An Unusual Case of Churg-Strauss Syndrome

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Churg-Strauss syndrome (CSS) is a very rare disease which combines the features of asthma, hypereosinophilia, multisystem necrotizing vasculitis and extravascular granuolma. To our knolwedge, there have been four reports in Korean journals, but none in dermatology journals. We report a 37-year-old male who had allergic rhinitis, pansinusitis, a history of atopic dermatitis, multiple prurigo-nodularis like skin lesions, non-fixed interstitial lung infiltration, hypereosinophilia and subclinical asthma. Clinical and histopathological features of his skin lesions were nonspecific where perivascular lymphocytic infiltration and mild lymphocytic vasculitis were shown. The presence of a pituitary mass and proximal myopathy as opposed to peripheral neuropathy have not been reported in previous reports of CSS. However, other features of our patient met the criteria of CSS.

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Key Words : Churg-Strauss syndrome, Subclinical asthma

Churg-Strauss syndrome(CSS), allergic granulomatosis and angiitis were first reported as a disease entity separate from polyarteritis nodosa in 1951 by Churg and Strauss[1]. About 40% had cutaneous findings and 6% had skin lesions as the initial manifestation in the previous reports. The most frequent cutaneous findings were erythema multiforme like lesions, purpura and cutaneous papulonodules on the extremities[2-5]. The diagnostic criteria are asthma, peripheral eosinophilia more than 10%, peripheral neuropathy, non-fixed lung infiltration on chest X ray, sinusitis and eosinophilic infiltration on biopsy of various organs. More than four must exist[6]. We describe one patient with CSS whose skin lesion features were nonspecific clinically and histopathologically. In addition, there was no apparent asthma or nuropathy, which made diagnostic difficult. However, a rapid response to systemic steroids was noteworthy and confirmed the diagnosis.

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CASE REPORT

A 37-year-old man presented with malaise, intermittent fever, arthralgia, impotence, severe generalized itching and proximal muscle weakness of the lower extremities. These symptoms had been present for 8 months and he had had a dry cough for 2 months. The severity of the symptoms fluctuated without specific treatment. He was referred to our department for the generalized itchy papular lesions, axillary and pubic hair loss. Two months previously, he was diagnosed as having toxic hepatitis, diabetes insipidus and panhypopituitarism at another hospital. His past medical history included chronic pansinusitis, allergic rhinits with generalized itching for 15 years. Skin signs included dermographism and generalized multiple relatively ill defined erythematous to brownish indurated papuloplaques and crusted papules (Fig 1 A,B,C). Laboratory investigations demonstrated peripheral eosinophilia(30%), hyper-IgE, hyper-IgA, mild leukocytosis, anemia with a raised sediment rate. Negative cANCA, pANCA, antinuclear antibodies, and the rheumatoid factor were demonstrated. The CK/LD level was within normal limits. No diagnostic results were revealed by test for the para-

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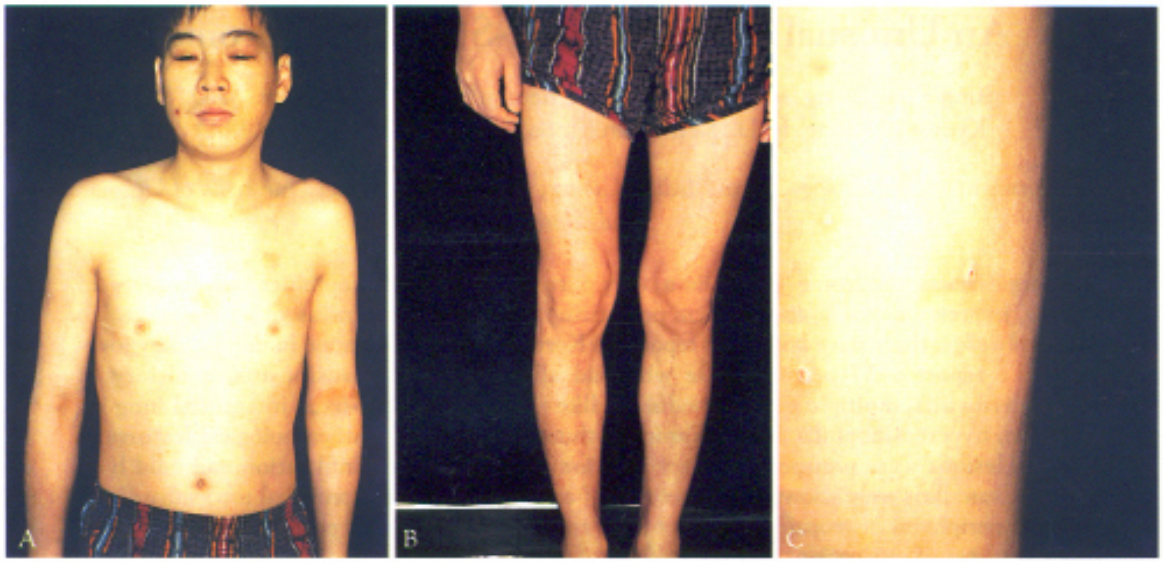


Fig. 1. A, B, C. Generalized multiple erythematous to brownish indurated papuloplaques and crusted brown to skin colored papules.

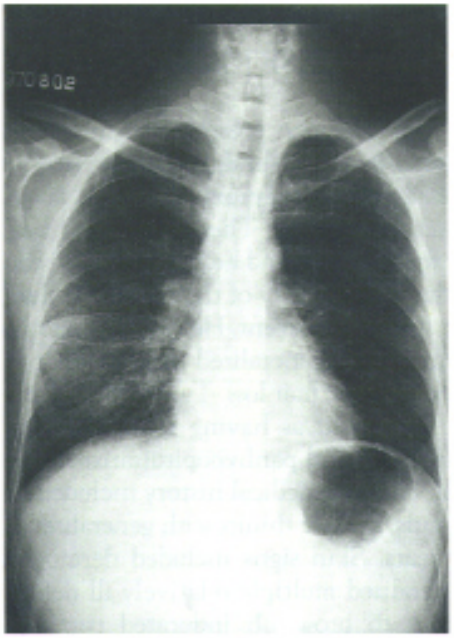


Fig. 2. Chest X ray revealed a linear reticular shadow in the right lower lobe of the lung.

site-specific Ig G antibody. A radiological examination revealed a linear reticular shadow in the right lower lobe of the lung (Fig 2). The infiltration of the lung was migratory and rapidly changing. He was also diagnosed as having bronchial asthma by a provocation test. A brain CT revealed a pituitary mass (Fig 3) which could explain his diabetes insipidus and panhypopituitarism. Electromyography was consistent with myopathy and nerve conduction velocity was within normal limits without polyneuropathy. A lung biopsy showed mostly eosinophilic infiltration around the bronchioles (Fig 4). His general symptoms and lung infiltration were slightly improved without any treatment 7 days after admission, when we carried out the skin biopsies. Multiple biopsies from the itchy nodular and papular lesions showed acanthotic epidermis, papillary dermal thickening with dense superficial and deep lymphoplasma cell infiltration with some eosinophils (Fig 5). Neither extravascular granuloma nor leukocytoclastic vasculitis was detected from the multiple skin biopsies.

Soon his condition was aggravated again and he received a high dose steroid administration (prednisolone 1 mg/kg with acute tapering) with thyroid and testosterone replacement. Peripheral eosinophilia was also decreased and he went into a long-lasting remission one month after starting steroid therapy.

DISCUSSION

Churg-Strauss syndrome(CSS) is a very rare dis-

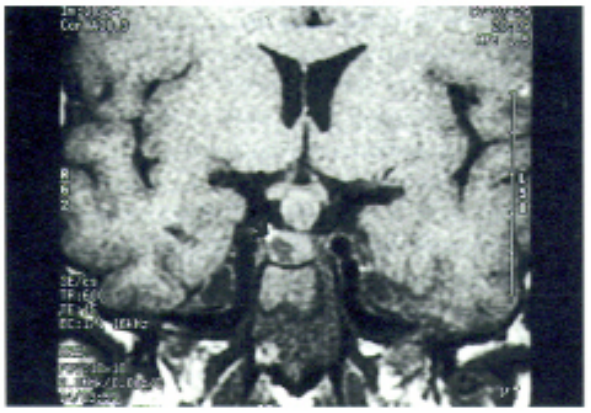


Fig. 3. Brain CT revealed a mass of pituitary gland.

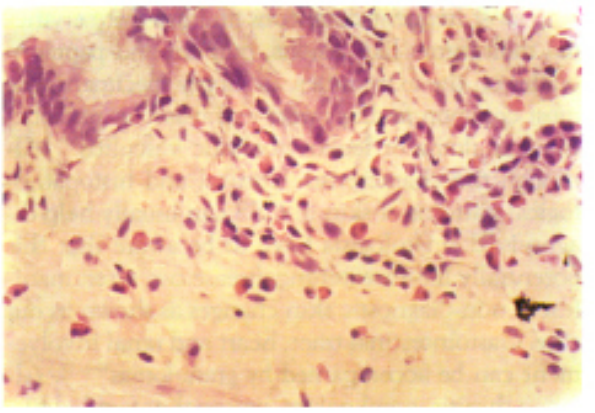
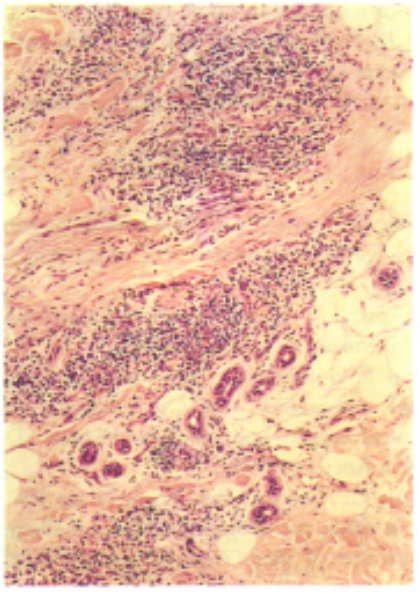


Fig. 4. Bronchoscopic biopsy showed mostly eosinophilic infiltration around the bronchioles (H&E, × 200).



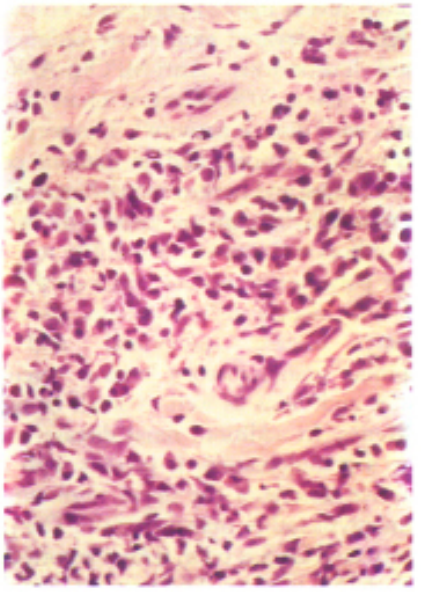


Fig. 5. A, B. Dense perivascular lymphoplasma cell infiltration with some eosinophils (A : H&E, ×40, B : H&E, × 200).

tinct disease which combines the features of bronchial asthma, transient lung infiltration, hypereosinophilia, multisystem necrotizing vasculitis and extravascular granuloma. Most patients are adults in the third and fourth decade and there is a slight male predominance[1-5]. The diagnostic criteria are that more than four of the following must be present : asthma, peripheral eosinophilia, neuropathy, nonfixed lung infiltration, sinusitis and eosinophilic infiltration on biopsy[6]. Asthma and necrotizing vasculitis are invariably present[2-6]. Previous investigations revealed that the symptoms of asthma usually preceded the occurrence of CSS by many years and asthmatic symptoms seemed to be severe at the onset of the CSS and progressed thereafter[2-4]. However, the occurrence of CSS is not necessarily correlated with symptoms of asthma as demonstrated by other reported cases including this case[5]. Pulmonary involvement usually showed transient patchy or nodular infiltration[7]. About 40% of CSS had cutaneous findings and 6% had skin lesions as the initial manifestation[5-9,8].

The most frequent cutaneous findings were erythema multiforme like lesions, purpura and papulonodules on the extremities. In biopsy specimens, the most common findings were extravascular necrotizing granuloma or leukocytoclastic vasculitis[8,9]. Although the characteristic clinical and histopathological patterns of skin in CSS can usually help diagnosis, dermatologists have to be aware that this potentially life-threatening disease can often manifest as non-diagnostic skin changes as shown in our case. Gastointestinal tract, heart and renal involvement can be accompanied by granulomas[10-12,19]. Allergic rhinitis, nasal polyps, personal or familial history of atopy, allergic reactions to inhalants, hypertension, arthralgia, arthritis, myalgia, fever, and abdominal pain can exist[8-12]. Laboratory investigations often reveal marked peripheral eosinophilia, hyper-IgE, an elevated erythrocyte sedimentation rate and leukocytosis. Anti-MPO(p-ANCA) is positive in about 70% of active phase cases and correlates with disease activity[13]. The pathogenesis of CSS is still unknown and an immune complex mediated disease and hyperresponsiveness of granulocytes to anaphylatoxins are suspected[14]. Although several authors still consider this entity to be a subgroup of polyarteritis nodosa, distinguishing featuers of CSS include asthma, allergic phenomena, pulmonary disease, peripheral and tissue eosinophilia, granulomatous lesions, vasculitis of capillaries and veins as well as small or medium-sized arteries. In addition, renal disease is rare[3,15]. Wegener's granulomatosis is present with ulceroproliferative lesions of the upper respiratory tract and hemoptysis rather than asthma[3,15]. The prognosis of untreated CSS is poor, with a reported 5-year survival of 25% of cases. The cause of death is pulmonary and cardiac disease[11,16]. Early and appropriate diagnosis is crucial for starting appropriate and, often, life-saving therapeutic measures. Gluococorticoid therapy increases the 5-year survival rate by more than 50%[16-21]. A combined regimen of cyclophosphami-de and alternate-day prednisone can be used[17]. Prednisone plus cyclophosphamide plus plasma exchanges as first-line treatment can be used for severe CSS[18]. A monthly bolus of cyclophosphamide or weekly low-dose methotrexate are being pursued with less toxicities[16]. The presenting case showed a dramatic response to high-dose prednisone without notable toxicities. In this case, CSS was very difficult to diagnose because asthma was detected only by a later provocation test and histopathological results of the skin and lung did not aid the diagnosis. Our case fulfilled the criteria although there were neither typical extravascular granuloma nor leukocytoclastic vasculitis in the skin and lung biopsies. Considering that CSS may have a spontaneously fluctuating course, the nonspecific findings without definite necrotizing vasculitis might reflect the improved state. Another interesting feature of this case was the combined pituitary mass. There have been many reports with peripheral neuropathy in CSS but no reports of a lesion of the central nervous system. Furthermore, our case had myopathy not peripheral neuropahty. We emphasize that diagnosing CSS can be very difficult with such nonspecific skin manifestations and the lack of non-granulomatous lesions in both the skin and lung. However, this rare disease can be treated effectively and dermatologists need to be reminded of its existence.

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