A 9-year-old boy had vision impairment in his left eye. Examination of the left eye included visual acuity of questionable light perception, microphthalmos, and a retrolental cyclitic membrane. Examination showed a fleshy, whitish-pink ciliary body mass. B-scan ultrasound confirmed a ciliary body mass with intralesional cysts and total retinal detachment. The right eye had visual acuity of 20/20 and otherwise was unremarkable. Orbital magnetic resonance imaging showed inhomogeneous posterior chamber signals with diffuse, marked, and irregular circumferential thickening of the inner choroid and retinal layer. The left ciliary body was thickened associated with a hypervascular lesion, 7 × 5 × 2.7 mm3 with intense and anomalous enhancement. The clinical and imaging findings were consistent with MECB. Therefore, left eye enucleation was performed. Histological examination showed neoplastic proliferation involving the inner surface of the ciliary body. The neoplasm also had cells with poorly differentiated neuroblastic morphology, arranged in rosettes with increased mitotic activity. Immunohistochemical stains showed positivity for neuron- specific enolase, MNF116 cytokeratins, CK 8/18, weak and focal reactivity for S100, and negativity for acidic glial fibrillar protein. There were no areas of heterologous differentiation nor invasion of the iris, cornea, or sclera. The real-time polymerase chain reaction con- ducted on formalin-fixed paraffin-embedded samples of tumor tissue revealed presence of a mutation in exon 25 (p.E1813D) of the DICER1 gene. The ultrasound examination of the head and neck lymph nodes revealed an enlarged thyroid with an inhomogeneous echogenicity, consisting of various mixed solid-cystic and iso-hyperechoic nodules. Fine-needle aspiration revealed numerous aggregates of thyrocytes in a microfollicular arrangement with a mildly increased nucleus-cytoplasmic ratio. Radiological examinations showed no masses or neoplasms in other organs.