remission, and 67.3% of them were disease free up to 6 months. These data suggest that MTX may be a good choice for the treatment of limited WG[10].

We treated our patient initially with prednisolone 60 mg/day (1 mg/kg/day). The ulcer rapidly healed within 2 weeks. After the ulcer was healed, 10 mg weekly of methotrexate was added and prednisolone was slowly tapered off. At present, the patient receives prednisolone 5 mg every other day and methotrexate 7.5 mg/day. Plastic surgeon was consulted for the treatment of the scarring alopecia. Since the clinical course of patients with limited forms of WG is still unpredictable, our patient needs a careful lifelong follow-up[11].

Conclusion

We present the case of 28 year old man who presented with rare clinical of chronic scalp ulcer in limited Wegener's granulomatosis and he well responded to high dose prenisolone and methotrexate.

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การรายงานผู้ป่วย limited Wegener's granulomatosis นำมาด้วยอาการแผลเรื้อรังที่หนังศีรษะ

สุพิชญา ไทยวัฒน์, กอบกุล อุณหโชค

Wegener's granulomatosis เป็นโรคที่เกิดการอักเสบแบบแกรนูโลม่า ซึ่งเกี่ยวข้องกับอวัยวะหลายส่วน, Wegener's granulomatosis ชนิดเฉพาะที่ จะไม่พบอาการที่เกี่ยวกับอวัยวะสำคัญ หรือเป็นอันตรายต่อชีวิตผู้ป่วย อาการทางผิวหนังโดยส่วนมากพบเป็นลักษณะจุดเลือดออกที่คลำได้, แผลที่มีลักษณะเนื้อตาย, ตุ่มนูนเล็ก และตุ่มนูนขนาดกลางโดยพบลักษณะทางพยาธิวิทยาหลายแบบ เช่น การอักเสบของเส้นเลือดที่เกี่ยวข้องกับเม็ดเลือดขาว การอักเสบของเส้นเลือดที่เกี่ยวกับแกรนูโลม่า และการอักเสบที่มีแกรนูโลม่าล้อมรอบ เราได้รายงานผู้ป่วย Wegener's granulomatosis ชนิดเฉพาะที่อาการมีนำมาเป็นแผลขนาดใหญ่ที่ศีรษะซึ่งตอบสนองดี ต่อการรักษาด้วยยากดภูมิต้านทาน