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Frontal Lobe Ependymoma: A Case Report

Frontal Lob Yerleşimli Ependimoma: Olgu Sunumu

CASE REPORT
OLGU SUNUMU

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ABSTRACT
ÖZET

A 15-year-old female patient with no previous health problems was admitted our clinic with syncope. Cranial computed tomography of the patient showed a 6.5x3.5 cm mass lesion with heterogeneous weak hyperdensity in the left frontoparietal region adjacent to the internal tabula. There was a hypodense zone of edema around the lesion. Following contrast medium injection, significant contrast enhancement was observed. In the anterior portion of the body of the left lateral ventricle and in the left frontal horn, we found obliteration which was secondary to the edema. The mass was excised by left frontotemporal craniotomy. Histopathological findings were found to be consistent with ependymoma, WHO Grade III. We discuss here a case with a diagnosis of an ependymoma with an extra-axial location and anaplastic histomorphology, in the light of current literature.

Key words: Frontal lobe, brain tumor, primary, anaplastic ependymoma

Daha önce sağlıklı olan 15 yaşındaki kadın hasta senkop belirtileri ile kliniğimize başvurdu. Kraniyal tomografide sol frontopariyetal bölgede internal tabulaya komşuluk gösteren heterojen zayıf hiperdens yapıda 6,5x3,5 cm boyutlarında kitle lezyonu tespit edildi. Lezyon çevresinde hipodens ödem sahası vardı. Kontrast madde enjeksiyonu sonrası belirgin kontrast madde tutulumu izlendi. Sol tarafta lateral ventrikülün gövde kesiminin anterior kısmında ve sol frontal hornda ödeme sekonder obliterasyon görüldü. Sol frontotemporal kraniyotomi ile kitle eksize edildi. Histopatolojik bulgularla olgu clear cell ependymoma WHO Grade III olarak değerlendirildi. Burada ekstra aksiyal yerleşimli ve anaplastik histomorfolojili ependimoma tanısı alan bir olgu güncel literatür ışığında tartışılacaktır.

Anahtar kelimeler: Frontal lob, beyin tümörü, primer, anaplastik epandimom

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Introduction

Ependymomas are central nervous system tumors arising from the ependymal cells lying on the inner surface of brain ventricles and along the central spinal canal. Ependymomas of supratentorial location account for 30-50% of all intracranial ependymoma cases. Of the supratentorial ependymomas, approximately 50% are hemisphere-located and have no connection with the ventricular system (1). The total extra-axial ependymoma phenomenon is quite rare (2).

Ependymomas account for 2-9% of all intracranial tumors. They are tumors typically arising from the ventricular system or the central canal's ependymal surface. They are more common in children and are of infratentorial location. In 60-70% of the cases, the tumor is located in the posterior fossa or the fourth ventricle. In adults, however, 33% of the cases are of infratentorial and 66% are supratentorially located (3). Supratentorial cortical ependymoma is a rare clinical entity where the ependymoma occurs in the cortex without any connection to the ventricular system.

There have been 15 such cases reported in the literature. We report the second case of a supratentorial extra-axial cortical anaplastic ependymoma with minimal cortical attachment in a 15-year-old girl who presented with syncope (4).

Case Report

A 15-year-old female patient with no previous health problems was admitted our clinic with syncope. A 6.5x3.5 cm mass lesion of heterogeneous weak hyperdense structure was detected in the left frontoparietal region adjacent to the internal tabula by cranial computed tomography. There was a hypodense zone of edema around the lesion. Following contrast medium injection, significant contrast enhancement was observed. In the anterior portion of the body of the left lateral ventricle and in the left frontal horn, we found obliteration which was secondary to the edema.

MR imaging revealed a lobular-contoured 42x52x60 mm solid tumoral mass lesion in the left frontal lobe with cortical, subcortical, and deep white matter involvement and surrounding edema. The left lateral ventricle was depressed due to a mass effect, compressing the adjacent cerebral cortical sulci, and showing hypointensity in

T1-weighted images, hyperintensity on T2-weighted images, and dense contrast enhancement following contrast material injection (Figure 1, 2).

The mass was excised macroscopically with left frontotemporal craniotomy. The postoperative period was free of any complications. A relatively smooth-edged medium-sized cellular tumor, infiltrating into the adjacent brain parenchyma, was observed in the histopathological study of the patient's operative materials. Although showed minimal nuclear atypia, the tumor cells were characterized by a clear perinuclear halo and had an appearance similar to that of oligodendroglioma. Widespread perivascular pseudorosette proliferation was notable in the tumor (Figure 3). We observed necrosis without palisading and microvascular proliferation in focal areas. There was marked mitotic activity, but the Ki-67 proliferation index was about 30%. Tumor cells showed strong and widespread glial fibrillary acidic protein (GFAP) expression (Figure 4). Epithelial membrane antigen (EMA) and cytoplasmic "dotted-line" staining were present (Figure 5). Cytoplasmic staining was observed to be widespread with vimentin staining, but was limited to a number of cells with S-100 staining. Pancytokeratin or synaptophysin expression was not present. Ki-67 proliferation index was 30% (Figure 6). These findings were considered to be consistent with anaplastic clear cell ependymoma, WHO Grade III. No residue was found in the control MR after a month. The patient had cranial radiotherapy and the follow-up is continuing.

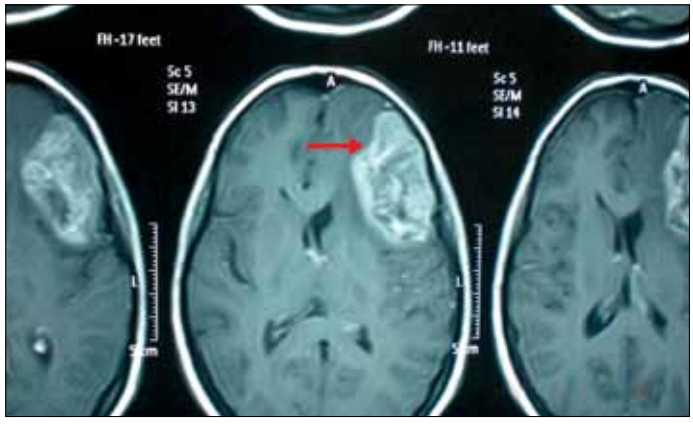

Figure 1. On the right, mass lesion markedly heterogeneous contrasted on the lobe contour following intravenous contrast material injection by which widespread hypointense edema was observed around the white material in the cortical subcortical area in the frontal lobe

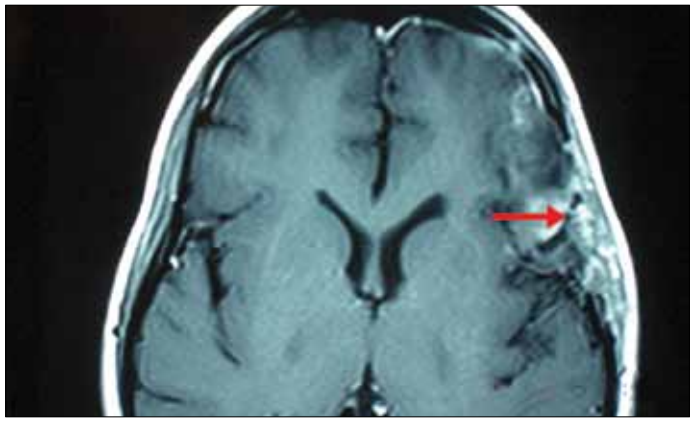

Figure 2. On the right, hyperintense signal changes belonging to the postoperative ependymoma in the frontal lobe, with a marked decrease in edema and mass effect (no contrast enhancement was observed related to residual tissue following intravenous contrast material injection

Discussion

There are four pathological subtypes of ependymoma, designated as myxopapillary ependymoma (WHO grade I), subependymoma (WHO grade I), ependymoma (WHO grade II), and anaplastic ependymoma (WHO grade III) (5). Ependymal rosettes and perivascular pseudorosette structures are the main histological findings. Perivascular pseudorosettes are formed by tumor cells radially surrounding blood vessels and are seen in a majority of ependymomas. True ependymal rosettes are formed by columnar cells arranged around a central lumen. Although these structures are diagnostic for ependymoma, they are found only in a minority of cases.

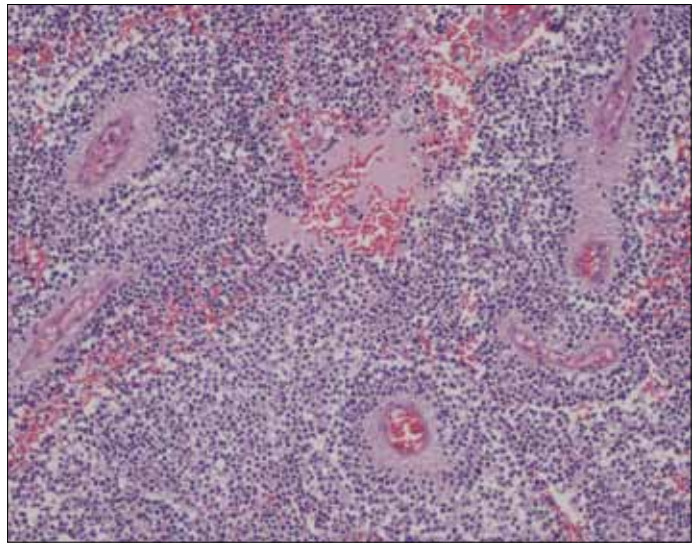

Figure 3. Perivascular pseudorosette structures and tumor cells with clear cytoplasm (H&E x100)

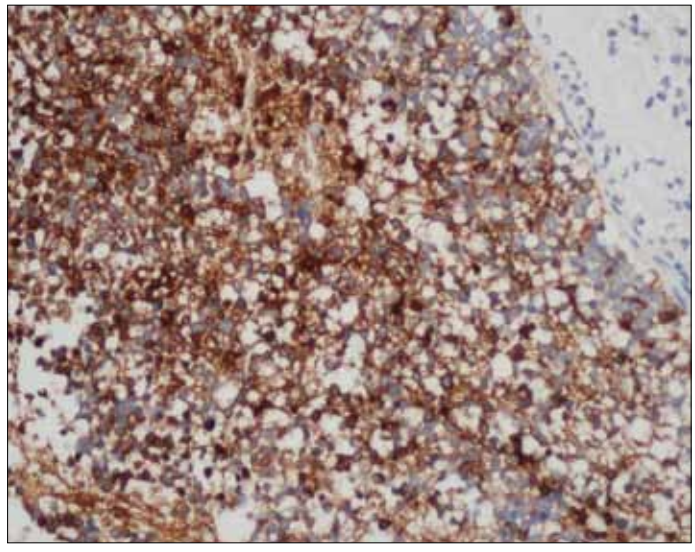

Figure 4. Strong and widespread GFAP positivity (GFAP x100)

Table 1. Supratentorial ependymomas without attachment to the ventricular system

Author et. al.	Year	Age	Sex	Pathology/Location	References
Hayashi et. al.	1994	13	M	Clear cell/parietooccipital	7
Vernet et. al.	1995	11	F	Ependymoma/frontal	8
Fujimoto et. al	1997	13	M	Clear cell/frontal	9
Saito et. al.	1999	63	F	Cellular/parietal	10
Sato et. al.	2000	41	F	Ependymoma/frontoparietal	11
Takeshima et. al.	2002	70	F	Anaplastic/frontal	12
Kojima et. al.	2003	56	F	Anaplastic/temporoparietal	13
Lehman et. al.	2003	10	F	Ependymoma/frontal	14
Moritani et. al.	2003	50	F	Anaplastic/temporal	15
Ono et. al.	2004	6	M	Ependymoma/frontal	16
Roncaroli et. al.	2005	52	M	Ependymoma/frontal	17
Roncaroli et. al.	2005	34	M	Ependymoma/temporal	17
Roncaroli et. al.	2005	11	F	Ependymoma/parietal	17
Miyazawa et. al.	2007	32	M	Anaplastic/parietal	6
Park et. al	2010	17	F	Anaplastic/frontoparietal	18

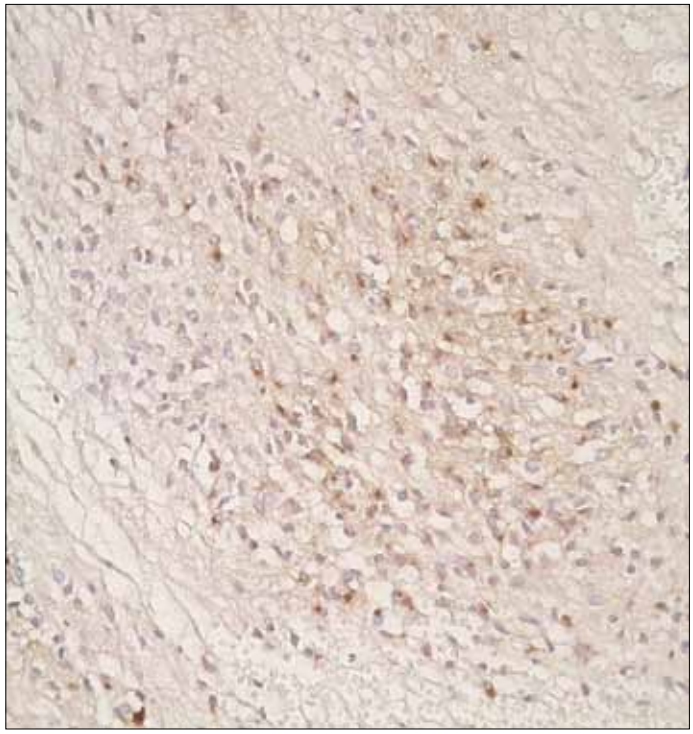

Figure 5. Epithelial membrane antigen showed cytoplasmic "dotted-line" staining (EMA x200)

Anaplastic ependymomas, on the other hand, are characterized by increased cellularity and marked mitotic activity. Microvascular proliferation and pseudopalisading necrosis are frequently seen. While perivascular pseudorosette structures are frequently found, true rosette structures are either infrequent or non-existent. Anaplastic ependymomas tend to remain well-defined and may sometimes be quite invasive. In the anaplastic type, metastasis through the CSF pathway is quite frequent. Ependymomas that are slow-

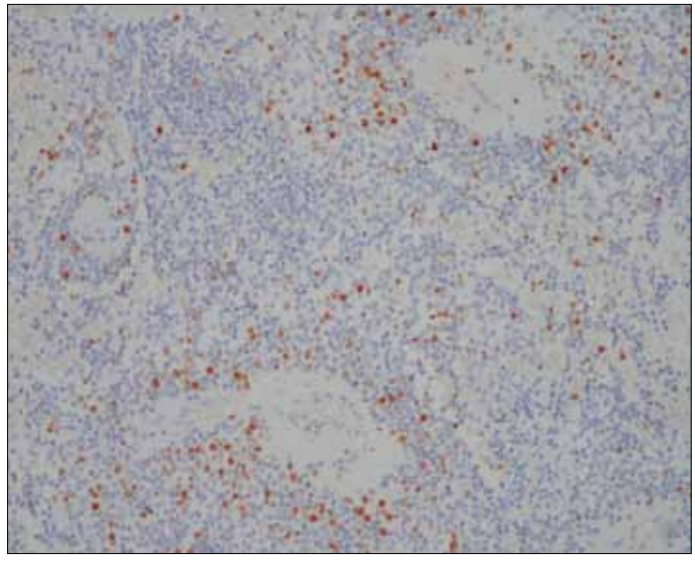

Figure 6. Neoplastic cells show high proliferation index with Ki-67 (Ki-67 x100)

growing and rarely show anaplastic changes have up to an 80% probability of recurrence. The case we have presented here is of an extra-axial location in the frontal lobe and is of anaplastic histology. It is the sixteenth case of a cortical supratentorial location and the sixth of a supratentorial location with anaplastic histology reported in the literature (Table 1) (6).

The radiological appearance of ependymomas is in the form of a smooth-edged mass. It may be accompanied by a cystic component and may present contrast in varying degrees. Hydrocephaly is frequent. Metastasis through seeding along the CSF pathway is frequently encountered and this condition is widespread in the anaplastic form. Resection is the primary mode of treatment. If no residue is detected in the postoperative control MRI and CSF

samples are free of malignant cells, radiation therapy is applied in limited areas.

Conclusion

We have presented here a case of anaplastic ependymoma of a supratentorial location treated by total excision. No residue was detected in the control MRI. Cranial radiation therapy was applied. Although supratentorial intraparenchymal ependymomas are rare occurrences, we are of the opinion that this pathology must be kept in mind in the differential diagnosis of intracranial masses.

Conflict of interest
No conflict of interest was declared by the authors.

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Authors' contributions: Conceived and designed the experiments or case: MY, DS. Examination and follow-up of the patient: UDD, EP, VU, SG, CT. Analysed the data: MY. Wrote the paper: MY, DS, UDD. All authors have read and approved the final manuscript.

Çıkar Çatışması
Yazarlar herhangi bir çıkar çatışması bildirmemişlerdir.

Hakem değerlendirmesi: Bağımsız hakemlerce değerlendirilmiştir.

Yazar katkıları: Çalışma fikrinin tasarlanması: MY, DS. Hastanın muayenesi ve takibi: UDD, EP, VU, SG, CT. Verilerin analizi: MY. Yazının hazırlanması: MY, DS, UDD. Tüm yazarlar yazının son halini okumuş ve onaylamıştır.

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