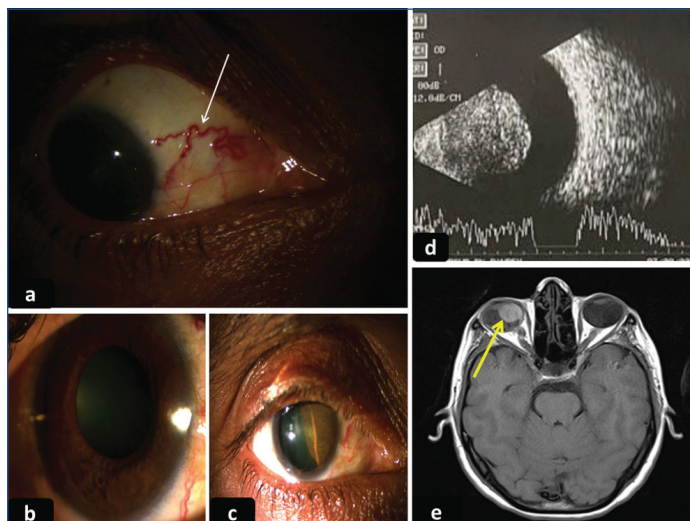


Sentinel Vessel Sign: A Shadow of the Subway Storm Setting into Strike Sight

RAKESH KUMAR JHA¹, JAYA KAUSHIK², SUNANDAN BHATTA³**Keywords:** Ciliary body melanoma, Melanoma, Ocular tumour

A 56-year-old woman presented with a complaint of painless diminished vision in the right eye for nine months. Best-corrected visual acuity was 20/400 Oculus Dexter (OD) and 20/20 Oculus Sinister (OS). Ophthalmological examination revealed a prominent solitary, tortuous, dilated episcleral sentinel vessel in the superonasal aspect of the right eye (white arrow, [Table/Fig-1a]). An undilated examination revealed a normal pupillary area [Table/Fig-1b], but a dilated examination revealed a retrolental dome-shaped dark-brown mass protruding into the vitreous cavity [Table/Fig-1c]. Ultrasonography revealed a 9 mm acoustically-solid mass in maximum dimension with medium internal reflectivity [Table/Fig-1d]. A diagnosis of ciliochoroidal melanoma was made. A Computed Tomography (CT) scan of the head excluded extraocular extension (yellow arrow, [Table/Fig-1e]). Metastatic work-up, along with radiological imaging, excluded distant metastasis. Given the involvement of the ciliary body (which carries a worse prognosis), the involved eye was enucleated after due counseling with the patient. Subsequent histopathological examination, along with immunohistochemistry analysis of the lesion, revealed it to be a grade III-pT1-stage malignant melanoma with no local invasion. After enucleation, subsequent follow-up examinations for one year excluded any recurrence.



[Table/Fig-1]: a) Solitary episcleral sentinel vessel in the superonasal aspect of bulbar conjunctiva (white arrow) of right eye; b) Usual grey reflex in the pupillary area on undilated, and; c) A retrolental dome-shaped dark-brown mass protruding into vitreous cavity on dilated ocular examination; d) Ultrasonography revealing acoustically solid mass with medium internal reflectivity; e) CT-scan head revealing hyperdense sharply marginated mushroom-shaped mass abutting the scleral wall (yellow arrow) with no extraocular extension.

DISCUSSION

Melanomas are usually asymptomatic and may remain clinically inapparent to the patient as well as to the clinician in the initial stage unless examined by widely dilating the pupil [1,2], especially ciliary-body melanomas, which may attain a large size without any clinically evident involvement of the adnexal ocular structures and may silently metastasize to the liver and lungs extrasclerally through emissary veins [3].

Ciliary body melanomas constitute approximately 10% of uveal melanomas [4], with an average age of 55 to 62 years at the time of diagnosis [5]. They have the worst prognosis, with a 10-year mortality rate reaching up to 30%-50% [5]. Vital organ dysfunction resulting from vascular metastasis is the primary cause of mortality among these patients. Visual symptoms usually occur late, either when the tumour obscures the pupil or contacts the lens, and hence, can go undetected by the patient until it attains a large size. Additionally, compared to uveal melanomas, these tumours have vastly different unusual presenting signs, with case reports of orbital inflammation [6], angle-closure glaucoma [7], unilateral pigmentary glaucoma [8], spontaneous hyphema [9,10], or the presence of metastases, sometimes to rare organs like the temporalis muscle at the time of diagnosis [11]. However, in all these reported cases, the sentinel vessel was the most consistent external feature.

Sentinel vessels are disproportionately dilated and tortuous episcleral blood vessels, which are commonly associated with ciliary body melanomas compared with posteriorly located uveal melanomas. They provide clues for the presence of underlying asymptomatic occult ciliary body melanoma [12]. Sometimes, sentinel vessels themselves may be occult, especially with small tumours in the early stages, visible only as trace conjunctival hyperemia. Marvasti AH et al., have illustrated scleral angiography for highlighting these occult hyperemic sectorial vessels for the detection of asymptomatic, suspicious ciliary body melanoma [13]. Scleral angiography in their case was able to detect vascular abnormalities in focal regions of the sclera that otherwise appeared normal on the slit-lamp examination. The occult vessels demonstrated an early arterial phase that quickly included venous filling with pinpoint leaks that expanded gradually with time. In present case, authors made them apparent by blanching the other vessels with topical phenylephrine instillation.

Because this sign is constantly associated with ciliary body melanoma, its presence is additionally helpful in differentiating it from a multitude of its differential diagnoses, like tumours of the ciliary body pigmented epithelium [14] and non pigmented epithelium [15,16], ciliochoroidal effusion, cysts, and several other tumours [17,18], and pseudotumours [19,20].

The episcleral sentinel vessel is a key clinical sign that points to a possible underlying malignancy and is helpful for its early detection to either salvage the eye or avoid life-threatening complications. It is important to be familiar with this sign, as small asymptomatic ciliary body melanoma may skip detection on screening ocular examination, and recognising it may provide a valuable clue for an in-depth clinical examination.

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PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Ophthalmology, Command Hospital, Lucknow, Uttar Pradesh, India.
2. Professor, Department of Ophthalmology, Command Hospital, Lucknow, Uttar Pradesh, India.
3. Assistant Professor, Department of Ophthalmology, Military Hospital, Agra, Uttar Pradesh, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Rakesh Kumar Jha,
Assistant Professor, Department of Ophthalmology, Command Hospital,
Cariappa Road, Lucknow Cantonment, Lucknow-226002, Uttar Pradesh, India.
E-mail: rkjhan2@y7mail.com

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