ejpmr, 2022,9(2), 542-544

SJIF Impact Factor 6.222

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

MULTISYSTEM INVOLVEMENT IN WEGENER'S GRANULOMATOSIS – A CASE REPORT

Dr. Ranjit Kaur[1], Dr. Ketaki Kainth[2] and Dr. Ashish Kumar[3]*

[1]M.D Medicine Civil Hospital Rajgarh Sirmaur H.P.
[2]Medical Officer Civil Hospital Nurpur H.P.
[3]M.D Medicine Civil Hospital Jawali Kangra H.P.

***Corresponding Author: Dr. Ashish Kumar**
M.D Medicine Civil Hospital Jawali Kangra H.P.

Article Received on 06/12/2021 **Article Revised on 26/12/2021** **Article Accepted on 16/01/2022**

ABSTRACT

A 36 years male presented with gradually progressive shortness of breath associated with decreased hearing and blurring of vision, with repeated episodes of sinusitis. Examination revealed saddle nose and high frequency hearing loss. Further, blood investigations showed anemia with acute kidney injury and hematuria with macroscopic proteinuria. cANCA was positive. Renal biopsy suggested pauci immune glomerulonephritis. FESS showed septal perforation with granuloma formation and necrosis. MRI brain showed findings suggestive of optic neuritis. This case presented with typical presentation of wegener's granulomatosis, symptoms ignored by patient for 4 years, and came to care only after renal failure has developed. Patient was managed with pulse therapy with cyclophosphamide and hemodialysis support.

KEYWORDS: Granulomatosis with polyangiitis, Wegener's granulomatosis, optic neuritis.

INTRODUCTION

Granulomatosis with polyangiitis, formerly called Wegener's granulomatosis is an uncommon disorder and is one of the medium and small vessel necrotizing vasculitis. The prevalence of the disease is about 3 in 100,000, with a slightly higher prevalence in men than in women (3:2). The peak incidence of the disease is at 50 to 60 years of age.

This disorder causes inflammation of the blood vessels in nose, sinuses, throat, lungs and kidneysleading to synptoms associated with organ involvement.

CASE REPORT

A 36 years male presented with complaints of insidious, gradually progressive easy fatiguability associated with exertional shortness of breath. No associated history of chest pain, orthopnoea, cough. History of decreased hearing in bilateral ears for 20 days associated with tinnitus, no history of discharge, earache. Later, patient noticed painless blurring of vision bilateral eyes without eye discharge or redness. Patient was evaluated and detailed past history revealed that patient has repeated episodes of nasal congestion and epistaxis with sore throat for 3 years for which he used to take over the counter treatment. General examination revealed patient had pallor, pedal edema, saddle shaped nose deformity(Fig. 1), sinuses in palate(Fig. 2). Eye examination visual acuity right and left eye finger counting closed to face and 3 metres respectively. Fundus showed hyperemia of disc. Pupillary reaction right eye showed RAPD. Ear examination showed sensorineural hearing loss. Pt was further evaluated with blood investigations.

Figure 1: Saddle nose deformity.

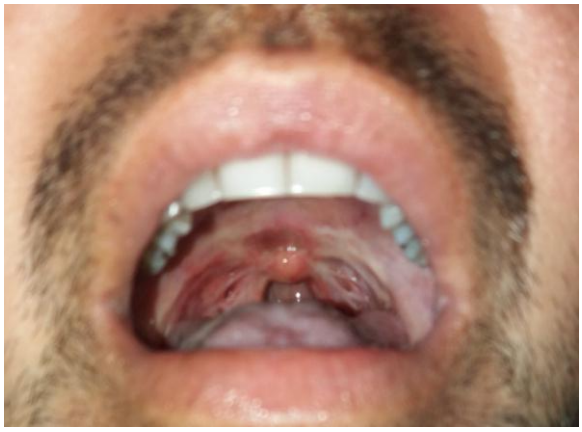

Figure 2: Palate sinuses.

Investigations

Hemoglobin: 7gm/dL, TLC: 11.9 thou/uL, platelets: 754 thou/uL, ESR: 140 mm1st hr. LFT within normal range.
BUN: 94 mg/dL, Serum creatinine: 11.94 mg/dL, serum calcium: 8.6 mg/dL, phosphorus: 6.1 mg/dL,
Urine protein: 3+, RBC's: 5-7 /HPF
24 hrs urine protein: 3509 mg/24hrs

ABG showed Metabolic acidosis with respiratory alkalosis.
iPTH: 265pg/dL (15-68)
S. iron: 21ug/dL (70-180)
% saturation: 11% (20-50)
TIBC: 191ug/dL (250-450)
S. ferritin: 263ng/dL (21-274)
ANA by hep 2 method: negative
cANCA: positive, (by EIA method : >100 U/mL)
pANCA: negative
AntiGBM: negative
C3/ C4: normal limits
USG abdomen: increased cortical echogenicity with decreased CMD.
HRCT chest: soft tissue nodules (9.2 x 10.2mm) in superior segment of RLL.
MRI Brain: Soft tissue and mucosal thickening in Frontal sinus on right side, Maxillary, sphenoid and mastoid air cells.

Normal study for brain parenchyma except for a calcified granuloma in temporal lobe. Bilateral optic nerve shows mild hypointense signal intensity (right > left), rest of obrit and optic nerve normal.

FESS: Crusts present B/l nasal cavity. Septal peroration at bony – cartilagenous junction, (punch bx taken :- cartilagenous tissue with inflammation & Granulation). Granulation present in lateral wall of left nose(Fig. 3). (punch bx taken :- Granulation tissue with areas of necrosis). Thick & unhealthy mucosa on lateral wall bilateral nostrils.

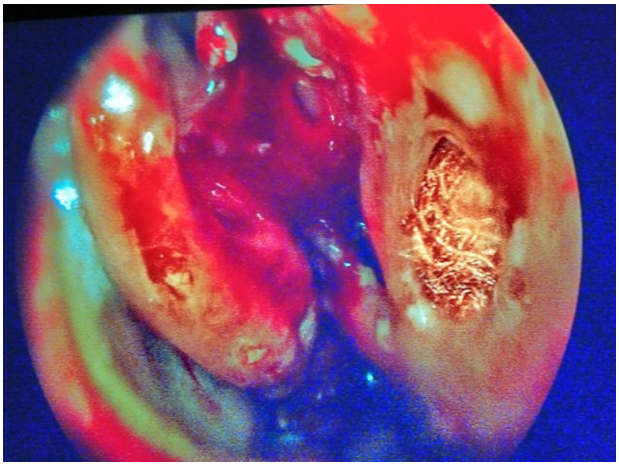

Figure 3: Fess image showing crusts and inflammation.

PTA: high freaquency hearing loss bilateral ear
Renal Biopsy: All glomeruli in the bx specimen shows global crescents.
One glomeruli showing segmental necrotizing lesion.
Acute tubular injury with interstitial edema & lymphocytes & monocytes infilterates. Pauci – immune crescentric Glomerulonephritis.

Patient managed with four rounds of hemodialysis followed by methylprednisolone pulse therapy followed oral prednisolone at 1mg/kg and injection cyclophosphamide at 15mg/kg q2wkly for 3 doses followed by consolidation phase q 3 weekly.
Patient discharged and advised follow up.

DISCUSSION

Although GPA was first described by Klinger as a form of polyarteritis nodosa (PAN), the unique nature of the disease was recognized earlier by Wegener. The term GPA was first introduced into the English literature in 1954 by Godman and Churg.

Wegener granulomatosis, or granulomatous vasculitis, is a disease that produces inflammation of the medium and small arteries and venules,[2,4] Necrotizing and crescentic changes are found in the glomeruli.[1] The process typically affects the upper and lower airways and kidneys.

This patient presented with the classic clinical triad of Wegener granulomatosis: upper respiratory, pulmonary, and kidney involvement.[2,3] A tissue biopsy is essential for the definitive diagnosis of Wegener granulomatosis. Initial therapy generally consists of cyclophosphamide and glucocorticoids.[5] Alternative regimens include 1) intravenous monthly cyclophosphamide instead of daily, oral cyclophosphamide; 2) methotrexate instead of cyclophosphamide in patients with mild disease, limited bone marrow reserve, or bladder toxicity; and 3) plasmapheresis, especially when anti–glomerular basement membrane antibodies are present or when severe pulmonary hemorrhage occurs.[6]

CONCLUSION

Wegners granulomatosis is an uncommon disease with common early presenting symptoms. Therefore its diagnosis requires a high index of suspicion so early treatment can be instituted and further organ damage can be prevented.

REFERENCES

1. Haas M, Eustace JA. Immune complex deposits in ANCA-associated crescentic glomerulonephritis: a study of 126 cases. *Kidney Int.*, 2004; 65(6): 2145–2152.
2. Manganelli P, Fietta P, Carotti M, Pesci A, Salaffi F. Respiratory system involvement in systemic vasculitides. *Clin Exp Rheumatol*, 2006; 24(2 Suppl 41): S48–S59.
3. Duna GF, Galperin C, Hoffman GS. Wegener's granulomatosis. *Rheum Dis Clin North Am.*, 1995; 21(4): 949–986.
4. Seo P, Stone JH. The antineutrophil cytoplasmic antibody–associated vasculitides. *Am J Med*, 2004; 117(1): 39–50.
5. White ES, Lynch JP. Pharmacological therapy for Wegener's granulomatosis. *Drugs*, 2006; 66(9): 1209–1228.
6. Levy JB, Hammad T, Coulthart A, Dougan T, Pusey CD. Clinical features and outcome of patients with both ANCA and anti-GBM antibodies. *Kidney Int.*, 2004; 66(4): 1535–1540.