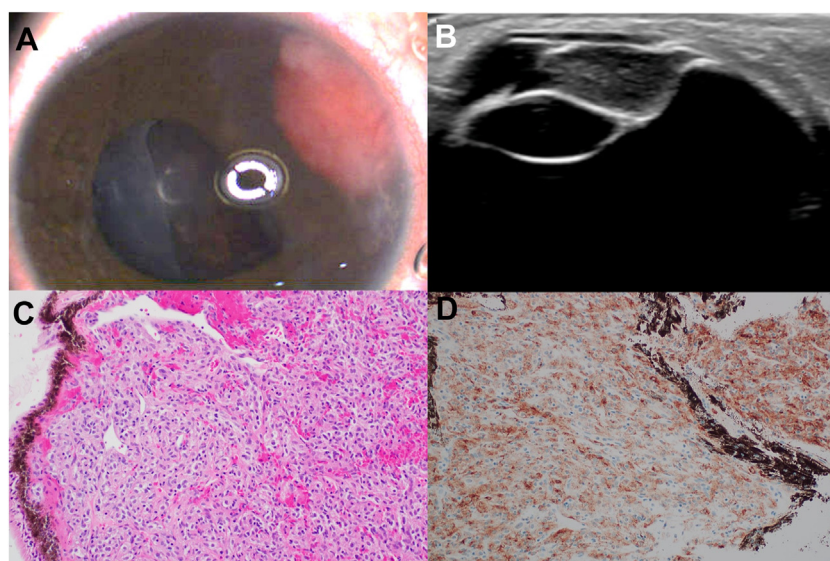


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Pictures & Perspectives



Ciliary Body Perivascular Epithelioid Cell Tumor

A 4-year-old girl presented with an enlarging iris mass. **A**, Examination revealed peripheral amelanotic vascular lesion with a larger retrolenticular portion. **B**, Ultrasound demonstrated a homogenous mass arising from the ciliary body (CB). The patient underwent excisional biopsy through a scleral tunnel incision. **C**, Pathology showed a low-grade neoplasm with plump epithelioid cells. Immunohistochemistry showed diffuse expression of HMB45 immunostain (**D**) and heterogeneous smooth muscle actin (SMA), while being negative for SOX10 and S100. On RNA sequencing *PRCC-TFE3* fusion was detected. Perivascular epithelioid cell tumors (PEComas) are rare benign mesenchymal tumors. *PRCC-TFE3* fusion has been described in choroidal and subconjunctival PEComa, and we demonstrate it with CB PEComa also. (Magnified version of Figure A–D is available online at www.aaojournal.org).

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