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Supratentorial primitive neuroectodermal tumor of the frontal lobe in a teenager

İsmail Gülşen[1], Hakan Ak[2]*, İlker Alaca[3], Mehmet Deniz Bulut[4], Alper Can[5], İrfan Bayram[6]
[1] YüzüncüYıl University, School of Medicine, Department of Neurosurgery, Van/TURKEY
[2] BozokUniversity, School of Medicine, Department of Neurosurgery, Yozgat/TURKEY
[3] Yüksekova State Hospital, Hakkari/TURKEY
[4]YüzüncüYıl University, School of Medicine, Department of Radiology, Van/TURKEY
[5]YüzüncüYıl University, School of Medicine, Department of Oncology, Van/TURKEY
6 YüzüncüYıl University, School of Medicine, Department of Pathology, Van/TURKEY

***Corresponding author**
Hakan Ak
Email: nrsdrhakanak@yahoo.com

Abstract: Primitive neuroectodermal tumors occur predominantly in children and adolescents. However, they represent less than 2.5% of childhood brain tumors. These tumors show aggressive clinical behavior and their general prognosis is poor. A 15-year-old girl was admitted with a headache lasting about 20 days and her radiological images showed a well-demarcated tumor with heterogeneous enhancement after gadolinium-diethylene triamine penta acetic acid (Gd-DTPA) administration in the left frontal lobe. Pathological diagnosis was concordant with supratentorial primitive neuroectodermal tumor. After surgical decompression, chemotherapy and radiotherapy were given. Patient is still under clinical follow up without recurrence. Surgery, chemotherapy, and radiotherapy compromise the main treatment of this pathology.
Keywords: supratentorial primitive neuroectodermal tumor, medulloblastoma, radiotherapy, surgery.

INTRODUCTION

Embryonal brain tumors include medulloblastoma, central nervous system (CNS) primitive neuroectodermal tumor (PNET), and atypical teratoid/rhabdoid tumor according to the 2007 World Health Organization (WHO) classification for tumors of the nervous system. PNETs occur predominantly in children and adolescents [1]. However, they represent less than 2.5% of childhood brain tumors [2]. They may be phenotypically poor differentiated or show divergent differentiation along neuronal, astrocytic and ependimal lines [1].

These tumors show aggressive clinical behavior and their general prognosis is poor [1]. The prognosis is poorer in infants with PNET who are less than two years old at the time of diagnosis. While 5-year survival rate for the patients with S-PNETs is 34% [3]. Radical resection, radiotherapy, chemotherapy, and or their combinations are the choices of treatment [4, 5]. Herein, we describe a SPNET without evidence of differentiation in a 15-year-old girl who is still under follow-up without recurrence after 36 months.

CASE REPORT

A 15-year-old girl was admitted with a headache lasting about 20 days. Her neurological examination was intact on the admission. T1-weighted magnetic resonance (MR) imaging demonstrated a well-demarcated tumor with heterogeneous enhancement after gadolinium-diethylene triamine penta acetic acid (Gd-DTPA) administration in the left frontal lobe (Fig.1a&b).

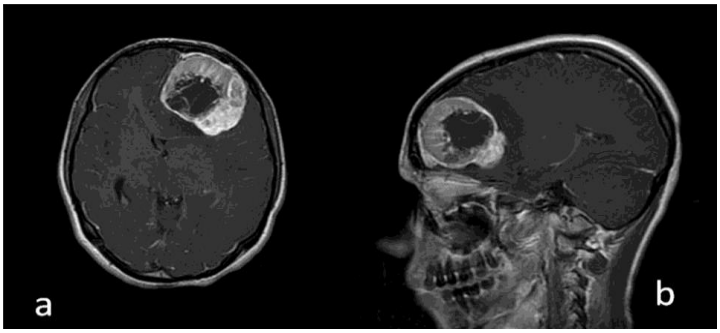

Fig 1a&b: shows a well demarcated highly contrast enhanced space occupying lesion which contains cystic and necrotic areas in the left frontal lobe which causes midline-shift

A left-sided frontal craniotomy was performed. The tumor was grayish, medium-hard, and hemorrhagic. It has a thin capsule and was well demarcated from the surrounding brain parenchyma. The medial part of tumor was fibrous and attached to the falxcerebri. Total tumor resection was performed micro surgically. She recovered well after the operation. The histological examination revealed Supratentorial primitive neuroectodermal tumor [World Health Organization (WHO) grade 4] (Figure 2a,b and c). Ki-67 proliferation index was 40%.

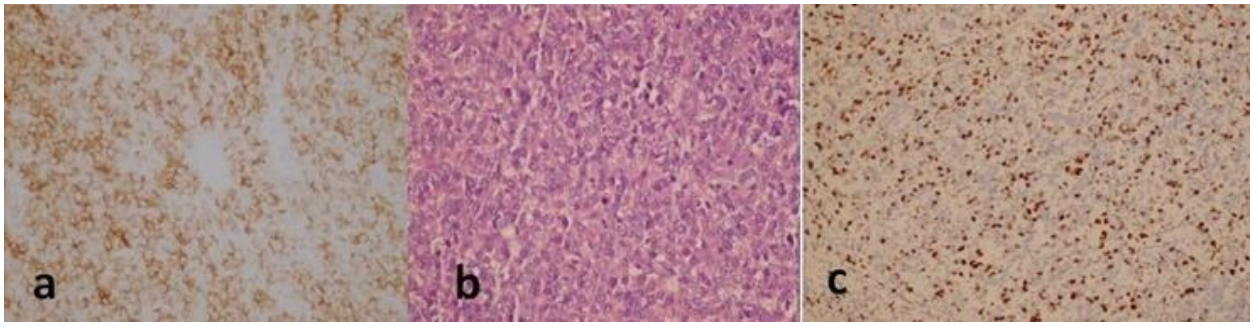

Fig-2(a): Diffuse membranous staining with CD56, a neuro endocrin marker, in primitive cells (immüno histochemical staining, magnification x400)

Fig-2(b):Shows a solitary mass composed of primitive cells which have narrow cytoplasm, uniform round nuclei, and high mitotic activity (H&E staining, magnification x400)

Fig-2(c): Shows highly mitotic activity with Ki 67 proliferation index marker (immuno histochemical staining, magnification x100)

Patient received chemotherapy and radiotherapy after the histopathological diagnosis. Radiotherapy with concurrent vincristine 2 mg weekly for 6 weeks in total was performed. Then a 6 cycles of adjuvant chemotherapy in 21 days CVP (cisplatin, cyclophosphamide, vincristine, and mesna) were given. She remains neurologically free and had no evidence of tumor recurrence at follow-up examination after 36 months (Fig 3a&b).

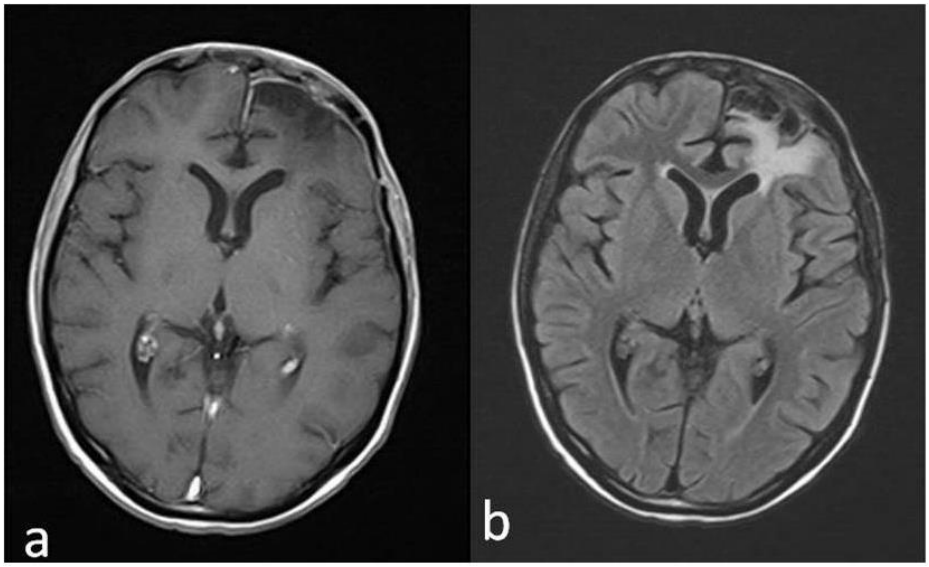

Fig-3a&b: Postoperative MR images obtained 36months after operation showing no evidence of recurrence

DISCUSSION

PNET's are mainly seen in the pediatric population and frequently located in cerebellum [6]. These tumors composed of neuro epithelial cells which are able to differentiate along the neuronal, astrocytic, muscular, or melanocytic lines, with high index of proliferation [1]. PNETs include neuroblastomas, ganglio neuroblastomas, medullo epitheliomas, ependymoblastomas, embryonal tumors with abundant neuropil and true rosettes, and not otherwise specified PNET tumors [1] Supratentorial PNETs are very rare and aggressive tumors, which corresponds to WHO grade IV [7]. They may contain cells displaying features of neoplastic astrocytes, oligodendrocytes, ependymal cells, neuronal cells, melanocytes, or mesenchymal cells [6]. Moreover, PNET located in cerebellum or supratentorium, describing ganglioglial

differentiation or showing coexistence with anaplastic ganglioma were reported in the literature [3,8].

Presenting symptoms are headache, nausea and/or vomiting, confusion and papilledema due to raised intracranial pressure [9,10]. A supratentorial PNET may present with focal neurological signs and deficits, such as limb paresis, aphasia, facial palsy and visual field defects, depending on the anatomic location of the tumor, especially in adults. Seizure may be a rare presenting symptom [9]. Moreover, supratentorial PNET presenting with intracerebral hemorrhage is also reported in the literature [11, 12].

PNET's are considered a challenging clinical entity due to its poor prognosis. When compared with high risk medulloblastoma patients treated with the same regimens, patients with supratentorial PNETs fare worse in terms of survival rates, suggesting that there is an intrinsic difference in the biology of these tumors [13]. The overall 5-year progression-free survival rate for patients who do not undergo resection ranges from 18% to 47% [7]. The reported overall survival of PNETs in the pediatric population ranges between 29% and 57% [14] For pediatric patients with supratentorial PNETs, the prognosis correlates with age, extent of necrosis, tumor dissemination and the presence of bad prognostic genetic features, such as, MYCN or MYCC gene amplifications and polysomies of chromosomes 2 and 8 [9,14] The prognosis is poorer in infants with PNET who are less than two years old at the time of diagnosis. While 5-year survival rate for the patients with S-PNETs is 34%, it is reported to be as high as 85% for the ones with a PNET arising in the posterior fossa [15]. It was reported that the most important prognostic factor in adults was considered to be the Ki-67 index [9]. A Ki-67 index greater than 30% demonstrated very poor outcome.

The complete resection, as possible as, along with radiation and chemotherapy has been shown to lead to better survival in several studies [7]. The extent of resection depends on several factors. Surgeon should be careful about the risks of postoperative neurological deficits. Supratentorial PNETs occurring in eloquent areas of the brain presents a particularly difficult situation because the risks of severe postoperative neurological deficit are relatively high. These tumors have a high tendency for spinal metastasis and approximately 30-50% of the patients with intracranial PNETs develop spinal metastases [16] Postsurgical irradiation of the whole craniospinal axis may be employed, even when there is no evidence of spinal seeding [9]. The employed mean dose is 54Gy for the brain and 31Gy for the spinal cord [9]. The exact role of stereotactic radio surgery employment, especially in cases of partially resected PNETs, remains to be defined.

Yang *et al.;* investigated the clinical features, treatment outcome and prognostic factors of pediatric supratentorial PNTs in a retrospective study in 28 patients. They assessed the prognostic importance of age, sex, size of tumor, M stage, extent of surgical resection, histological features, immuno histochemical labeling indices (Ki-67, p53), and apoptotic index. They reported that 25 patients who completed planned adjuvant therapy, showed a 73% rate of 3-year survival. They concluded that univariate analysis showed a correlation between the presence of tumor necrosis and extent of resection and survival. Patients with a high Ki-67 labeling index (>10%) tended to have shorter survival. In multivariate analysis, tumor necrosis showed statistical significance [10].

CONCLUSION

In the light of the available data about these tumors shows that we know a little about management and treatment of them. Future studies and case series will increase our knowledge base.

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