Granulomatosis with polyangiitis

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Abstract

Granulomatosis with polyangiitis is a small and medium vessel vasculitis. It may affect any organ system. Neurological involvement, though less frequent, mostly manifests as peripheral neuropathy. Central nervous system (CNS) involvement as autoimmune cerebritis is rare but may be devastating. We report a patient having CNS involvement in the form of Parkinsonism long before the development of typical upper and lower airway symptoms. Granulomatosis with polyangiitis was diagnosed on the basis of cavitary lung lesions along with epistaxis, granulomatous inflammation from fine needle aspiration (FNAC) of lymph node and high titer of c-ANCA. The patient was commenced on intravenous methylprednisolone followed by mycophenolate mofetil and responded to treatment. Autoimmune cerebritis may present with subacute manifestation of diffuse CNS involvement.

Introduction

Granulomatosis with polyangiitis, formerly referred as Wegener's granulomatosis, is a distinct clinicopathologic entity characterized by granulomatous vasculitis of the upper and lower respiratory tracts together with glomerulonephritis.[1] It is a rare multisystem autoimmune disease of unknown etiology. Its hallmark features include necrotizing granulomatous inflammation and pauci-immune vasculitis in small and medium sized blood vessels.[2]

It is strongly associated with cytoplasmic anti-neutrophil cytoplasmic antibodies (c-ANCA). This is a complex and potentially serious disease. But with prompt diagnosis, granulomatosis with polyangiitis can be treated effectively. It can affect any organ, though mostly involving the kidneys and respiratory tract. Neurological manifestations are less common. This case report details a patient whose earliest symptom was Parkinsonism, with later development of respiratory manifestations.

Case Report

A 38 year old Bangladeshi gentleman, father of one child, presented with resting tremor of both hands developing over one year, fever, nose bleeding and nasal crusting for 3 months. Initially, tremor was unilateral, later became bilateral. But he had no gait abnormality or rigidity. For this symptom he visited neurologists several times and was on levodopa-carbidopa for 6 months. His symptoms did not improve. Thereafter, for the last 3 months, he developed intermittent high grade fever, epistaxis and nasal crusting. Gradually respiratory distress developed over the month before admission. He had no history of bronchial asthma, atopy or any past cerebrovascular disease. Upon admission, clinical examination demonstrated marked anemia, bilateral lacrimal gland enlargement, injected conjunctiva with serous discharge from eyes. Crusting with blood stained discharge was present in both nostrils. Patient was febrile with temperature 103°F, SPO_2 was 90% in room air. Bilateral enlarged submandibular lymph nodes were present (right one 3 × 3 cm, left one 2.5 × 2 cm), both were firm, non-tender and mobile. Chest auscultation revealed prolonged vesicular expiration with wheeze and bilateral diffuse inspiratory crackles. Patient had resting tremor of both hands with cogwheel rigidity and positive glabellar tap.

After the chest radiographic findings of multiple cavitary lesions along with epistaxis, nasal crusting and FNAC of lymph node showing granulomatous inflammation, provisional diagnosis of granulomatosis with polyangiitis was suggested. Strongly positive c-ANCA (45 U/L) and granulomatous inflammation on FNAC of lymph node confirmed granulomatosis with polyangiitis. Pulmonary tuberculosis was excluded on the basis of negative Mantoux test, negative QuantiFERON-TB gold test and sputum negative for TB bacilli for successive 3 days. Our final diagnosis was

Table I

Investigation parameters

Investigation	Results
Complete blood count	6.6 g/dL, MCV -78.7 fL, MCH – 24.3, TC – 16,700/µL, platelet – 6,93,000/µL
Erythrocyte sedimentation rate	>140 mm
C-reactive protein	342 mg/dL
Urine routine examination	Albumin - present; RBS – 3-6/HPF; Pus cells – 5-10/HPF
S. creatinine	0.7 mg/dL
Peripheral blood film	Mild anisochromia and anisocytosis with rouleaux formation, anemia of chronic disease
Serum alanine aminotransferase	66 U/L
Urine for dimorphic RBC	Not found
Sputum for C/S	No growth
FNAC from submandibular lymph nodes	Right: Suppurative granulomatous inflammation Left: Suppurative inflammation

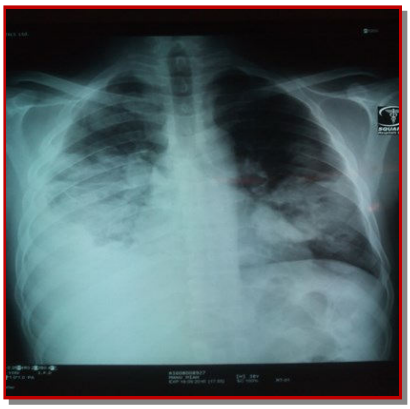

Figure 1: XRay chest P/A view showing multiple bilateral cavitary lesions with right basal consolidation with effusion

granulomatosis with poly-angiitis with Parkinsonism due to cerebritis and pneumonia. An MRI of brain was done prior to admi-ssion in our hospital which was unremarkable.

Patient received two units of packed red blood cells. Intravenous methylprednisolone was commenced immediately, for 3 days followed by oral prednisolone 1 mg/kg body weight with plan for slow tapering. Intravenous ceftriaxone and clindamycin were started to cover secondary bacterial infection. Respiratory distress improved with sulbutamol and steroid nebulization. Sulphamethoxazole and trimethoprim combination was given for pneumocystis jiroveci prophylaxis and isoniazide 150 mg for TB prophylaxis. He was also started oral mycophenolate mofetil. For remission induction we did not choose cyclophosphamide as patient's wish for future offspring. Rituximab was planned for future flare if any. Gradually patient became afebrile and his generalized well being was improved. For tremor, we continued levodopa-carbidopa and trihexyphenidyl hydrochoride.

Discussion

Most commonly granulomatosis with polyangiitis affects the sinuses, lungs and kidneys, but can also affect the eyes, skin, nerves (both peripheral and central). But it can affect almost any organs of body. Therefore, a wide range of symptoms may develop.

For 90% of people, the first symptoms appear in the respiratory tract- nasal congestion, nose bleeds, shortness of breath and cough with bloody phlegm. Other early symptoms may include joint pain, decreased hearing, skin rash, conjunctivitis or episcleritis.[3] Saddle nose deformity due to nasal septal cartilage destruction is a characterized abnormality. Although CNS (central nervous system) manifestations are rare presentation of granulomatosis with polyangiitis, cerebral small vessel vasculitis occurs in approximately 4% of patients.[4] It has been associated with seizures, intracranial hemorrhage, transient ischemic attacks, arterial and venous thrombosis. Mostly the peripheral nervous system is affected, resulting in mononeuritis multiplex or polyneuropathy.[5] In this patient, the earliest manifestation was neurological, with later development of typical upper airway and lung symptoms. This atypical presentation resulted in delay of diagnosis.

Secondary CNS involvement by systemic vasculitis occurs in less than one fifth of patients but may be devastating.[6] A prompt recognition and aggressive treatment is crucial to avoid paramount damage and dysfunction. Despite lack of specific findings in MRI of brain, with the typical cavitary lung lesions and upper airway involvement along with positive c-ANCA, cerebral vasculitis was the likeliest explanation for Parkinsonism in this patient.

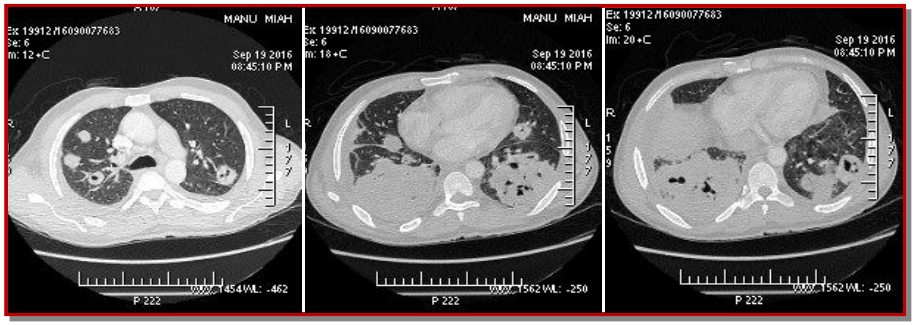

Figure 2: HRCT chest showing bilateral consolidation with multiple cavitary lesions and pleural effusion

Conclusion

We described a patient with granulomatosis with polyangiitis, who developed Parkinsonism as the earliest manifestation long before any other organ involvement. A high degree of suspicion is of utmost value for diagnosing vasculitis, especially when unusual neurological symptom in a young patient could not be explained otherwise.

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