Chronic meningitis with multiple cranial neuropathies: A rare initial presentation of Wegener's granulomatosis

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Abstract

Wegener's granulomatosis (WG) is a systemic necrotizing vasculitis that affects the small blood vessels. It mainly affects the upper and lower respiratory tract and kidneys. Central nervous system (CNS) involvement is rare, and has been reported only in about 8% of cases during the course of illness. Initial presentation with neurologic affection, particularly chronic hypertrophic meningitis is very unusual. We report the case of a 34 year old male who presented with chronic hypertrophic meningitis and multiple cranial nerve involvement as the initial manifestation, without respiratory and renal symptoms. This case highlights the difficulties in diagnosing a rare disease with rarer presentation, and at the same time illustrates that Wegener's granulomatosis should be considered in the differential diagnosis of chronic meningitis.

Key Words

Chronic meningitis, cranial neuropathies, Wegener's granulomatosis

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Introduction

Wegener's granulomatosis (WG) is a systemic necrotizing vasculitis that mainly affects the upper and lower respiratory tract and kidneys.[1] Central nervous system (CNS) involvement is rare, and has been reported only in about 8% of cases during the course of illness.[2] Initial presentation with CNS affection, particularly chronic hypertrophic meningitis is even rarer. Very few cases of Wegener's granulomatosis have been reported with such initial presentation.

Case Report

A 34 year old male presented to our department with two months duration of headache, right earache, and vomiting. Two weeks later, he developed right facial numbness, inability to close the right eye, deviation of the mouth to left, loss of taste sensation in the right half of tongue, slurring of speech with nasal intonation, and difficulty in swallowing with nasal regurgitation of liquids. There was no history of weakness or sensory impairment in limbs, blurring of vision, hearing impairment, cough, fever or loss of appetite. There was no history of ear discharge, joint pains or rashes. He had a history of recurrent mild sinusitis since last 10 years; however, it was never serious enough to warrant medical intervention. He had no history of hypertension, diabetes, tuberculosis, or any other chronic illness.

His vitals and general physical examination were normal. Fundus examination did not reveal any abnormality. He had right 5th and 7th, and bilateral 9th, 10th cranial nerve palsies. There were no signs of meningeal irritation. Rest of the neurological examination was unremarkable. Systemic examination, including respiratory system was normal. He had undergone evaluation elsewhere prior to admission with us. His hemogram, fasting blood sugar, renal functions, liver functions and serum electrolytes were within the normal limits. Chest radiograph did not reveal any abnormality. Cerebrospinal fluid (CSF) analysis showed lymphocytic pleocytosis, with normal sugar and proteins. Magnetic resonance imaging (MRI) brain showed an inflammatory duromeningeal process with asymmetric and prominent enhancement in the right hemisphere suggestive of hypertrophic pachymeningitis [Figure 1]. He was treated with anti tubercular therapy (ATT) and oral steroids; however, there was no improvement in the response.

After hospitalization, we re-evaluated him to diagnose an alternative etiology. His erythrocyte sedimentation rate (ESR) was 50 mm/h (normal- 0-15 mm/h). The test results for antinuclear antibodies (ANA), Human immunodeficiency virus (HIV), hepatitis B surface antigen (HBsAg), Venereal Disease Research Laboratory (VDRL) and brucella serology

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were negative. Serum Angiotensin Converting Enzyme (ACE) level was within normal limit. Urine examination revealed microscopic hematuria with mild proteinuria. Repeat CSF analysis showed lymphocytic pleocytosis with moderately raised proteins and no atypical (malignant) cells were found. Bacterial antigen test, gram staining, Acid-fast bacilli (AFB) staining, India ink preparation and culture for bacteria and fungus were negative. CSF tuberculosis polymerase chain reaction (TB PCR)and VDRL test were negative. CSF ACE level was normal.

Computed Tomography (CT) thorax was planned to look for possible evidence of pulmonary tuberculosis and sarcoidosis. It showed multiple cavitating nodular opacities in the right lung field [Figure 2]. The otolaryngologist assessed him for earache, and advised CT scan of the temporal bones and paranasal sinuses, which then revealed bilateral mastoiditis and chronic rhino-sinusitis [Figures 3 and 4]. With these radiologic findings, a strong possibility of WG was suspected, which was further confirmed by positive Cytoplasmic antineutrophil cytoplasmic antibodies (c-ANCA) staining and nasopharyngeal biopsy which revealed an ulcer with chronic granulomatous inflammation [Figure 5]. He was put on oral cyclophosphamide, and the dosage of prednisolone was increased. Headache, vomiting, and earache disappeared within days. On follow up, the patient has displayed significant clinical improvement.

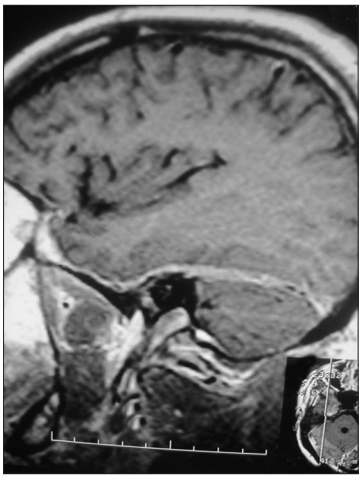

Figure 1: Magnetic resonance imaging brain of the patient in the study showing dural thickening and abnormal enhancement

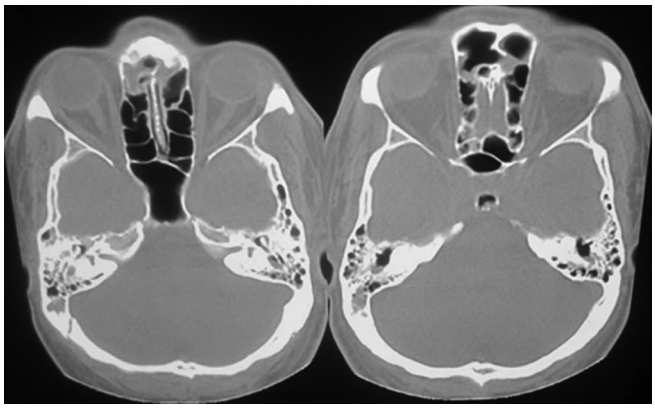

Figure 3: Non Contrast Computed Tomography scan of the temporal bone of the patient in the study showing bilateral opaque mastoid air cells and tympanic cavities

Discussion

WG is a rare disease. It was initially described as a granulomatous affection of the upper and lower respiratory tract and kidney. Currently it is recognized as a necrotising, small vessel vasculitis that can involve nearly every organ or tissue in the body.[3] More than 90% of the patients initially present with upper or lower airway symptoms or both.[2] Neurological involvement during the course of the disease is usually in the form of mononeuritis multiplex and cranial neuropathy. Headache, cerebritis, seizures, stroke, meningitis, diabetes insipidus, hydrocephalus, external ophthalmoplegia, myelopathy, and myopathy are other manifestations which are seen less frequently.[4] CNS involvement usually occurs during the later stages of the disease,[3] and is seen in about 8% of the cases. Three mechanisms are involved in the

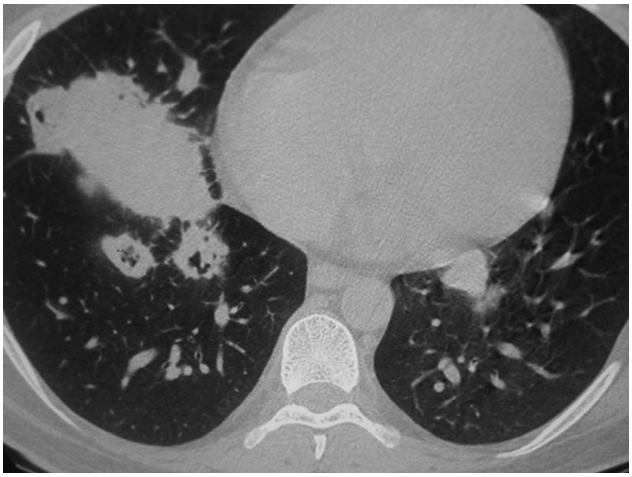

Figure 2: Computed Tomography scan of the thorax of the patient in the study showing multiple cavitating nodular opacities in right lung field

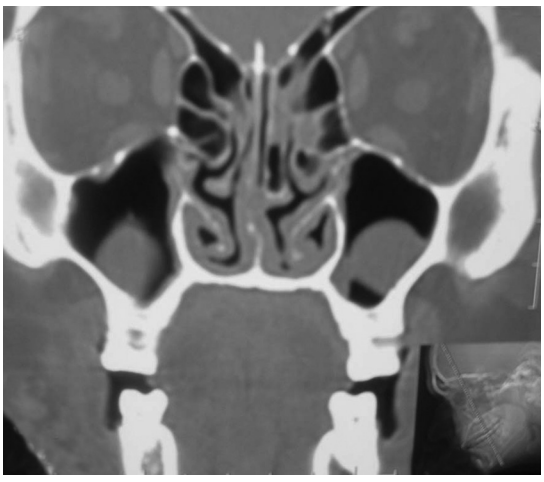

Figure 4: Non Contrast Computed Tomographyscan the paranasal sinuses of the patient in the study showing chronic rhino-sinusitis

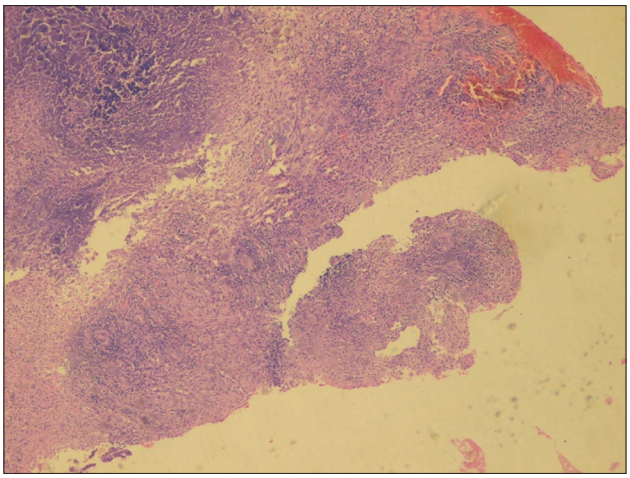

Figure 5: Nasopharyngeal biopsy of the patient in the study showing chronic granulomatous inflammation

pathogenesis of the different CNS manifestations seen in WG: a) vasculitis of the nervous system; b) contiguous invasion of granuloma from extracranial sites; c) and, remote intracranial granuloma.[4] Our patient presented with chronic hypertrophic pachymeningitis with multiple cranial neuropathies without any classical symptoms of WG. This unusual presentation delayed the diagnosis. He was initially given ATT considering the possibility of neurotuberculosis, which is endemic in our country and may present with similar features.[5] However, in absence of appropriate response he was investigated further, and eventually the diagnosis of WG was established. Meningeal involvement at any stage of disease is rare (1%).[2,6] In a large case series of 324 patients of WG, meningitis was found only in 2 patients.[6] In another series of 158 patients, none of the patients was found to have meningitis.[2]

Untreated WG universally has a fatal course. Treatment with a combination of glucocorticoids and cyclophosphamide leads to a marked symptomatic improvement in more than 90% of patients, and 75% of patients achieve complete remission.[1] We also found excellent response to this combination treatment in our patient. In WG with chronic meningitis, an early diagnosis and proper treatment may prevent irreversible cranial nerve dysfunction and other sequelae, like communicating hydrocephalus.[4]

Conclusion

Chronic meningitis with multiple cranial neuropathies may be the initial presenting feature of Wegener's granulomatosis. Such presentation is rare and needs a high degree of suspicion as early diagnosis and treatment can prevent morbidity and mortality.

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