Photo Essay

Central retinal vein occlusion as the initial presentation in leptomeningeal carcinomatosis

Anusha Venkataraman, Bijnya Birajita Panda, K Nagarajan[1]

A 72-year-old male presented with complaints of sudden diminution of vision in his left eye associated with a severe headache, nausea and vomiting of 15 days duration. He gave a history of noncompliance to chemotherapy for gastric adenocarcinoma (after partial radical gastrectomy) 7 months back. Best-corrected visual acuity in the right eye was 20/40 and no light perception in the left eye. An afferent pupillary defect was detected in the left eye. Fundoscopy revealed papilledema in both eyes with a central retinal vein occlusion (CRVO) in the left eye [Fig. 1]. There was no involvement of cranial nerves other than both optic nerves. Visual fields tested by confrontation field testing were within normal limits for the right eye. Magnetic resonance imaging (MRI) brain and orbit showed irregular dilatation of ventricles with periventricular hyperintensities suggestive of extraventricular obstruction. Thickening and enhancement of bilateral optic nerve sheath complex [Fig. 2] were also noted raising the possibility of leptomeningeal carcinomatosis (LC). Cerebrospinal fluid (CSF) cytology revealed signet ring cells [Fig. 3] as seen in adenocarcinoma. He was referred to the oncology for intrathecal chemotherapy and palliative radiotherapy but due to his deteriorating general condition he died within 2 months.

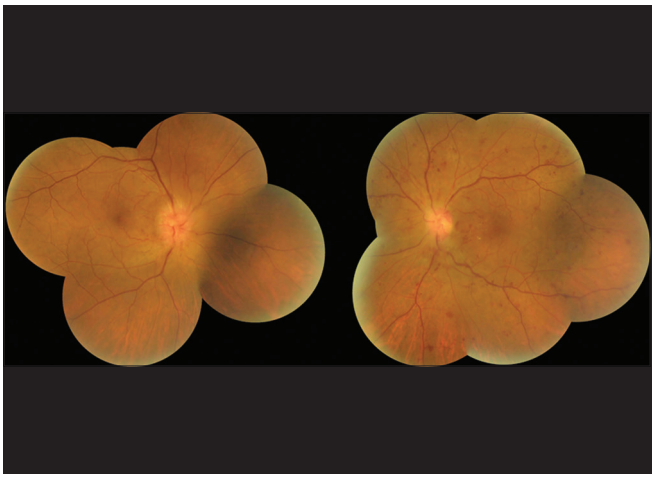

Figure 1: Colour fundus montage photographs showing papilledema in both eyes and central retinal vein occlusion in left eye

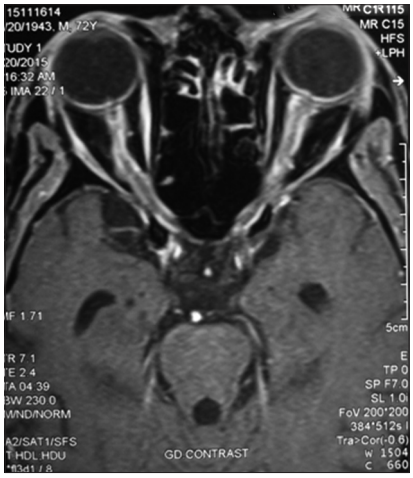

Figure 2: Magnetic resonance imaging of brain and orbits showing bilateral optic nerve sheath thickening with lumpy infiltration along the sheath margins

Discussion

The incidence of retinal vein occlusion varies from 2/1000–8/1000 people, seen typically above 65 years of age with conditions such as diabetes mellitus, hypertension, collagen vascular diseases, and hyperviscosity syndromes.[1-3] A neoplastic cause is, however, rare. LC, an infiltration of the pia mater and the arachnoid membrane by malignant cells is less commonly associated with solid tumors; lung, breast and melanoma being the ones most often reported. Gastric cancer complicated by LC as observed in our patient is very rare. It is estimated to occur in 0.16% of all cases of gastric cancer,[4] of which 87% have disseminated disease.[5] The most common ocular manifestations in LC include visual loss

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Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/0301-4738.181745

Departments of Ophthalmology and [1]Radiology, All India Institute of Medical Sciences, Bhubaneswar, Odisha, India

Correspondence to: Dr. Bijnya Birajita Panda, Department of Ophthalmology, All India Institute of Medical Sciences, Bhubaneswar, Odisha, India. E-mail: bigyan_panda@yahoo.co.in

Manuscript received: 17.07.15; Revision accepted: 16.01.16

Cite this article as: Venkataraman A, Panda BB, Nagarajan K. Central retinal vein occlusion as the initial presentation in leptomeningeal carcinomatosis. Indian J Ophthalmol 2016;64:225-6.

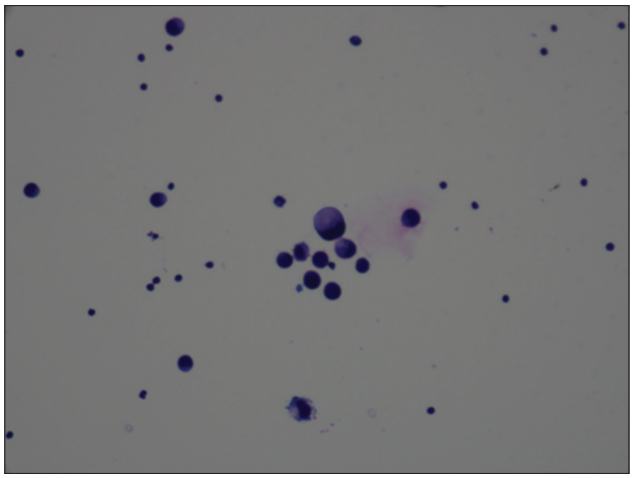

Figure 3: Cerebrospinal fluid specimen with May–Grunwald–Giemsa stain under ×40 showing few signet ring cells (cell nucleus pushed to the periphery) in a background of lymphocytes and arachnoid cap cells

due to optic atrophy or optic neuritis, followed by diplopia due to cranial nerve palsies, ptosis, papilledema, anisocoria, exophthalmos, orbital pain, scotomas, hemianopsia, and nystagmus.[6] CRVO as an initial presentation in LC as seen in our patient with an inadequately treated gastric adenocarcinoma is not reported in literature until date except for a single case report of combined central retinal artery and CRVO in a patient with breast carcinoma.[7] Hence, an elderly patient presenting with CRVO and a no PL eye in the context of malignancy warrants high suspicion and detailed evaluation including MRI and CSF cytology to diagnose this rare association.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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