Case Report: A Rare Yet Life-Threatening Mimicker of Chronic Conjunctivitis

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ABSTRACT

Wegener's granulomatosis (WG) is a necrotizing granulomatous inflammation. A 40-year-old lady of Malay descent presented with unresolving bilateral painful red eyes for three weeks. Clinical examination revealed that best corrected vision acuity of 6/9 for both eyes. Slit lamp examination revealed diffuse scleritis. Other Investigations result like UFEME blood cell: 2+, C-reactive protein 70.06 mg/L and ESR of 125 mm/h suggestive on acute inflammations. Connective tissue screening revealed cytoplasmic ANCA was positive and was supported by Anti-Serine Protease3 (PR3) 68. All the investigation results revealed that she had Wegener's granulomatosis with ophthalmology manifestation in the form scleritis. Patient was treated with guttae Maxidex QID to reduce cells that present in anterior chamber and oral ibuprofen 400 mg thrice daily. Subsequently, oral prednisolone and oral cyclophosphamide with oral Bactrim were commenced. Patient responded well and redness resolved. There are many differential diagnoses for chronic conjunctivitis but to rule out connective tissue disease should be one of the primary differential diagnoses in young female. Oral immunosuppressive and Trimethoprim/ Sufmethoxazole (Bactrim) were been found beneficial and symptoms were resolved. Wegener's granulomatosis is a great mimicker as exemplified in this case. This disease can be misdiagnosed and maltreated as conjunctivitis. Thus, the authors wish to emphasize that WG is one the differential diagnoses that need to be considered in a person with bilateral scleritis.

Keywords: Wegener's granulomatosis, scleritis, chronic conjunctivitis

INTRODUCTION

Wegener's granulomatosis (WG) is a necrotizing granulomatous inflammation involving small to medium vessels in many organs.[1] Various tests are needed for diagnosis of Wegener's granulomatosis, based on the clinically, radiological and serological findings.[2] WG is a great mimicker. Chronic conjunctivitis is one of the rare manifestations of WG. Here, the authors report a case of Wegener granulomatosis with ocular involvement.

CASE PRESENTATION

A 40 year-old-Malay-female, presented with episodic bilateral eye redness which did not resolve for the past three weeks. Patient however denied any ocular trauma or contact with foreign body. It was associated with dull aching pain upon eye movement only. Further examination revealed that her best corrected vision acuity for both eyes was 6/9. There was no relative afferent pupillary defect. Slit lamp examination revealed non-necrotizing anterior diffuse scleritis bilaterally. There were cells presented in anterior chambers of both eyes. Slit lamp examination diffuse scleritis bilaterally. Indirect ophthalmoscopy with mydriatics drops revealed a normal fundus. Figure 1 shows the initial appearance of the eyes. Cardiovascular, respiratory and abdominal examinations were unremarkable. The intraocular pressure for both eyes was at 14 mmHg. Blood and urine investigations were done (see Table 1).

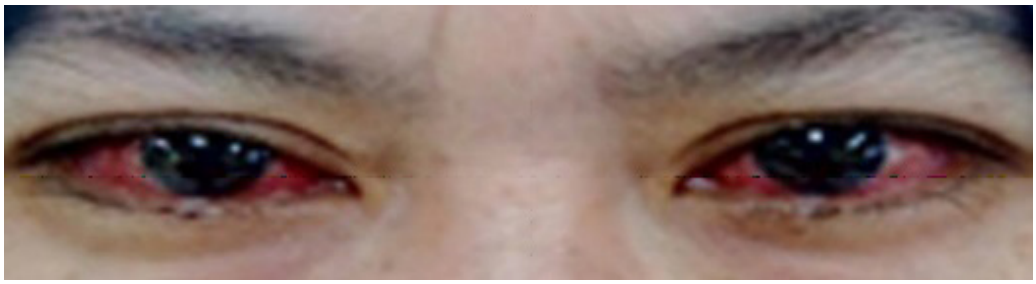

Figure 1 Initial appearance of the eye

Table 1 Investigations

Full blood count	
White Cell Count	12,200/ml
Haemoglobin	11.6 g/dL
Platelet	495,000/ml
Erythrocyte Sedimentation Rate	125
MCV	75.5 fL
MCH	23.3 pg
MCHC	30.9 g/dL
Biochemical parameters	
C-Reactive Protein	70.06 mg/L (<5)
Creatinine	59 mmol/L
Urine Analysis	
pH	6.5
Blood	2+
Protein	Negative
Red Blood Cell Cast	Negative

Immunological parameter	
Antinuclear antibodies	Negative
C-ANCA	Positive
P-ANCA	Negative
RA	Positive
MPO	< 0.0(CU)
PR3	68 (CU)

On investigations, she had hypochromic microcytic anaemia which was confirmed via full blood count accompanied by raised inflammatory markers (Table 1). TB workup for her was negative. She was screened for Staphylococcal Aureus nasal carriage which turned out to be negative. Her urine analysis showed the presence of red blood cells which was later resolved upon treatment commencement. Routine chest x-ray revealed a suspicious right upper lobe with cavitation. So, CT scan of thorax was done. Her CT scan of thorax showed dense consolidation foci at other lobes with ground glass opacity as shown in Figure 2. Immunological parameter revealed cytoplasmic ANCA was positive and was supported by Anti-Serine Protease3 (PR3) 68 (normal < 3.0 CU). Based on these evidences, diagnosis of WG was made.

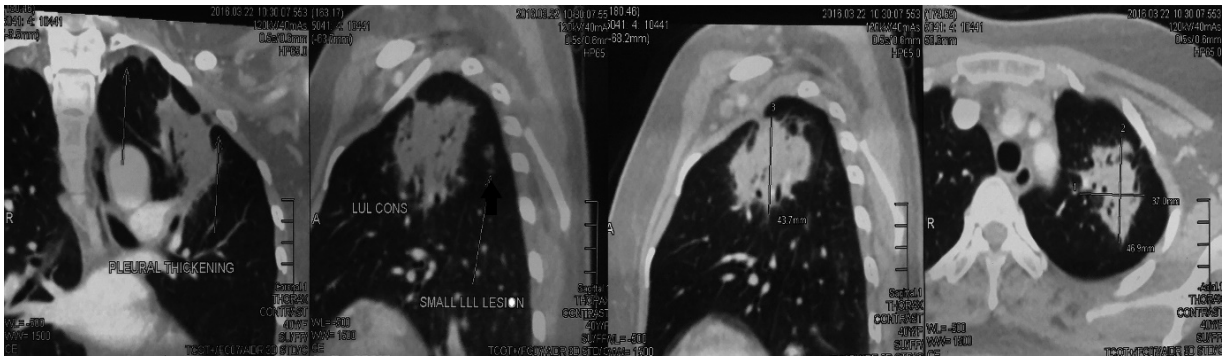

Figure 2 Left upper lobe focal dense consolidation (dark-coloured arrow) with speculation and adjacent fibrosis and pleural thickening on a background of ground glass opacity

Patient was treated with 6-hourly guttae Dexamethasone to reduce cells that present in anterior chamber. In additional to that, the authors added oral ibuprofen 400 mg thrice daily to reduce inflammation. However, redness of both eyes did not resolve. Immunosuppressive therapy in the form of oral prednisolone and oral Azathioprine was initiated with prophylaxis Trimethoprim-Sulfamethoxazole (Bactrim). Patient responded well and eye redness resolved (see Figure 3). Currently she is doing well and on regular follow-up.

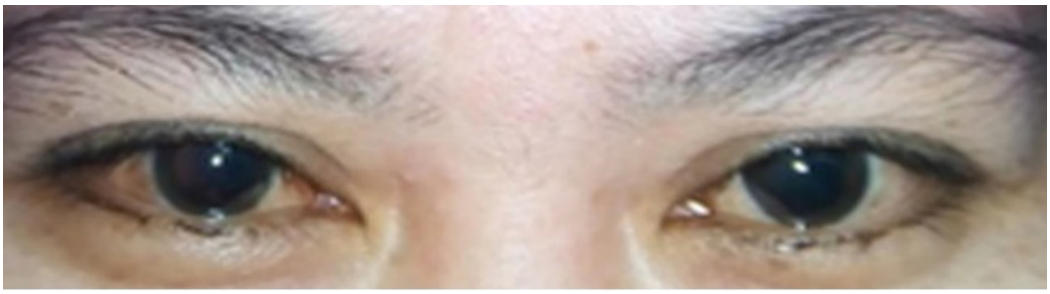

Figure 3 After commencement of treatment

DISCUSSION

Chronic conjunctivitis is one of the rare manifestations of WG. The non-responding nature of conjunctivitis coupled with the lung radiological findings lead to suspect vasculitis. High resolution computer tomography of thorax later confirmed a left upper lobe dense consolidation with speculation with adjacent fibrosis and pleural thickening on a background of ground glass appearance. These radiological features are in consistent with Wegener's granulomatosis.[3] A set of investigations are needed for supportive diagnosis tools. Antineutrophil cystoplasmic antibody test (ANCA) is an important diagnostic criterion for WG. More specific for WG is c-ANCA is an autoantibody directed against the neutrophil serine protease.[2] The c-ANCA test has a high sensitivity (96%) for WG.[4] Tissue biopsy from the suspected region of lesion is essential as to give a confirmatory histology diagnosis. This patient refused to do tissue biopsy. On investigation, raised inflammatory markers supported WG. The aetiology of WG is unknown and postulated causes might be autoimmune origin, genetic predisposition, connective tissue disease, viral or hypersentivity interaction.[5] Clinical presentations are heterogeneous and can be either insidious or acute. The patient was started with oral prednisolone of 1 mg/kg per day and oral Azathioprine. Both her eyes and constitutional symptoms resolved with treatment (Figure 3). Staphylococcus

Aureus is one of the aetiological factors for WG in some patient and serve as trigger factor.[6] Trimethoprim /Sulfamethoxazole (Bactrim) has been found beneficial and prevent S. Aureus infections.[7] As such she was on Trimethoprim/ Sulfamethoxazole (Bactrim) apart from her immunosuppressant therapy.

CONCLUSION

Connective tissue diseases may be one of the differential diagnoses for female in reproductive age. There are many differential diagnoses for chronic conjunctivitis but connective tissue disease especially WG should be kept in mind as one of the primary differential diagnoses in female. High index of suspicion can prevent mistreatment and eventually prognosis of the disease which may be fatal if untreated.

ACKNOWLEDGEMENTS

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CONFLICT OF INTEREST

The authors declare that they have no competing interests.

CONSENTS

Written informed-consent was obtained from the patient to publish the case with its related pictures. A copy of the written consent is available for review by the Chief Editor of this journal.

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