A case of lung mass with no perception of light: Extreme ocular presentation of granulomatosis polyangiitis

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Key words: Granulomatosis polyangiitis, panophthalmitis, scleritis

A 60-year-old female presented with sudden painful visual loss and photophobia in left eye (LE) since 15 days. LE had undergone cataract surgery eight months back. Visual acuity was 6/6 in right eye, and no perception of light (PL) in LE. Right eye was unremarkable, whereas LE had congested scleral vessels [Fig. 1a and b]. Anterior chamber showed

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Received: 26-Apr-2019 Revision: 08-Jul-2019 Accepted: 23-Jul-2019 Published: 22-Oct-2019 intense inflammation with perilenticular fluffy exudates. Vitreous cavity was full of yellowish exudates with subretinal exudation seen hazily with ophthalmoscope. Sonography of LE showed vitreous exudates, shallow retinal detachment, and periocular exudation around the optic nerve. Infective sclerouveitis/panophthalmitis was presumed. Investigations revealed neutrophilia (~85%), raised ESR (78 mm/h), and a lung mass/large nodule in left lung [Fig. 1c]. After discussion with the pulmonologist, Contrast Enhanced Computed Tomography (CECT) chest and orbits, and Cytoplasmic antineutrophil cytoplasmic antibodies (c-ANCA) were ordered. Intensive antifungal regime was started with oral indomethacin suspecting pulmonary aspergilloma-related endogenous infection.

Two days hence, significantly high c-ANCA (17.69 U/ml) was detected. Later radioimaging revealed multiple cavitary lesions in both lungs (L > R) [Fig. 1d] and periscleral inflammation [Fig. 1e]. The diagnosis was revised to granulomatosis polyangiitis (GPA). High-dose intravenous steroids for three days and single high-dose cyclophosphamide were given with supportive topical therapy, and later the patient was shifted to similar oral therapy. One month later, she was painless, anterior chamber was clear, and scleritis had dampened with scleral thinning, but vision persisted to be low (PL+). The lung mass had reduced considerably on skiagram.

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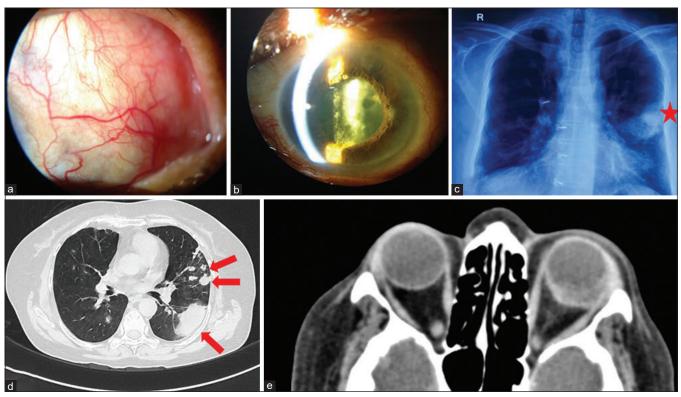


Figure 1: (a and b) Slit lamp examination of left eye showing congestion of deep episcleral and scleral vessels, and fluffy-fibrinous exudates around the lens. (c) Chest X-ray showing left lung mass/lucent round lesion (cavitary nodule, star). (d) CT chest showing multiple large nodules in both lungs. Left lung nodules are conglomerating to make a mass in posterior segment of left upper and lower lobes (arrows). (e) CT orbit showing left eye scleritis (posterior scleral thickening) with intravitreal/subretinal inflammatory collection

GPA is progressive with associated mortality if not treated timely. In total, 11% can have scleritis as initial presentation.^[1-3] This case was marked by the severity of its presentation leading to the initial misdiagnosis of infective etiology. Even the initial blood and imaging workup were suggestive of fungal infection, but lack of risk factors and general condition were against fungal endogenous infection.^[4,5] Continued surveillance with clinical suspicion, aided by aggressive multidisciplinary interaction led to the accurate management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed. Financial support and sponsorship Nil.

Conflicts of interest

There are no conflicts of interest.

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