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Horner's Syndrome in Wegener's Granulomatosis: A Case with Pulmonary Involvement

Canan Eren Dağlı[1], Mehmet Sayarlıoğlu[2], Murat Özdemir[3], Hayriye Sayarlıoğlu[4], Nurhan Köksal[1]

[1]Kahramanmaras Sutcu Imam University Chest Diseases, Kahramanmaras, Turkey
[2]Kahramanmaras Sutcu Imam University Rheumatology, Kahramanmaras, Turkey
[3]Kahramanmaras Sutcu Imam University Ophtalmology, Kahramanmaras, Turkey
[4]Kahramanmaras Sutcu Imam University Nephrology, Kahramanmaras, Turkey

Abstract
Wegener's granulomatosis(WG) is an uncommon inflammatory which rarely is reported to cause Horner's syndrome. We report a case of HS in a patient with WG caused by pulmonary involvement. A 47 year-old man presented with cough, fever, and chest pain. On exam, he was noted to have ptosis and miosis on the left eye. Chest X-ray and thorax computed tomographic scan of the chest revealed a cavitary lesion in the apicoposterior segment of the left upper lobe. HS caused by pulmonary manifestation of WG is rare and such a disease process needs to be considered in the differential diagnosis.

Key words: Horner's syndrome, pulmonary lesion, Wegener's granulomatosis

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INTRODUCTION

WG is a systemic inflammatory disease characterized by necrotizing granulomatous inflammation of the upper and lower respiratory tract, glomerulonephritis, and systemic vasculitis. It is a systemic vasculitis of the small and medium sized arteries and veins and is a relatively rare disease of unknown etiology. Ocular disease is the presenting manifestation in 8-16% of patients, and proptosis, ophthalmoplegia, dacryoadenitis and nasolacrymal duct obstruction, and sclerokeratitis are common [1-3]. In previous reports, HS was presented in several WG patients with generally central etiology [4-6]. We report a very rare case with HS caused by pulmonary manifestation of WG.

CASE REPORT

A 47 year-old man presented with symptoms of fall down of left eyelid, cough, fever, and chest pain in January 2007. Five years ago, he underwent pneumonectomy and was diagnosed as WG with histological biopsy and C-ANCA positivity in another medical center. He was treated with endoxan and prednisolon for one year when he was admitted to our hospital.

Radiologic examination including chest X-ray and thorax computed tomography revealed bilateral, multiple cystic and cavitary nodular lesions. The greatest of them (7*6.5*7.5 cm) located in the apicoposterior segment of the upper lobe of left lung (Figure 2). On ophthalmic examination, a mild ptosis and miosis were detected in the left eye (Figure 1). Any other ophthalmic manifestations of WG were not detected. Adrenaline 1:1000 test showing postganglionic involvement of sympathetic route was positive in the left eye. In this test, adrenaline 1:1000 was instilled into both eye. In a preganglionic lesion, no pupil dilates but the Horner pupil dilates and ptosis may be temporarily relieved [3]. In our patient, we thought that HS was caused by the lesion in the apicoposterior segment of the upper lobe of the left lung.

DISCUSSION

American College of Rheumatology describes a patient with WG if two or more of the following criteria are present: [1] nasal or oral inflammation, [2] abnormal chest radiograph, [3] abnormal urine sediment, or [4] granulomatous inflammation on biopsy. The sensitivity of this classification system is 88% [1]. In addition, c-ANCA has 85-96 % sensitivity in WG [1]. Our case was diagnosed previously as WG with positive c-ANCA-test and specific histopathology findings in another medical center.

In January 2007, our case was admitted with symptoms of HS. The positive adrenalin 1/1000 test showed postganglionic etiology. Therefore, we thought that the lesion in the upper lobe of the left lung compressed on the servical sympathetic route and caused HS.

In WG, ocular disease is the presenting manifestation in 8-16% of patients, and eventually develops in 28-87 %. Necrotizing sclerokeratitis and proptosis caused by orbital inflammation are most commonly seen [1,2]. Reported neuro-ophthalmologic manifestations include palsies of the se-

Corresponding Author: Dr. Canan Eren Dağlı, Kahramanmaras Sutcu Imam University, Chest diseases, Kahramanmaras, Turkey Phone: +90 344 221 23 37 E-mail: cananerdagli@yahoo.com

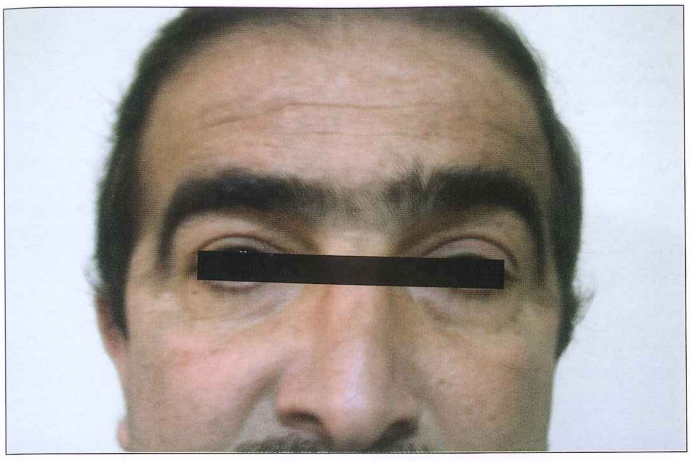

Figure 1. The patient has miosis and ptosis

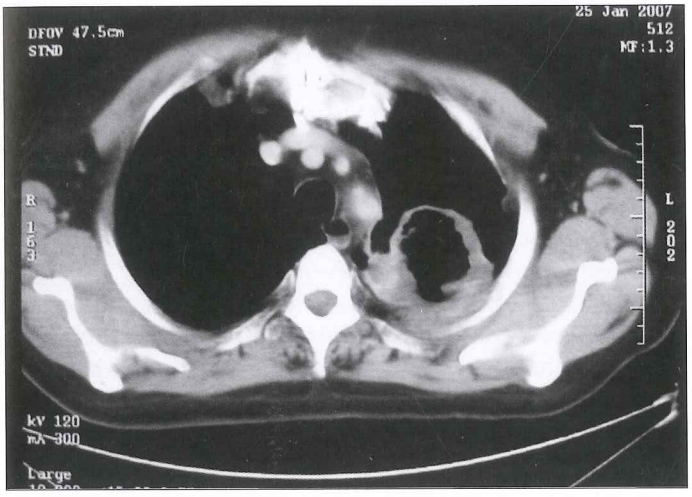

Figure 2. 7.5x7x6.5 cm. mass on the left upper lob on thorax CT

cond, third, fourth, and sixth cranial nerves [4]. HS as manifestation of WG is extremely rare: a few cases are reported [4-6]. All of these cases had central etiology. Four patients with active WG had HS, which was the only neurological involvement in one. Two others had multiple mononeuropathy and one a multiple cranial neuropathy. HS occurred in the active stage and improved with treatment of WG. In our case, there was not central nervous system involvement and there was no other ocular or orbital pathology.

Our patient is a very rare case with HS caused by pulmonary involvement of WG without central nervous system etiology. Therefore, our case illustrates the need to consider WG in the differential diagnosis of HS and we want to emphasize that HS may develop due to pulmonary involvement in WG patients.

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