The recent description of the findings of extracutaneous malformations with bilateral grade III microtia, along with a right scalp hemangioma, was first reported in this patient. A review of the literature shows 3 prior reports of similar cases. In 1978, Pascual-Castroviejo described a female patient with right anotia and ipsilateral grade III microtia with a scalp hemangioma (Figure 1). The hemangioma was not present at birth but began to grow when the child was about 2 months of age while the ear deformity was congenital. The patient had a normal left OAE. Sound field testing demonstrated a normal 2D ECHO. Although the precise pathogenesis of HFM is incompletely understood, theories indicate that HFM may develop as a result of inadequate migration of neural crest cells and adherence of the neural crest to the embryo. Leading theory suggests that a genetic defect with mesenchymal disruptions and embryonic hemorrhage could lead to HFM.

Pascual-Castroviejo described a series of patients with hemangioma and ipsilateral grade III microtia and ipsilateral scalp hemangioma in 1996. In the majority of PHACES syndrome patients, a segmental, morphologically diffuse hemangioma of the face is seen. In 7 patients described in a previous report, the hemangioma was excised and an ordered workup was performed. In the majority of PHACES syndrome patients, a segmental, morphologically diffuse hemangioma of the face is seen. In 7 patients described in a previous report, the hemangioma was excised and an ordered workup was performed. In the majority of PHACES syndrome patients, a segmental, morphologically diffuse hemangioma of the face is seen. In 7 patients described in a previous report, the hemangioma was excised and an ordered workup.