

Case Report

SUPRATENTORIAL HEMANGIOBLASTOMA IN A CHILD

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ABSTRACT

The authors present a 10 year old girl with convulsions refractory to chemotherapy. Neuroimaging revealed a right temporal mass which proved to be a hemangioblastoma upon its total removal.

Key Words: Hemangioblastoma, Supratentorial, von-Hippel Lindau

INTRODUCTION

Hemangioblastomas are primarily tumors of the cerebellum. They are known to occur in association with von Hippel-Lindau disease. Supratentorial localization for this tumor is rare and less than 90 such cases have been reported in the literature. Supratentorial hemangioblastoma in children is even rarer; so far only six children 11 years old or younger have been reported (1-6).

CASE REPORT

This 10 year old girl was admitted because of uncontrolled epileptic seizures. She had been taking 200 mg/day diphenylhydantoin for the last year. The cranial computerized tomography (CT) performed at another center demonstrated a right temporal lobe mass lesion measuring 2.7 x 2.3 cm. It had regular contours with dense enhancement except the posteromedial region (Fig. 1).

The cranial magnetic resonance (MR) showed heterogenous hypointense mass on T1W images and hyperintense on T2W images (Figs. 2a and 2b). The mass was supratentorial and enhanced diffusely (Fig. 2c).

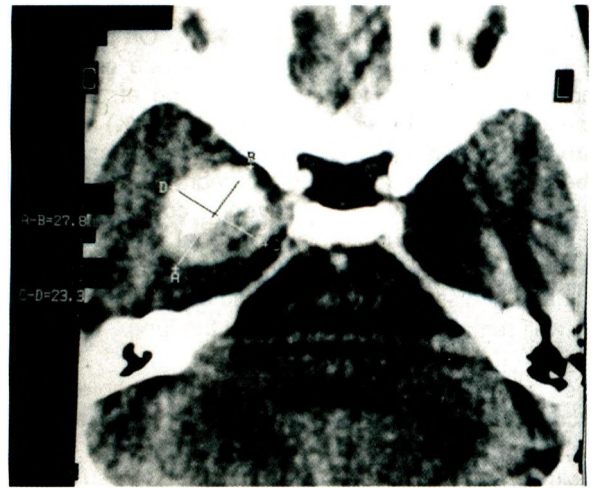
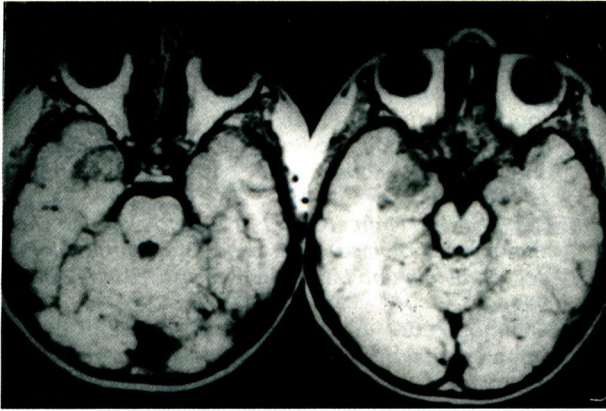
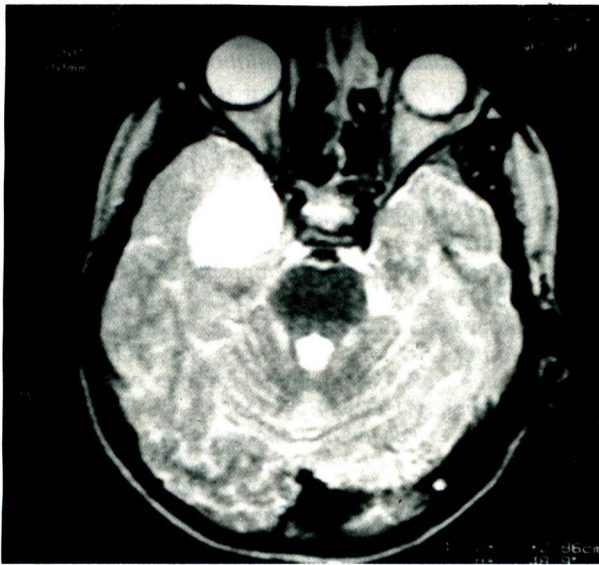


Fig.1: Postcontrast transverse CT revealed an enhancing mass within the right temporal lobe. See dense enhancement, hypodense region posteromedially.

The physical and neurologic examinations were normal. The patient was operated on June 13, 1996. A right frontotemporal craniotomy was performed. Following transsylvian dissection, a cortical incision of one cm. was made at uncus. The pathology was reached in 2 mm. depth of the incision. The tumor was grayish in color. It did not show a clear cleavage. It was remarkably vascular, but moderate hemorrhage occurred. There was no macrocyst. The tumor measuring approximately 3x2.5x2.5 cm. was totally removed. Postoperative period was uneventful. Diphenylhydantoin was continued at the dose of 200 mg/day and she did not experience any epileptic seizures postoperatively. The histopathologic diagnosis was hemangioblastoma. Abdominal ultrasound and ophthalmologic examination performed upon this diagnosis were normal. The family history was negative. A cranial CT performed 24 hours postoperatively and cranial MR three months



a



b



c

postoperatively were insignificant. MR performed six months postoperatively revealed a recurrent mass smaller than one cm. The patient was kept under control. The last cranial MR on September 1997 showed that the mass had reached its preoperative dimensions. Meanwhile she had no complaints. Papilledema was noted on neurologic examination. The patient was reoperated on September 10, 1997 with total removal of the mass. There were no seizures and there was no recurrence during the follow-up of one year.

DISCUSSION

Hemangioblastomas constitute 1.5 to 2 % of all intracranial tumors and account for 7 to 12 % of posterior fossa tumors. They are benign tumors, malignant spread is rare. The cerebellum is the hemangioblastoma's most frequent site, but hemangioblastoma can also be found in the area postrema of the medulla or within the spinal cord, particularly in those patients with von-Hippel Lindau. Supratentorial hemangioblastomas are rare. Since its first description in 1902 less than 90 supratentorial hemangioblastomas have been reported in the literature, including localizations like pituitary fossa, ventricles and leptomeninges. Supratentorial hemangioblastoma occurring in children is extremely rare. The presented patient is the seventh case of a supratentorial hemangioblastoma occurring in a child 11 years old or younger (1-6). Macroscopically hemangioblastomas of the cerebellum are pinkish or yellow in color. They are associated with a macrocyst in 70 % of the cases with a mural nodule within the cyst (7). Solid cerebellar hemangioblastomas have also been reported. Entirely solid hemangioblastomas occur in 30 to 40% of the cases and are the most common morphologic type if in the supratentorial compartment. The distinction of solid hemangioblastoma from either renal cell carcinoma metastasis or another lesion of concern in von Hippel-Lindau patients, or angioblastic meningioma may at times be extremely difficult on histopathology. The cystic nature of the mass, a peripheral, pial based mural nodule of solid tissue which enhances markedly with IV contrast and large vessels within and/or the periphery of the mass are the most important features. Small or supratentorial hemangioblastomas may be

Fig.2: Transverse short TRITE images demonstrated 2.5 x 2.8 cm mass in the right temporal region. The mass is heterogeneously hypointense relative to the brain tissue (a). T2 W image, the mass is homogeneously hyperintense (b). Gd-DPTA administration showed homogenous enhancement, implying the solid nature of the mass (c).

completely solid and enhance. CT findings of the presented patient were generally consistent with the findings in the literature except for the presence of a macrocyst. CT reveals a tumor density similar to the brain, homogenous enhancement after contrast and little perilesional edema.

Intratumoral hemorrhage can occur occasionally in association with hemangioblastoma. Even in seemingly total removal of the tumor 3 to 10% recurrence rates have been reported. In such situations the recommended therapy is reoperation and multiple interventions are possible if necessary (7). The reason for recurrence, which we also experienced, is not clear. The operative mortality has dropped to 0 % (8) from the 25 to 30% mortality rates of the 1940's (9). Postoperative deaths, in most cases, are associated with posterior fossa or a supratentorial hematoma or with brain stem infarction.

The beneficial effects of conventional radiotherapy in local control of incompletely resected hemangioblastomas has been reported (10).

Stereotactic radiosurgery has helped in volumetric reduction of the tumor mass (11), in control of the progression of the tumor (12) and has contributed to the complete disappearance of an intrasellar hemangioblastoma which was subtotally resected and received conventional radiotherapy postoperatively.

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