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Churg-Strauss Syndrome : A case report

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รายงานผู้ป่วยกลุ่มอาการ Churg-Strauss

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ภาควิชาอายุรศาสตร์ คณะแพทยศาสตร์ มหาวิทยาลัยขอนแก่น

รายงานผู้ป่วย 1 ราย เป็นผู้ป่วยชายอายุ 29 ปี มาโรงพยาบาลศรีนครินทร์ ด้วยอาการไอเรื้อรัง มีอาการอ่อนแรงและปวดบริเวณแขนและขาทั้ง 2 ข้างมาประมาณ 3 เดือน ร่วมกับมีอาการปวดตามข้อต่าง ๆ และมีตุ่มกดเจ็บใต้ผิวหนังขึ้นทั่วร่างกาย ผลการตรวจร่างกายและการตรวจวินิจฉัยทางห้องปฏิบัติการ เข้าได้กับกลุ่มอาการ Churg-Strauss ซึ่งเป็นกลุ่มอาการที่พบได้น้อยมาก และยังไม่เคยมีรายงานมาก่อนในประเทศไทย

A case of 29 year-old male presented with history of chronic cough, muscle pain, arthralgia, progressive weakness of upper and lower extremities and palpable tender small cutaneous nodules for three months. Physical examination and all of laboratory investigations were compatible with Churg-Strauss syndrome, which is a rare disease and never been reported in Thailand.

INTRODUCTION

Churg-Strauss syndrome (CSS) or allergic granulomatosis is the one of three systemic diseases in which a necrotizing vasculitis is seen in association with neerotizing granuloma.[1] This syndrome was recognized as an entity seperated from periarteritis nodosa in 1951.[2] Clues in the diagnosis of CSS are[1-10](1) respiratory symptoms, usually asthma or allergic rhinitis (2) pronounced eosinophilia both in circulating blood and in the lesions (3) the presence of pulmonary infiltration (4) involvement of arteries and veins (5) the pressence of palisading granuloma with central necrosis extravascularly in the connective tissue

CASE REPORT

A 29 year-old male patient came to Srinagarind Hospital with the history of pain

and weakness of both upper and lower extremities 3 months before admission. He noted that the weakness began at both feet and gradually increased in severity. One month later he developed arthralgia and palpable discrete small tender cutaneous nodules over his trunk and extremities. He had the past history of allergic rhinitis and chronic cough for 10 years, and he also had fever and progressive weight loss about 10 kg. in these 3 months.

On physical exammination, he was febrile, tachypnea and tachycardia, markedly thin; and small discrete tender cutaneous nodules size 3-5 mm. was palpable over his trunk and extremities. Bilateral ulnar, peroneal and superficial cervical nerves were enlarged. Both lungs had coarse crepitation. Neurological exammination showed right wrist drop, left foot drop, muscles power grade II-III and loss of pinprick sensation in gloves and stockings pattern at both upper and lower extremities.

Laboratory findings showed mild anaemia (Hct 26 vol%) peripheral blood eosinophilia (Eo 30%) ESR 54 mm/hr, positive rheumatoid factor, positive HBsAg, chest x-ray showed infected bronchiectasis of right lower lung field. Renal angiogram was normal. Other laboratory investigations including antinuclear factor, VDRL, B_1C, were normal. IgE level was not done because the laboratory was not available.

Histopathological studies of muscle, superficial cervical and sural nerves showed focal hyalinization of wall of medium and small sized blood vessles with cell infiltration around and within blood vessles wall, composed of neutrophils, mononuclear cells, eosinophils and nuclear dust.

The patient was treated with prednisolone 60 mg/d and systemic antibiotics for infected bronchiectasis. Clinical improvement was noted within 2 weeks. The last follow up, one and a half year later, he had got well, no wrist drop, no foot drop and the cutaneous nodules was not noted.

DISCUSSION

The diagnosis of Churg-Strauss syndrome in this patient is based on clinical clues of (1) history of allergic rhinitis but not asthma (2) peripheral blood eosinophilia and eosinophilic infiltration also noted in the lesions and (3) necrotizing vasculitis of small and medium sized arteries. Chumbley et al.[3] had noted that history of asthma or allergic rhinitis may precede the development of systemic vasculitis by up to 30 years. Occasionally, there is a dramatic remission of asthma with the onset of vasculitis but in some cases the vasculitic illness coincide with an exacerbation of asthma. Several cases in the literature have all the features of CSS, but instead of asthma, chronic bronchitis, bronchiectasis were recorded.[3,7] Each of the major histological features of CSS (tissue infiltration by eosinophils, extravascular granulomas and necrotizing vascultitis) may occur in isolation, or may coexist. Granulomas are most commonly observed in the heart, liver, spleen and lungs.[7,10] They may, therefore, be missed by tissue biopsy. HBsAg are commonly associated with periarteritis nodosa but not with CSS.[11-14] In this patient, the presence of HBsAg may be explained by the high incidence of carrier rate of HBsAg in Thai population.

CSS responds well to corticosteroids,[15] as in this patient. Five-year survival rate has been increased from 4% in presteroid era to 62%, and median survival is 9 years. Some cases of CSS require immunosuppressive agents, as adjuncts to steroid treatment. Cooper et al had described two cases who responsed to steroid and azathioprine therapy.[10] Cyclophosphamide and chlorambucil have also been used successfully.[16]

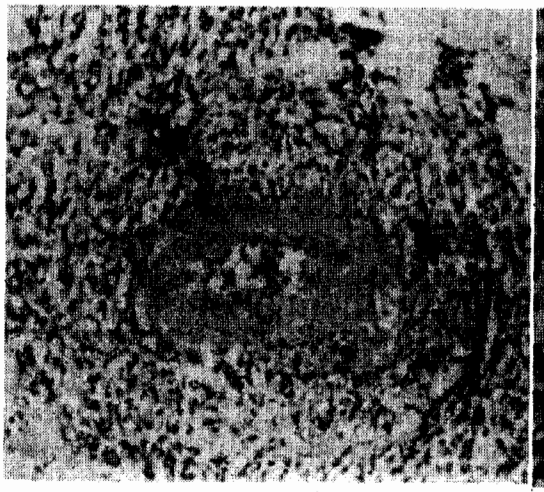

Figure 1 Histology of muscle showed hyalinization of wall of medium sized artery with cells infiltration around and within blood vessle wall (H & E × 50)

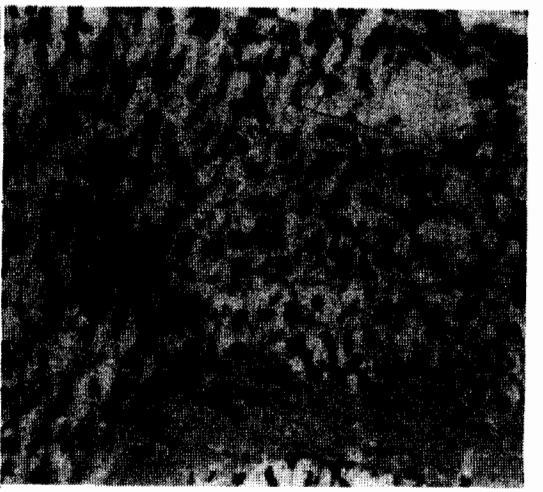

Figure 2 Cells infiltration composed of eosinophils, monomuclear cells and neutrophils (H & E × 200)

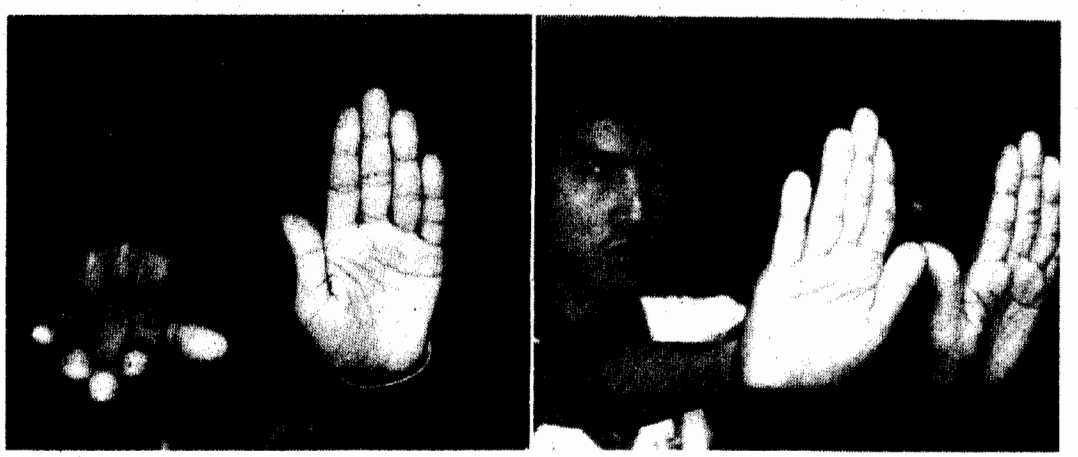

Figure 3 Demonstrate right wrist drop before treatment (a), and recovery one and a half years after treatment (b).

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