An aneurysmal bone cyst is a rare tumor-like lesion which can affect any part of the skeleton. It is a disease of childhood and adolescence. Reports of its occurrence on the skull base in children are rare. A 22-month-old male patient was admitted to our ENT department with a sudden protrusion of the right eyeball. Radiologically, a cystic, well-defined and contrast enhanced mass on the medial-cranial orbital wall with beginning destruction of the frontal skull base was detected. Histological assessment of a biopsy, which was taken by medial orbitotomy, showed giant-cellular and fibrohistiocytic changes. Definitive histological diagnosis after removal showed an aneurysmal bone cyst. If there is evidence for aggressive, expansive growth, an aneurysmal bone cyst should be included into the ENT-differential diagnosis of orbital tumors. It is not possible to confirm diagnosis from clinical or radiological data. Early biopsy is essential for a reliable diagnosis even if histological assessment is challenging.