Abstract

OBJECTIVE: This article describes our experience with two patients who presented with unusual tumors in the cerebellar vermis and cerebral aqueduct. Although sparing the fourth ventricle proper, both tumors had histological features consistent with the rare diagnosis of a rosette-forming glioneuronal tumor of the fourth ventricle, of which only 19 cases have been reported previously. A review of the clinical features and courses of all 21 cases is presented and management recommendations are given.

CLINICAL PRESENTATION: Patient 1 was a 42-year-old man who presented with a headache of 1 day's duration and no neurological signs, in whom magnetic resonance imaging disclosed a nonenhancing mass lesion occupying the proximal cerebral aqueduct. Patient 2 was a 38-year-old woman with a long history of intermittent giddiness, no neurological signs, and a magnetic resonance imaging scan that demonstrated a nonenhancing and subtle abnormality in the cerebellar vermis.

INTERVENTION: Biopsy was performed on both lesions, the first endoscopically and the second via craniotomy. The only postoperative complication was short-lived double vision and poor upgaze in Patient 1.

CONCLUSION: These cases demonstrate that the rosette-forming glioneuronal tumor may be more accurately categorized as an infratentorial tumor rather than a tumor of the fourth ventricle. Because the literature indicates that this is a tumor with little potential for malignant behavior and considerable morbidity can accompany attempts at resection, a conservative management approach would seem well advised. If this tumor is to be managed conservatively, because of the paucity of extended follow-up data, long-term radiological and clinical surveillance is strongly recommended.