A 20-year-old woman presented with a 3-year history of progressive proptosis of the right eye (A). Her father had a history of multipleosteomas. Congenital hypertrophy of the retinal pigment epithelial was not found on retinal examination, except for retinal folds. Multiplewell-circumscribed masses of high radiodensity (B-D), and multiple unerupted supernumerary teeth (C-D) were observed on computedtomography. The osteomas of the right ethmoidal sinus wereremoved due to the threat of the intraorbital structure.