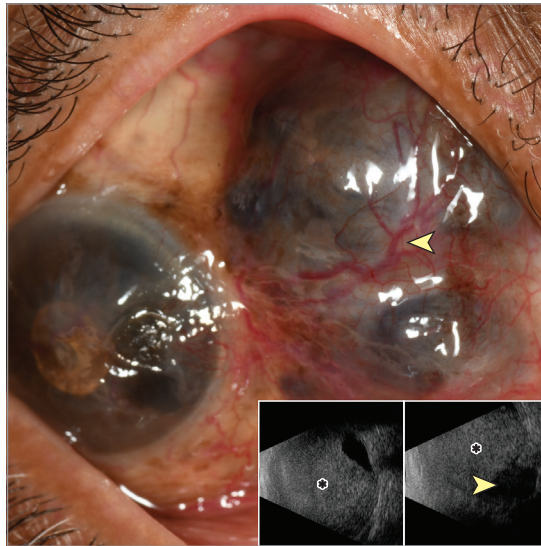


## JAMA Ophthalmology Clinical Challenge

## Subconjunctival Mass 30 Years After Melanoma Resection

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**Figure 1.** Imaging of the right eye at 30 years' follow-up after partial lamellar sclerouvectomy. External photography documenting a superonasal, multilobulated, pigmented lesion with episcleral feeder vessels (arrowhead) located under Tenon fascia. Ultrasonography (inset images) showing homogenous echodensity within the globe (asterisks) with an echolucent extrascleral component (inset, arrowhead).

**A 70-year-old Asian-Indian male** was referred for evaluation of a slow-growing conjunctival mass in the right eye over 27 years. There was no pain or discharge. He had previously undergone partial lamellar sclerouvectomy (PLSU) 30 years prior for ciliochoroidal melanoma in the right eye, followed by pars plana vitrectomy and scleral buckle 1 year later for retinal detachment (RD) with vitreous hemorrhage (VH) in the right eye. On examination, visual acuity was no light perception OD and 20/25 OS, with intraocular pressures of 33 mm Hg and 17 mm Hg in the right and left eyes, respectively. Examination results of the left eye were normal. Anterior segment examination in the right eye revealed thin conjunctiva with a nontender multilobular pigmented lesion in the superonasal quadrant (Figure 1), underlying Tenon fascia and surrounded by prominent episcleral feeder vessels (Figure 1, yellow arrowhead). The cornea showed nasal epithelial defect and flat anterior chamber with cataract and iris neovascularization. There was no view to the posterior segment. Ultrasonography revealed a homogenous echodensity within the vitreous cavity (Figure 1 insets, asterisks) measuring 21 mm in thickness, along with an echolucent episcleral mass (Figure 1 insets, arrowhead) 7 mm thick. In addition, spontaneous vascular pulsations (SVP) were noted within the intraocular component (Video).



Multimedia

## WHAT WOULD YOU DO NEXT?

- A. Scleral patch graft
- B. Magnetic resonance imaging of the head and orbits
- C. Incisional biopsy
- D. Observation

 CME Quiz at [jamacmelookup.com](http://jamacmelookup.com)

## Diagnosis

**Extraocular extension of recurrent ciliochoroidal melanoma**

## What to Do Next

**B.** Magnetic resonance imaging of the head and orbits

## Discussion

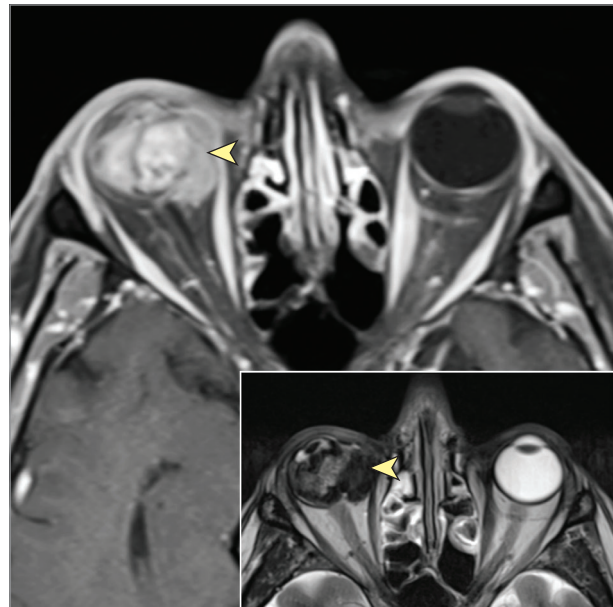
The preferred treatment modalities for uveal melanoma (UM) include plaque radiotherapy or enucleation, depending on tumor extent.<sup>1</sup>

However, PLSU has also been a reasonable procedure for anteriorly located iridociliochoroidal melanoma for several decades.<sup>1-3</sup> This technique of local surgical resection involves creating a hinged, partial-thickness scleral flap to excise the tumor. It avoids radiation complications, such as radiation retinopathy, maculopathy, and papillopathy, as well as the visual penalties associated with enucleation.<sup>2</sup> However, in the early postoperative period (48 hours to 8 weeks), PLSU is associated with RD in 2% to 12% of patients, with VH in 27% to 29% of patients, and with local recurrence in 5% to 11% of patients.<sup>2,3</sup>

Much like the initial diagnosis of UM, the diagnosis of tumor recurrence is made by clinical observation and presents with either-edge recurrence, increase in thickness, or new orange pigment.<sup>4</sup> Posterior segment examination is often limited due to various post-surgical findings, including total RD, VH, or dense cataract, as in this case. In this scenario, certain ultrasonographic features played a crucial role. Apart from the echogenicity and morphology of the intraocular structures, ultrasonography was important in detecting a solid mass with SVP, seen as localized pulsating figures within the mass (Video).<sup>5</sup> These figures denote intrinsic vessels and are pathognomonic of a viable intraocular tumor, as opposed to VH, which would be inert, without pulsations.<sup>6</sup> Although reported in literature, SVP are often overlooked. On the other hand, changes in Doppler ultrasonography are correlated with tumor regression, specifically a decrease in peak systolic frequency following plaque radiotherapy as well as Cyberknife (Accuray) radiotherapy.<sup>7,8</sup>

In this case, SVP was critical in diagnosing tumor recurrence, ruling out postsurgical scleral ectasia and VH. Therefore, scleral patch graft or observation are inappropriate management options. Incisional biopsy is not ideal for melanoma, as it would risk tumor dissemination across the ocular surface, eyelid, and orbit. Gadolinium-enhanced magnetic resonance imaging performed to explore the extent of orbital tumor involvement showed the mass to be hyperintense on fat-suppressed, T1-weighted images (Figure 2) and hypointense on T2-weighted images (Figure 2 inset), suggestive of orbital melanoma recurrence.<sup>5</sup> Limited exenteration with enucleation was performed to remove the globe and extraocular extension in 1 specimen. Histopathology confirmed recurrent ciliochoroidal melanoma with extraocular extension.

UM recurrence can be slow and mimic an ocular surface lesion externally or VH ultrasonographically. Fortunately, tumor recurrence following plaque radiotherapy is rare. However, in the early postoperative period, RD is seen in 1.5% and VH in 4% of patients.<sup>9,10</sup> Ultrasonography focusing on SVP is a relevant clinical tool for tu-



**Figure 2.** Magnetic resonance image showing T1-weighted, fat-suppressed, gadolinium-enhanced image with enhancing intraocular mass and extraocular extension (arrowhead) in the right eye. On T2-weighted imaging (inset), the extraocular extension is with low signal (arrowhead), typical of melanoma.

mor detection. These findings, along with gadolinium-enhanced magnetic resonance images, are valuable surrogates for detecting tumor recurrence.

### Patient Outcome

At 2-month follow-up, the enucleated socket was healed. The patient was planned for a custom-fit prosthesis and orbital radiotherapy. Periodic systemic surveillance for metastatic disease was advised.

### ARTICLE INFORMATION

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### REFERENCES

- Caminal JM, Lorenzo D, Gutierrez C, et al. Local resection in choroidal melanoma: a review. *J Clin Med*. 2022;11(23):7156. doi:10.3390/jcm11237156
- Mirzayev I, Gündüz AK, Okçu Heper A. Partial lamellar sclerouvectomy surgery for anteriorly located uveal tumour resection: a 20-year experience. *Eye (London)*. 2022;36(5):969-977. doi:10.1038/s41433-021-01545-7
- Shields JA, Shields CL, Shah P, Sivalingam V. Partial lamellar sclerouvectomy for ciliary body and choroidal tumors. *Ophthalmology*. 1991;98(6):971-983. doi:10.1016/S0161-6420(91)32194-8
- Binkley EM, Rodriguez SM, Hyer DE, Hinz CJ, Boldt HC. Edge creep: increased pigmentation at the border of choroidal melanomas treated with plaque brachytherapy. *Ocul Oncol Pathol*. 2023; 9(1-2):56-61. doi:10.1159/000531006
- Foti PV, Travali M, Farina R, et al. Diagnostic methods and therapeutic options of uveal melanoma with emphasis on MR imaging-part I: MR imaging with pathologic correlation and technical considerations. *Insights Imaging*. 2021;12(1):66. doi:10.1186/s13244-021-01000-x
- Aironi VD, Chougule SR, Singh J. Choroidal melanoma: B-scan spectrum. *Indian J Radiol Imaging*. 2007;17(01):8-10. doi:10.4103/0971-3026.32691
- Wolff-Kormann PG, Kormann BA, Riedel KG, et al. Quantitative color Doppler imaging in untreated and irradiated choroidal melanoma. *Invest Ophthalmol Vis Sci*. 1992;33(6):1928-1933.
- Kaak C, Kakkassery V, Scheef BO, et al. Evaluation of choroidal melanoma vascularization by color Doppler flow imaging: an option for follow-up tumor control assessment after CyberKnife®? *Medicina (Kaunas)*. 2021;57(6):553. doi:10.3390/medicina57060553
- Bianciotto C, Shields CL, Pirondini C, Mashayekhi A, Furuta M, Shields JA. Vitreous hemorrhage after plaque radiotherapy for uveal melanoma. *Retina*. 2012;32(6):1156-1164. doi:10.1097/IAE.0b013e3182340cc1
- Beykin G, Pe'er J, Hemo Y, Frenkel S, Chowers I. Pars plana vitrectomy to repair retinal detachment following brachytherapy for uveal melanoma. *Br J Ophthalmol*. 2013;97(12):1534-1537. doi:10.1136/bjophthalmol-2013-303331