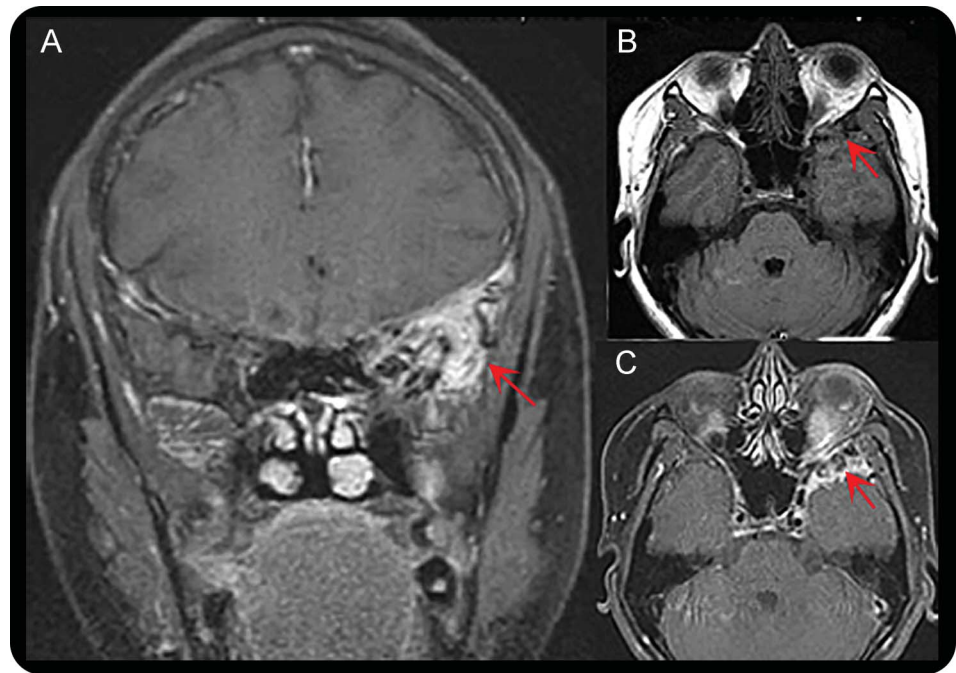


Intracranial epithelioid hemangioendothelioma causing subacute loss of vision

Figure 1 Brain MRI



(A) Coronal T1 with contrast, (B) axial T1 without contrast, and (C) axial T1 with contrast showing left sphenoid mass (arrows).

A 37-year-old woman presented with left eye pain, headaches, and vision loss. MRI showed a left sphenoid mass, with optic nerve compression and proptosis (figure 1). The left eye had minimal reactivity to light, scleral erythema, and proptosis. A metastatic lesion or a lymphoma was suspected; however, pathology showed an epithelioid hemangioendothelioma (figure 2). There was no extracranial disease. Intracranial epithelioid hemangioendothelioma is rare, with around 40 reports. Thirty-two percent show local invasion, mortality is 15%, 24% recur, and 15% metastasize.¹ It is associated with the WWTR1/CAMTA1 fusion protein.² The treatment is surgery, with unclear roles for adjuvant therapy.¹

Jose M. Pacheco, MD, J. Clay Goodman, MD, Jacob Mandel, MD

From the Department of Internal Medicine, Division of Hematology and Oncology (J.M.P.), Departments of Pathology & Immunology and Neurology (J.C.G.), and Department of Neurology (J.M.), Baylor College of Medicine, Houston, TX.

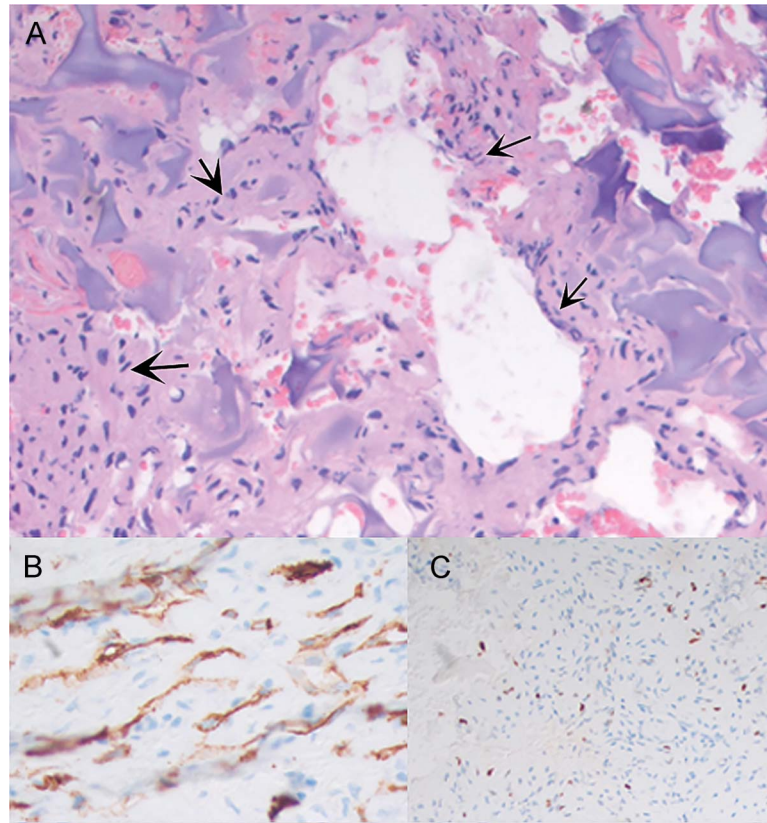
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Correspondence to Dr. Pacheco: jose.pacheco@bcm.edu

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(A) Vascular channels with interspersed spindle cells (arrows). (B) Staining for CD34, one of the characteristic positive stains in this neoplasm. (C) Ki-67 showing low mitotic activity.

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