



# Spontaneous globe rupture from an intraocular melanotic schwannoma

Hannah Yoo, BA<sup>a</sup>; Jordan Ueberroth, MD<sup>a</sup>; Khushali Shah, MD<sup>b</sup>; Codrin Iacob, MD<sup>b</sup>; Bryant Carruth, MD<sup>a</sup>

Department of Ophthalmology, SUNY Upstate Medical University, Syracuse, New York<sup>a</sup>  
Department of Ophthalmology, New York Eye and Ear Infirmary of Mount Sinai, New York, New York<sup>b</sup>

## INTRODUCTION

- Melanotic schwannoma (MS) is a rare neoplasm composed of Schwann cells containing melanosomes in different stages of maturation.<sup>1,2</sup> It is typically observed in spinal nerve roots or cranial nerves,<sup>3,4</sup> and intraocular involvement is uncommon.<sup>5</sup>
- It is often misdiagnosed as malignant melanoma.<sup>6</sup>
- Although previously reported as clinically benign, there may be a propensity for local recurrence and metastasis.<sup>7,8</sup>
- There are no standard guidelines for treating melanotic schwannoma.
- Here, we describe a case of melanotic schwannoma that resulted in a ruptured globe.

## CASE PRESENTATION

A 76-year-old Caucasian woman with multiple vasculopathic risk factors on aspirin daily and no known ocular history was transferred from an outside hospital to Upstate after experiencing acute bleeding from the right eye with complete loss of vision.

Over the previous month, she complained of progressively worsening right-sided headache with right eye pain and redness. There was no recent trauma or eye injury and no prior history of cancer or bleeding diathesis.

During initial examination, the patient was found to have a ruptured right globe with visual acuity of no light perception. Scleral perforation and expulsion of intraocular contents were observed.

Due to severity of injury, the patient underwent enucleation.

## PATHOLOGY REPORT

The specimen showed the following findings:

- An unencapsulated mass of spindle cells in fascicles (Antoni A pattern) with well delineated from the surrounding choroid (Figure 3A).
- Interspersed collagen (Luse bodies) (Figure 3B).
- There was moderate nuclear pleomorphism with intranuclear pseudoinclusions and sparse mitosis.
- Areas of necrosis.
- Bruch's membrane was intact but there was demonstration of extension along the emissary canal of the long posterior ciliary nerve without involvement of the orbital fat (Figure 3C).
- Immunohistochemical stains:
  - Positive: vimentin, S-100, HMB-45, Melan A, reticulin, collagen IV and MITF (Figure 3D)
  - Negative: GFAP, CAM 5.2, neurofilament protein and epithelial membrane antigen.
  - The immunohistochemical stains of the mass were distinctly different from that of the surrounding choroidal melanocytes.

### Final diagnosis:

**Melanotic schwannoma, non-psammomatous type**

## RESULTS

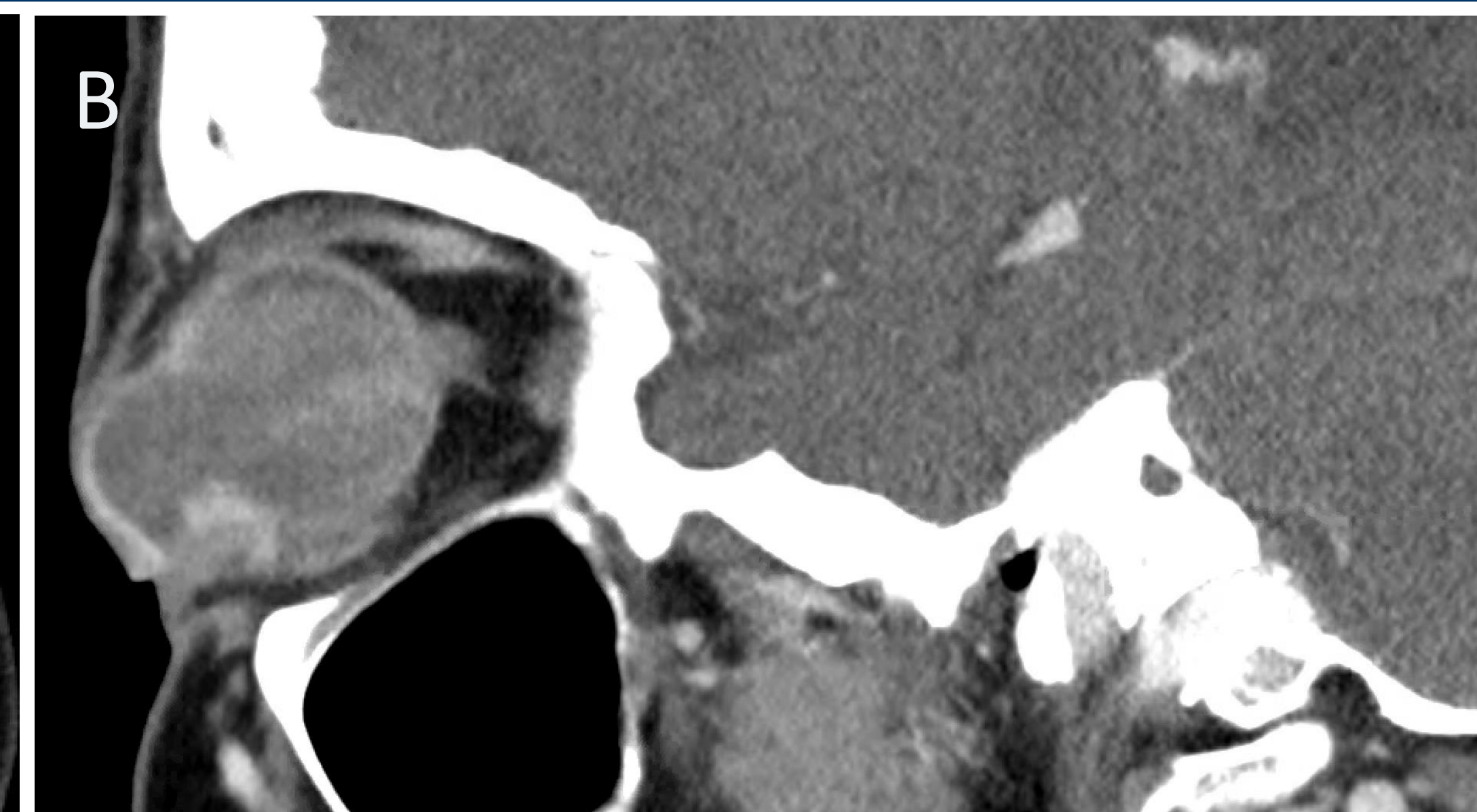
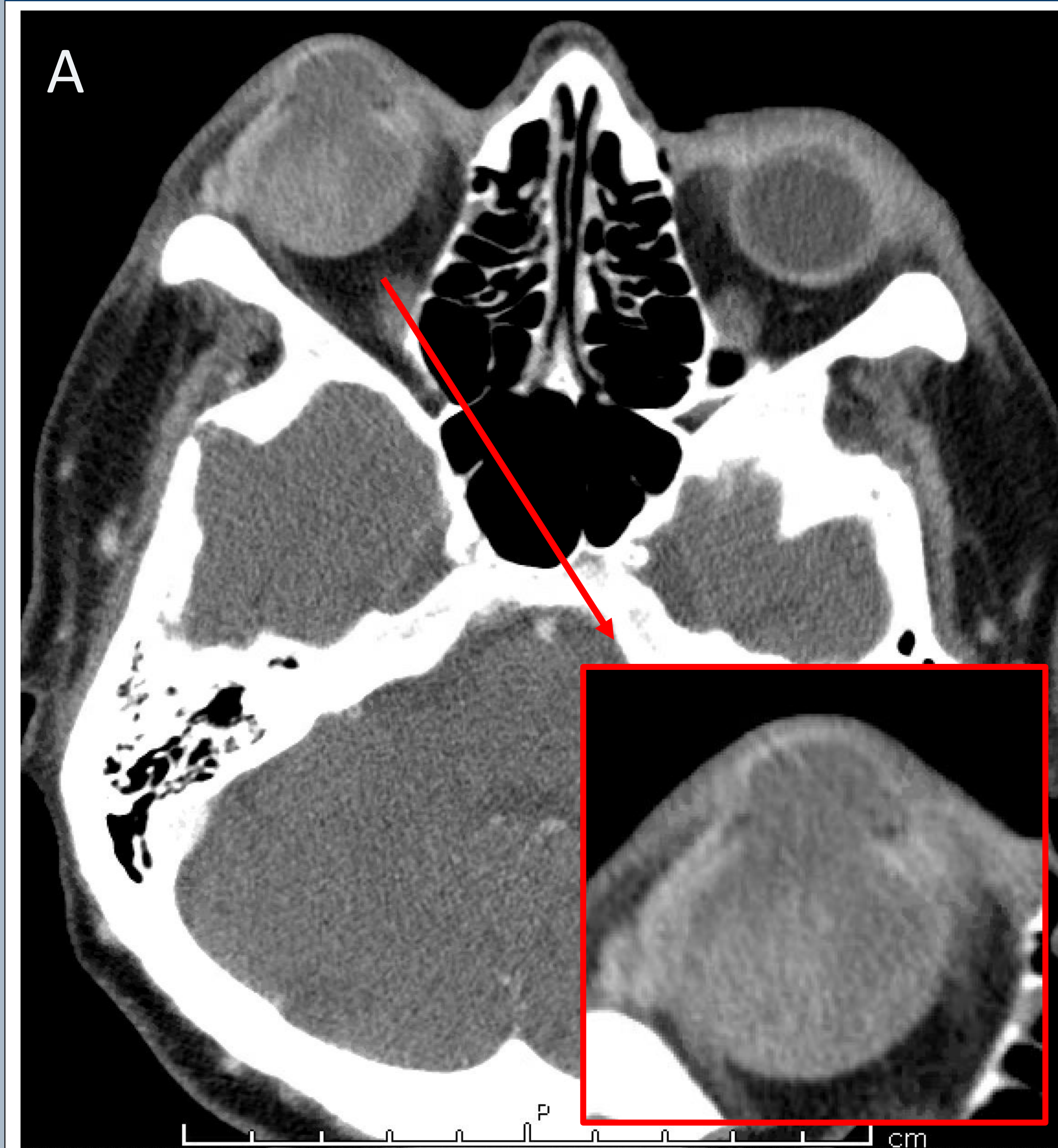


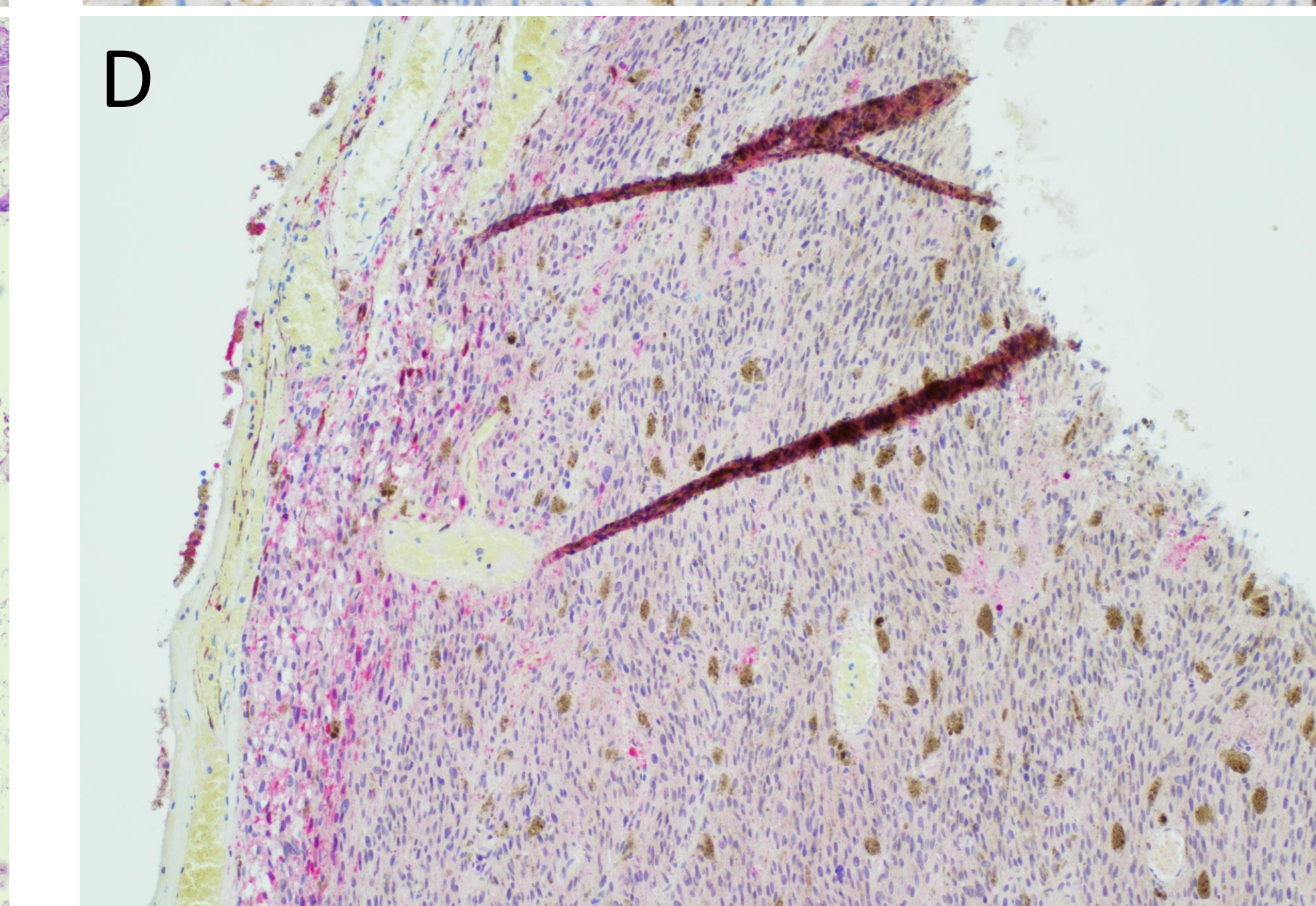
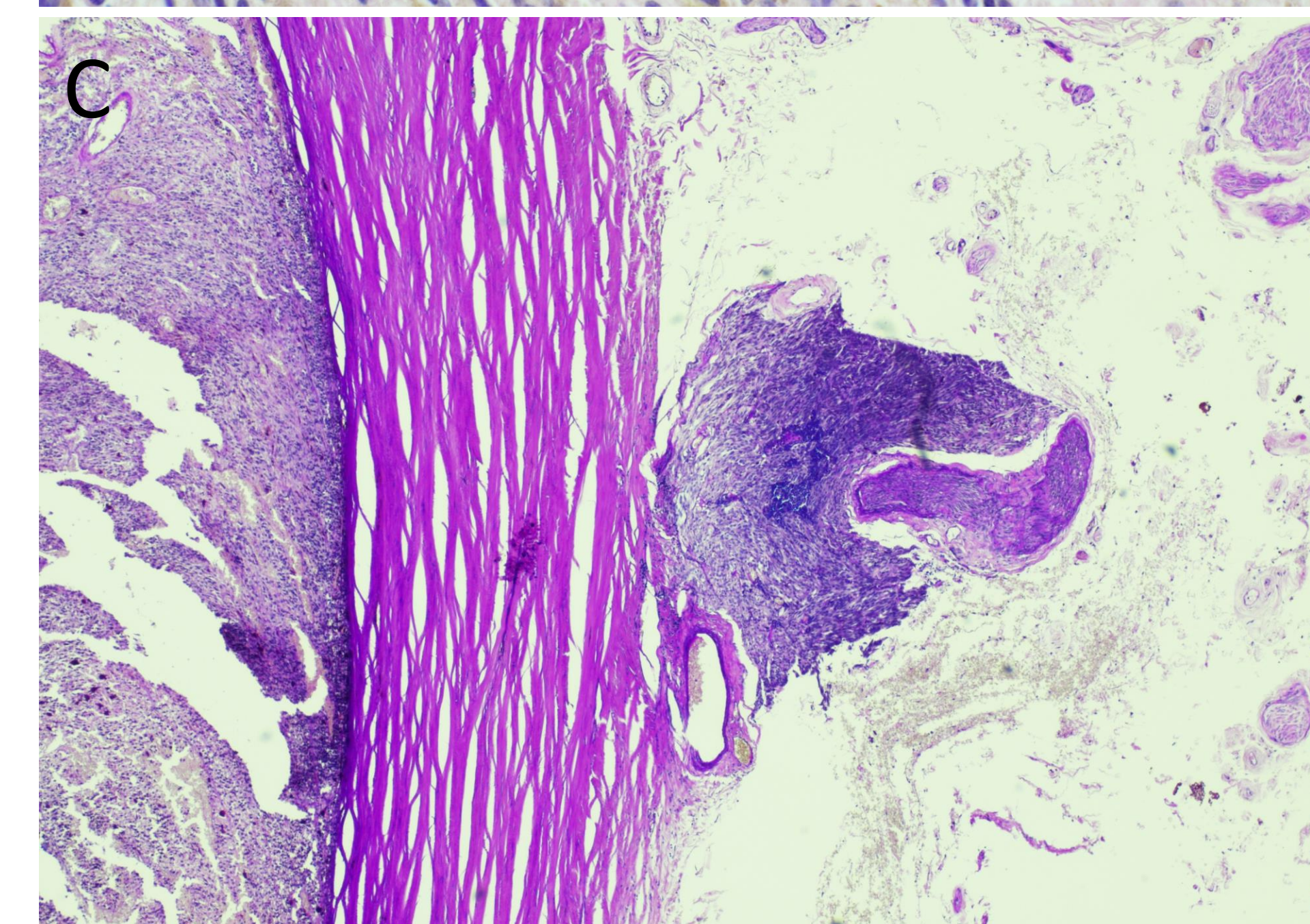
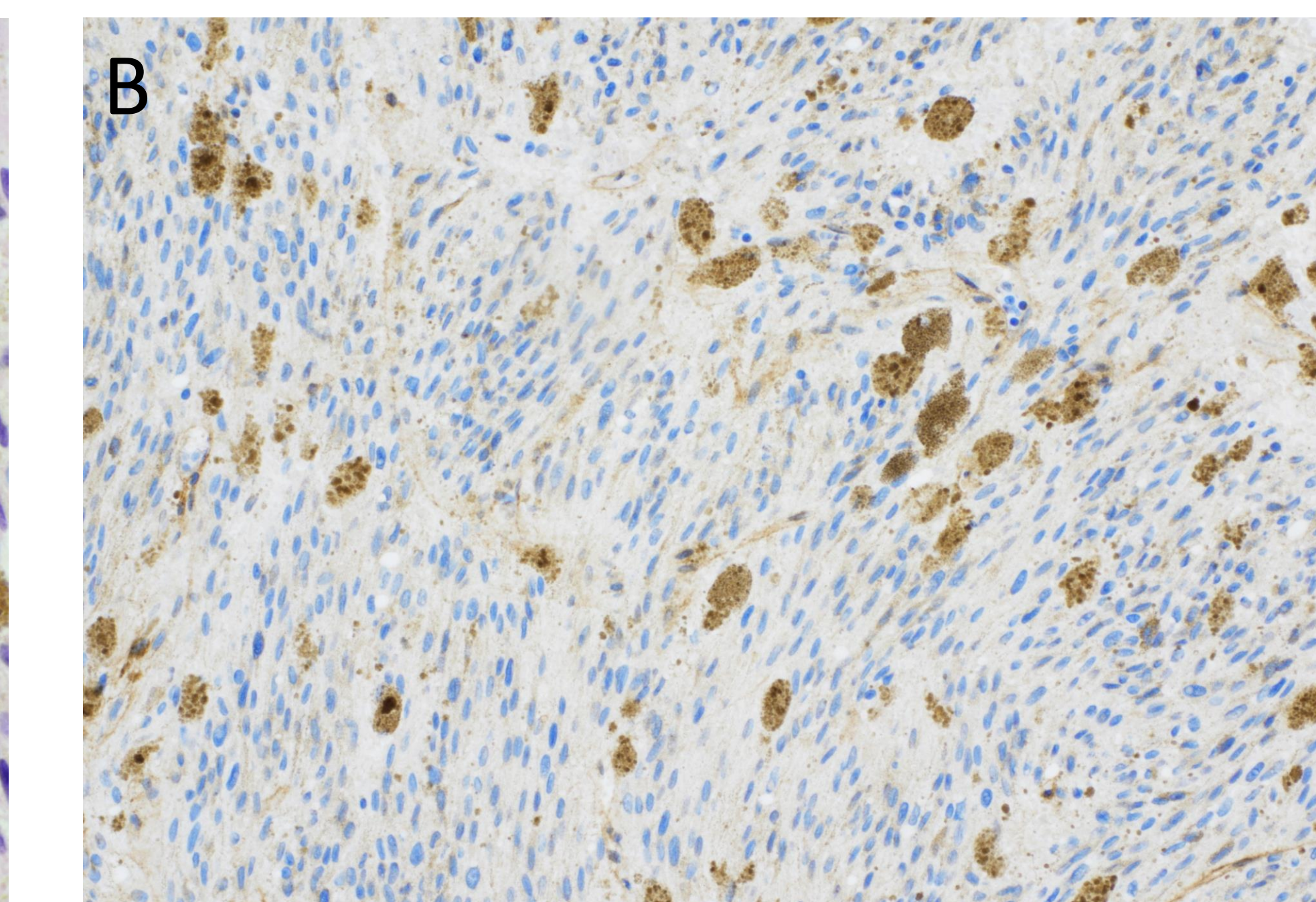
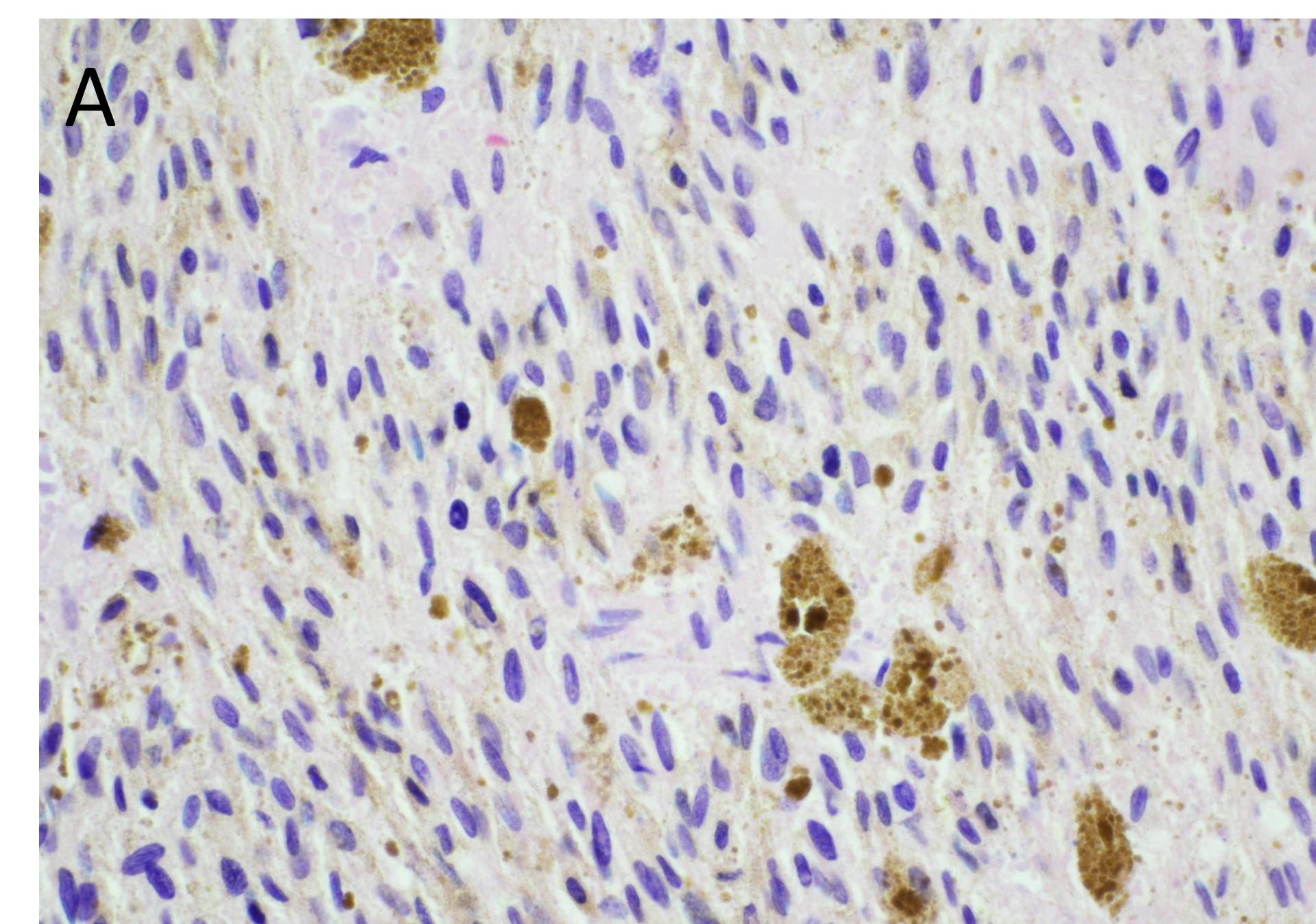
Figure 1. Computed tomography of the orbits.

Hyperdensity of the vitreous body, disorganization of the anterior chamber, and herniation of intraocular contents in the right eye. Axial (A) and Sagittal (B) views.

Figure 2. Clinical photo of the melanotic schwannoma prolapsed from the right ruptured globe. Size 20 mm x 10 mm.



Figure 3. A. Spindle cells in Antoni A pattern [H&E 40x]; B. Luse bodies [Collagen IV 10x]; C. Extrascleral extension along emissary canal of long posterior ciliary nerve [PAS 4x]; D. Tumor immunoreactive for S-100 [S-100 20x].



## DISCUSSION

- Schwannoma (neurilemoma) is a benign peripheral nerve sheath tumor known to rarely affect the globe as an amelanotic choroidal mass.<sup>6</sup>
- MS is well described to affect spinal nerve roots or peripheral nerves in the head or neck.
- MS may be psammomatous or non-psammomatous, with half of psammomatous cases being associated with Carney complex (endocrine abnormality, cardiac myxoma and skin changes).<sup>4</sup>
- Intraocular MS involves the uveal tract and is clinically indistinguishable from choroidal melanoma.
- There are no reported cases of MS causing a ruptured globe.
- The cells had moderate nuclear pleomorphism and low Ki-67 proliferation index of approximately 5%, differentiating from melanoma.<sup>9,10,11</sup>
- The 5-year rate of local recurrence and metastatic spread is 15-35% and 26-44%, respectively.<sup>8</sup>

## CONCLUSION

- We report an uncommon occurrence of melanotic schwannoma which resulted in a ruptured globe.
- Proper diagnosis with light microscopy and immunohistochemistry and close monitoring for local recurrence or metastasis are crucial for preventing a poor outcome.
- Patient outcome: the patient returned for a 2-week post-op visit and was doing well. Her pathology report was received the following week and attempts were made to refer the patient to the oncology service, but she has not been reachable.

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