Supratentorial Leptomeningeal Hemangioblastoma Mimicking A Meningioma Without Von Hippel-Lindau Complex

Von Hippel-Lindau Kompleksi Olmadan Meningiomayı Taklit Eden Bir Supratentorial Leptomeningeal Hemanjioblastoma Olgusu

Ömer L. İYİGÜN[1]
Cengiz ÇOKLUK[1]
Kerametin AYDIN[1]
Levent YILDIZ[2]
Cemil RAKUNT[1]
Fahrettin ÇELİK[1]

1 Department of Neurosurgery, School of Medicine, Ondokuzmayıs University, Samsun, Turkey
2 Department of Pathology, School of Medicine, Ondokuzmayıs University, Samsun, Turkey

ABSTRACT

We present a 61-year-old man with a 3-month history of headache, hemiparesia and speech disturbance. Computerized tomography (CT) of the brain showed well defined, diffusely contrast enhanced and frontoparietally located solid mass. Histopatologic diagnosis of the tumour was hemangioblastoma. In this report, we present clinic, radiologic and histophologic characteristics of the patient and discussed the case under the light of the literature.

KEY WORDS: Supratentorial hemangioblastoma, Von Hippel Lindau syndrome, brain tumour.

ÖZ

Üç aydır konuşma bozukluğu, hemiparezi, başağrısı hikayesi ile gelen 61 yaşında erkek hastayı sunduk. Çekilen CT'de iki sınırlı diffüz kontrast tutan ve frontoparietal lokalizasyonlu solid kitlesi mevcuttu. Tümör total çıkarıldı. Histopatolojik tanısı hemanjioblastoma olarak değerlendirildi. Bu vaka sunumunda hastanın klinik, radyolojik, histopatolojik karakteristikleri literatür eşliğinde tartışıldı.

ANAHTAR SÖZCÜKLER: Supratentoral hemaonjioblastoma, Von Hippel Lindau sedromu, beyin tümörü.

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Correspondence address:
Kerametin AYDIN
Ondokuzmayıs University Medical Faculty
Department of Neurosurgery
55139 Samsun, Turkey
Tel : +90 362 4576000/3652 int
Fax: : +90 362 4576041
e-mail : kaydin@omu.edu.tr

INTRODUCTION

Hemangioblastomas are histologically benign and rare neuroaxial tumors (2, 3, 5, 6, 7, 8, 9). They may occur sporadically or as multiple tumors in Von Hippel Lindau's (VHL) disease (1, 6, 8) These tumors are most commonly localized within the cerebellum and less commonly occur in the medulla oblongata or spinal cord, and rarely in the supratentorial compartments (2, 6, 8). Hemangioblastomas have a tendency to be cystic lesions with a mural nodule. Pure solid tumors are less common than cystic tumors (8).

In this report, a patient with a supratentorial solid hemangioblastoma is presented and the relevant literature is reviewed.

CASE REPORT

A 61-year-old man was admitted to our neurosurgical department because of a three-month history of headache and fifteen days onset of hemiparesia and speech disturbance. His personal history denoted a hospitalization because of suspected cerebrovascular disease eight years ago. Hematological values were within the normal physiological ranges. CT scan of the brain showed a well circumscribed, diffusely contrast-enhanced and solid right frontoparietal leptomeningeal tumor mimicking a meningioma with dense peritumoral edema and midline shift (Figure 1). The patient was operated on by standard craniotomy under general anesthesia. The purplish-colored and well-defined mass was removed totally by using microsurgical technique under the operating microscope. The hemiparesia and speech disturbance continued in the postoperative period without progression.

The histopathological features were large vacuolated stromal cells and a fine capillary network. Stromal cells varied in size and had hyperchromatic nuclei. Some stromal cells revealed clear cell morphology when stained with oil-red-o for lipids. A single row of endothelial cells lined the capillary network and these cells stained immuno-histochemically with CD31.

The histopathological diagnosis was hemangioblastoma (Figure 2).

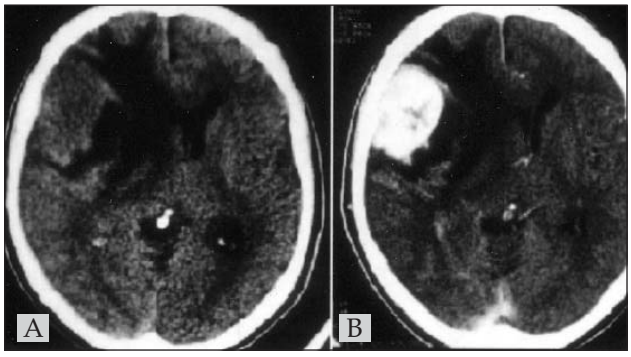

Figure 1. CT studies of the brain.
A. Non-contrast enhanced CT revealed right solid mass with perilesional edema and midline shift.
B. Contrast-enhanced CT scan of the brain showed well circumscribed, diffusely contrast-enhanced and solid right fronto-parietal leptomeningeal tumor mimicking a meningioma with dense peritumoral edema and midline shift

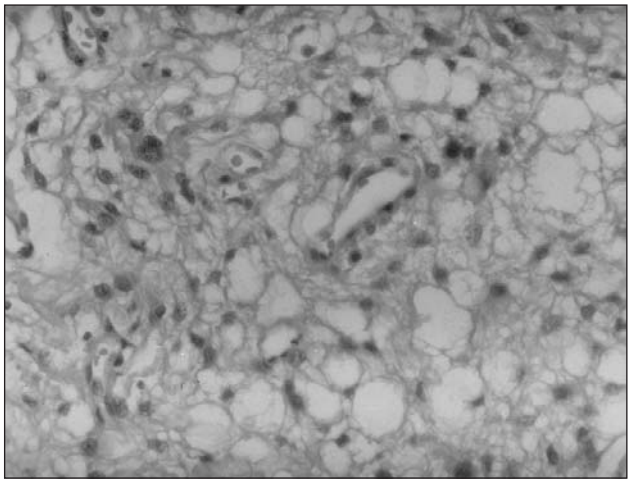

Figure 2. The histopathological diagnosis was hemangioblastoma
A. Vacuolated stromal cells and fine capillary network (X200 HE)
B. Large, clear, cytoplasmic cell in the tumor area (X400 HE)

DISCUSSION

Hemangioblastomas localized outside the posterior fossa are rarely encountered in neurosurgical practice (2, 3, 5, 6, 7, 8, 9). These tumors generally occur within the cerebellum, comprising 7-12% of all posterior fossa tumors (6). Solitary or multiple supratentorial hemangioblastomas are distinctly rare and can be associated with VHL disease as a part of the disease complex. The family history may be positive in 4-20 % of cases (1, 6, 8).

Hemangioblastomas may occur in the brain parenchyma of all lobes, and in the corpus callosum, ventricles, choroid plexus and leptomeninges (2, 4, 6, 10). Leptomeningeal location of the tumor is extremely rare and accounts for 5.6% of supratentorial cases (8).

Pure solid hemangioblastomas are rare in supratentorial and infratentorial locations (8). Two third of these tumors have a cystic component and the remaining tumors show pure solid characteristics (6,8).

The radiological features of supratentorial lesions may closely resemble those of hemangioblastoma in the infratentorial fossa (6).

CT and magnetic resonance imaging can easily detect supratentorially and infratentorially located tumors, and expose some characteristics such as whether the tumor is pure solid, cystic or mural nodule, and any peritumoral edema. On contrast-enhanced CT, the solid part, mural nodule and cyst wall may hold the dye diffusely. On an unenhanced scan, the mural nodule or the solid part of the tumor is isodense to brain tissue. Peritumoral edema is not common in these tumors. Calcification has not been reported (6).

CT appearance of the leptomeningeal supratentorial solid hemangioblastomas resembles meningiomas.

The goal of this report is to describe some uncommon features of supratentorial hemangioblastomas. Supratentorial leptomeningeal solid hemangioblastomas are extremely uncommon in the medical literature. The leptomeningeally located hemangioblastomas show diffuse contrast enhancement on CT and imitate the radiological characteristics of similarly located meningiomas. It is therefore fairly difficult to distinguish these tumors from meningiomas.

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