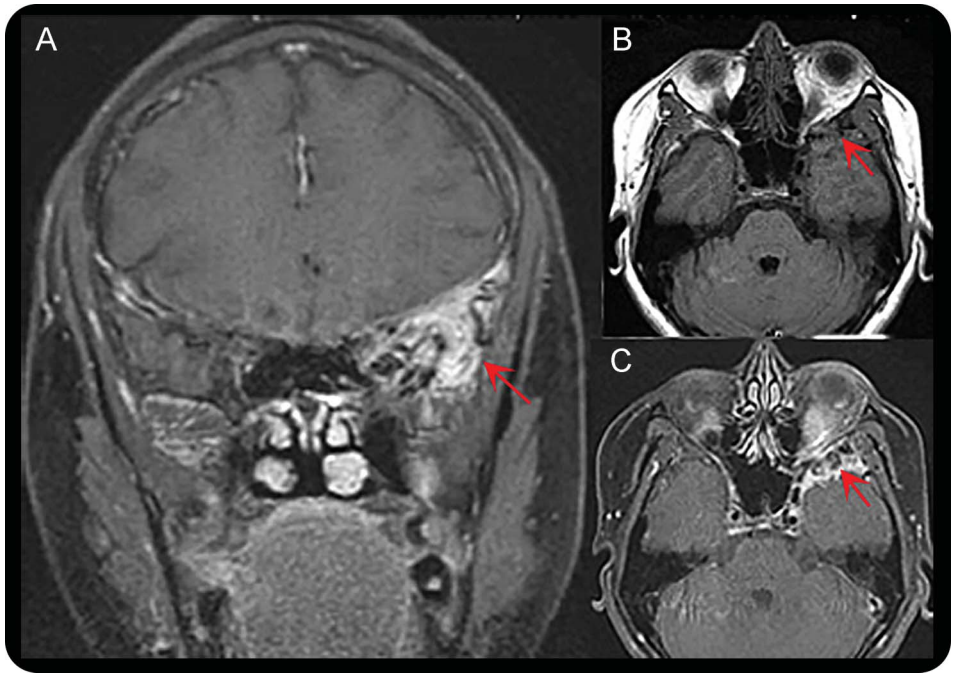
Intracranial epithelioid hemangioendothelioma causing subacute loss of vision

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]

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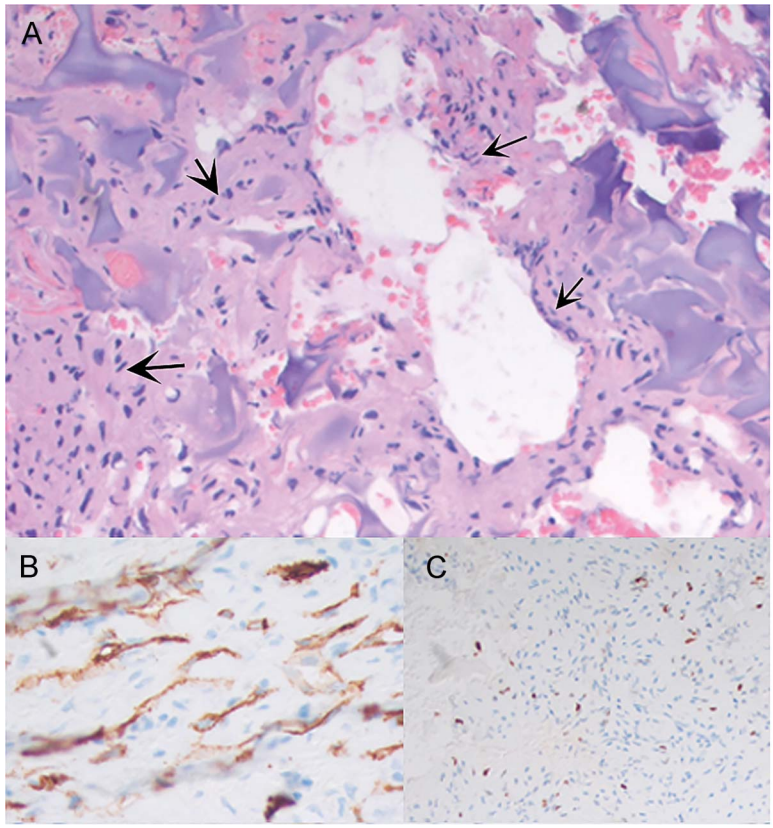
2. Tanas MR, Sboner A, Oliveira AM, et al. Identification of a disease-defining gene fusion in epithelioid hemangioendothelioma. Sci Transl Med 2011;3:98ra82.

Figure 2 Histopathology



(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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