Atypical optic neuritis as the presenting feature of testicular malignancy: Rare case report with review of literature

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A 23-year-old male presented with unilateral acute onset, painless, progressive diminution of vision. Initial clinical and radiological findings were consistent with optic neuritis. The patient was started on pulse steroids leading to visual recovery. However, the patient relapsed within one week. Repeat imaging revealed metastatic lesions near the orbital apex. Orbit is an unusual site of metastasis from testicular tumors and only 3 cases of testicular synovial sarcoma with orbital metastasis have been reported. A thorough systemic evaluation should be done in cases of atypical and relapsing optic neuritis to rule out the causes of optic neuritis (ON) masquerade syndrome.

Key words: Atypical optic neuritis, pulse steroids, testicular synovial sarcoma

Optic neuritis (ON), as the term suggests, literally refers to an inflammatory pathology of the optic nerve resulting from an idiopathic, demyelinating, inflammatory, infectious, or an infiltrative process. "Typical" ON most commonly represents non-inflammatory demyelination of the optic nerve. "Atypical" cases of ON belong to the subset of causes other than non-inflammatory demyelinating processes. Of these, rarely, ON may be the initial presenting feature of systemic malignancy, thus constituting the "Optic Neuritis Masquerade Syndrome". We report a rare case of testicular synovial sarcoma masquerading with symptoms of typical ON.

Case Report

A 23-year-old male patient presented with acute onset, painless, progressive diminution of vision in the right eye for 3 days. Examination revealed a visual acuity of 5/60 Snellen, not improving with pinhole. The right eye revealed abnormal color vision as per Ishihara charts while contrast sensitivity with Pelli-Robson charts was reduced to log 0.45. The right eye also had a Wagner grade 3 relative afferent pupillary defect (RAPD). Fundus examination showed a hyperemic edematous optic nerve head with congested peripapillary retinal nerve fiber layer [Ref. Fig. 1]. The left eye was unremarkable.

Visual field testing revealed high fixation losses and generalized depression of visual field in the right eye. Magnetic resonance imaging (MRI) of brain and orbit showed altered signal intensity in right optic nerve without any evidence of demyelinating lesions in brain. A diagnosis of idiopathic optic neuritis (ON) was made and patient was started on pulse therapy with 1 gm intravenous methyl-prednisolone for 3 days followed by 1 mg/kg oral prednisolone for 11 days. On day 3 after steroid pulse therapy, the patient's best corrected visual acuity (BCVA) improved to 6/6 in the right eye, color vision was normal and contrast sensitivity improved to log 1.50.

However, 10 days after stopping oral steroids, the patient again presented with similar symptoms in the right eye. Color

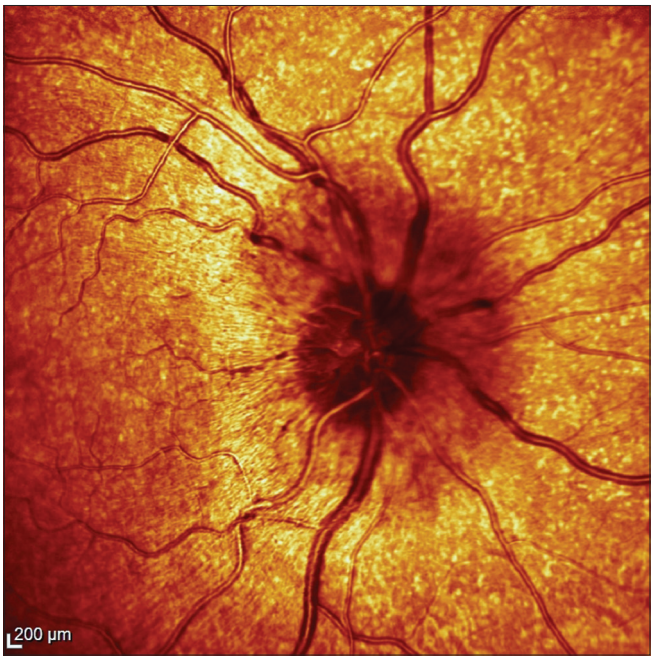

Figure 1: Fundus photograph showing blurring of disc margins, disc hyperemia with peripapillary congestion

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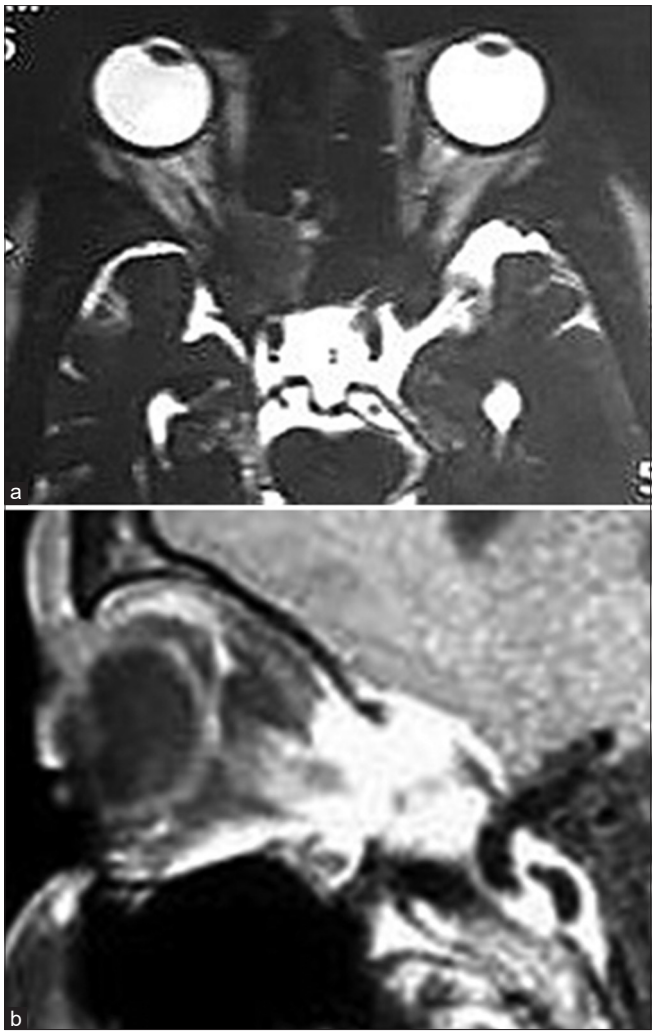

Figure 2: (a) (T1-weighted sagittal MRI showing) and (b) (T2-weighted sagittal MRI showing): Altered signal intensity lesions involving the right orbital apex encasing the right optic nerve in the optic canal and extending into the sphenoid sinus and posterior ethmoidal air cells

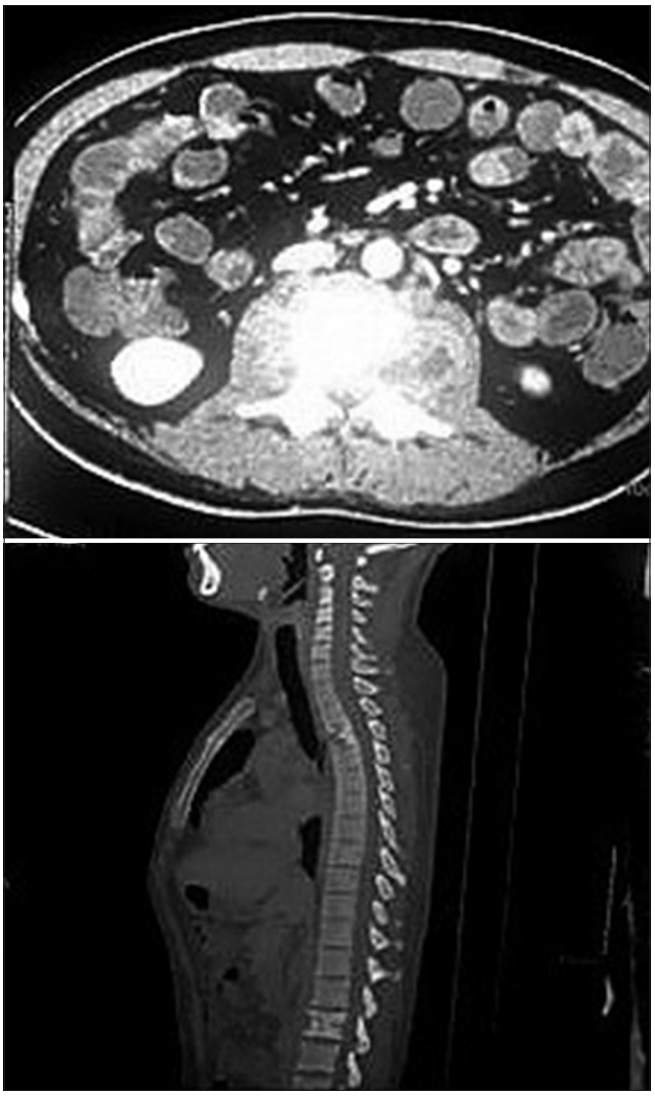

Figure 3: CECT spine showing lytic lesions involving the vertebral body, along with heterogeneously enhancing soft tissue lesion around L2 extending into bilateral psoas muscles

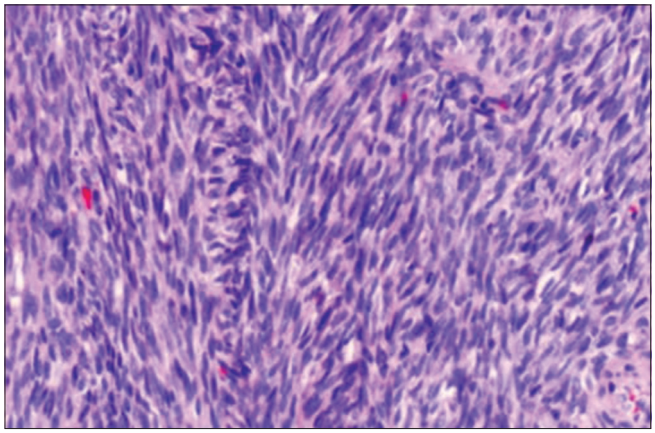

Figure 4: Histopathological H and E slide depicting testicular synovial sarcoma

vision and contrast sensitivity were reduced with a grade 4 RAPD in the right eye. MRI of brain and orbit was repeated, which revealed altered signal intensity lesions involving the right orbital apex encasing the right optic nerve in the optic canal and extending into the sphenoid sinus and posterior ethmoidal air cells along with post contrast heterogeneous enhancement [Ref. Fig. 2a and 2b]. Also, an extra-axial heterogeneously enhancing lesion was seen in the left parietal lobe causing adjacent bone destruction suggesting metastatic infiltration.

Contrast enhanced computerized tomography (CECT) of thorax showed hetergeneously enhancing enlarged paraaortic and left hilar lymph node with hypodense core. The left lung showed multiple small solid nodules involving the superior and posterobasal segment of the left lower lobe. CECT of spine showed similar findings suggestive of lytic lesions involving the vertebral body, along with heterogeneously enhancing soft tissue lesion around L2 extending into bilateral psoas muscles [Ref. Fig. 3].

Computed tomography (CT) guided biopsy from vertebrae reported mesenchymal tumor on histopathological examination

[Ref. Fig. 4]. Serum human chorionic gonadotropin (HCG), lactate dehydrogenase (LDH), and alpha-fetoprotein (AFP) levels were raised. The patient was diagnosed with testicular synovial sarcoma and referred to the medical oncology department for chemotherapy. The patient was started on intravenous (IV) Inj. ifosfamide 2 gm, Inj. mesna 300 mg, and Inj. adriamycin 30 mg. As per the recommendations of the medical oncology team, IV dexamethasone was started for ophthalmic involvement in the 3rd chemotherapy cycle. The patient is now on follow-up.

Discussion

ON is a common neuropathy affecting young adults that may affect intraocular portion of the optic nerve termed "Papilitis" or retrobulbar portion termed "Retrobulbar neuritis", respectively. "Typical" or demyelinating ON may be associated with multiple sclerosis and is the most common form affecting young patients. "Atypical" cases of ON belong to the subset of causes other than non-inflammatory demyelinating processes, namely inflammatory, infectious, or an infiltrative process.[1]

Rarely, however, metastatic spread to orbit from a primary malignancy elsewhere could manifest as clinical features of ON. Moreover, symptoms and signs might even improve temporarily with pulse steroid therapy in such patients, making the diagnosis even difficult. Approximately 1%–13% of all orbital tumors are secondary metastasis from primary malignancy, and about 2%–5% patients with systemic malignancy develop orbital metastasis.[2-5] Breast, lung, and prostate are the common sites for primary malignancy along with secondary involvement of orbit with the breast being the most common site in both sexes.[6] Metastasis to orbit generally presents clinically with proptosis, diplopia, decreased vision, or palpable mass.[2]

Our patient presented with clinical features of typical ON without any radiological evidence of alternative etiologies and improved dramatically on systemic pulse steroids. It was only after the relapse after 10 days of stopping systemic steroids that extensive search for secondary causes was done and the patient was diagnosed to be having testicular synovial sarcoma with extensive metastasis to lungs, vertebrae, and orbits. Testicular malignancies usually affect adolescents and young adults; metastasis occurs to lung, retroperitoneal lymph nodes, liver, mediastinal lymph nodes, brain, kidney, gastrointestinal tract, bones, adrenals, peritoneum, and spleen in decreasing order of frequency.[6] Orbit is an unusual site of metastasis from testicular tumors and only 3 cases of testicular synovial sarcoma with orbital metastasis have been reported in literature till date.[7-10] Testicular synovial sarcoma is a rare malignancy and has been reported in literature only once.[11] To have one with metastasis to orbit is even rarer. We believe that this is the first reported case of a primary testicular synovial sarcoma with secondary metastasis in the orbit.

Conclusion

Rarely, ON may be the initial presenting feature of systemic malignancy, thus constituting the "Optic Neuritis Masquerade Syndrome". Diagnosis in such cases requires clinical precision, a high index of suspicion, and a detailed systemic evaluation.

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.

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Conflicts of interest

There are no conflicts of interest.

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