Infantile Intracranial Hemangiopericytoma: Case Report

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ABSTRACT

Hemangiopericytoma (HPC) is a rare tumor in childhood and accounts for approximately 3% of all soft tissue sarcomas in this age group. Intracranial location is extremely rare. Furthermore, tumors are even more rare in the first year of life and are referred to as infantile hemangiopericytoma. Previously, nine infantile intracranial HPC cases have been reported in the literature. This is a case report of an infantile intracranial HPC and review of the literature.

Key Words: Infantile, Intracranial, Hemangiopericytoma

ÖZET

İnfantil İntrakranial Hemanjioperisitoma: Vaka Sunumu

Hemanjiyoperisitoma nadir görülen bir tümör olup, çocukluk çağı yumuşak doku tümörlerinin %3 kadarını oluşturur. İntrakraniyal lokalizasyon çok nadirdir. Bununla birlikte tümör hayatın ilk bir yılı içerisinde de çok nadir olarak görülür ve bu dönemde görülen tümörlere infantil hemanjiyoperisitoma adı verilir. Literatüre bakıldığında şimdiye kadar dokuz infantil intrakraniyal hemanjiyoperisitoma vakasının bildirildiği görülmektedir. Burada infantil intrakraniyal hemanjiyoperisitoma tanısı alan bir vakayı sunarak literatürü gözden geçirdik.

Anahtar Kelimeler: İnfantil, İntrakraniyal, Hemanjiyoperisitoma

INTRODUCTION

Hemangiopericytoma (HPC) is a rare tumor in childhood and accounts for approximately 3% of all soft tissue sarcomas in this age group.[1-5] The majority of these tumors occur in adults and only 10% of cases occur in children.[6,7] The most common primary site of HPC is extremity, especially the lower extremity. The retroperitoneum is the second most common site of disease followed by head and neck region and the trunk.[1] Intracranial location is extremely rare.[8-9] Furthermore, tumors occuring in the first year of life are even more rare and are referred to as infantile hemangiopericytoma. Previously, nine infantile intracranial HPC cases have been reported.[6,7,10] We have reported a case of infantile intracranial HPC and reviewed the literature.

CASE REPORT

A 12 month-old male had been admitted to an outside medical center with previous complaints of convulsions. He had no neurological or systemic abnormalities on physical examination and after the undercontrol of convulsions; phenobarbital had been started as preventive therapy.

On the 20th day of the preventive phenobarbital therapy, convulsions had restarted but neither vomiting, fever nor trauma determined. The systemic examination was normal and except unconsciousness the other neurological findings were normal. A cranial computed tomography (CT) had been performed. There was a non-homogenious, hyperdense, 5.5 x 4.5 cm dimensional lobulated mass, wide peripheral oedema at the right frontotemporal region, shift to left and compression to right frontal horn of lateral ventricles, third ventricle and mesencephalon (Figure 1). Afterwards the patient was referred to our hospital. During surgical operation, mass was removed only parsially because of massive bleeding. The specimen was hemorrhagic and determined as hemangiopericytoma on pathological examination (Figure 2). On postoperative neurological examination, patient was apathic and his muscles were tense and deep tendon reflexes were increased. The other systemic examinations were normal. Liver and kidney function tests, abdominal ultrasonography and bone scan were normal. A regimen of chemotherapy including temozolomide (200 mg/m^2/day 1-5 days), etoposide (100 mg/m^2/day 1, 2. days), carboplatin (250 mg/m^2/day 1, 2. days) was started. There was 28 days of interval between every course. An oral antineoplastic agent temozolomide of 100 mg capsules were given with nasogastric tube to the patient. Patient was well tolarated this teraphy and except a modarate mucositis no toxicity occured. No febrile neutropenia attact occured either. The evaluation after 4 course of chemoteraphy showed minimal regression at residual mass on CT scan and chemoteraphy continued. After 8 courses of chemoteraphy patient complained of vomiting and lethargy. Pupillas were unisocoric and pupil reactions were positive. Muscle tonus was generally increased, deep tendon reflexes were active and gloscow coma scale was 6. The blood count values were; Htc 29%, leucocyte 4200/mm^3, thrombocyte 150.000/mm^3 respectively. Erythrocyte transfusion was performed to patient. Values for blood Na was 116/mEq/l, blood osmolality was 240 mosmol and urine Na was 57 mmol/l respectively. With this findings SİADH was thought and fluid restriction was performed. Hyponatremia recovered at the followup. At CT scan, massive communicated hydrocephally and a

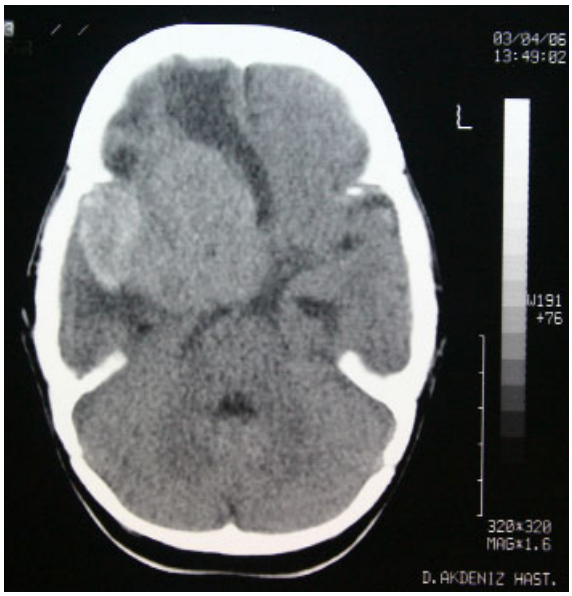
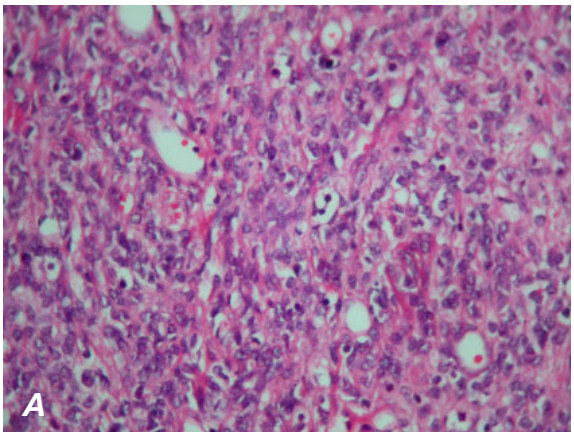
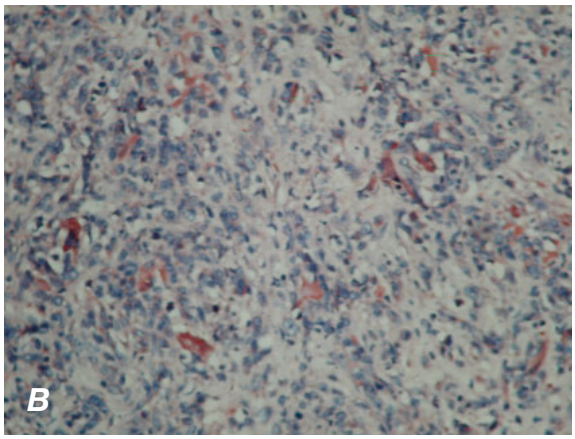


Figure 1. On non-enhanced CT, in the frontotemporal region non-homogeneous, hyperdense, lobulated, well demarcated mass with significant peripheral oedema is seen. The mass is extending to the left side of the mid-line and lateral part of the mass is more hyperdense referring hematoma.





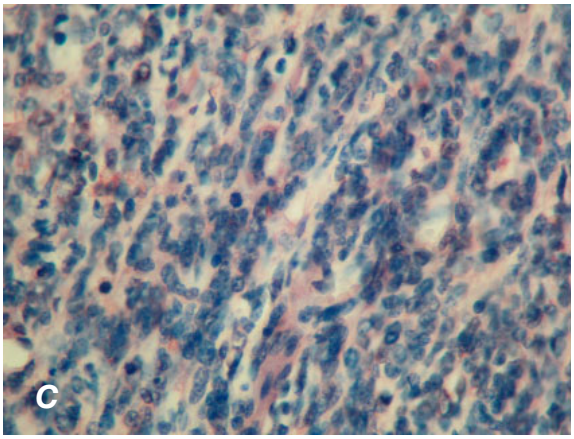


Figure 2 A. Perivascular cells with round-oval, orto and hyperchromatic nuclei, eosinophilic cytoplasm and high mitotic activity are seen. Interstitial lymphocyte infiltration is observed (Hematoxilen-eosin x40 obj).

B. Endothelial cells staining with factor VIII antibody is seen (immunperoxidase x20 obj).

C. Cytoplasmic S-100 protein positive staining of pericytes with increased mitotic activity is noticed (immunoperoxidase x20 obj).

globally rezidual mass which was covering all temporopariatel region and causing shift was demonstrated. Patient was entubated and was taken to pediatric intansive care unit for followup with mecanic ventilation. An external ventricular drenage was performed for hydrocephaly. The patient died due to progression of the mass on the sixth month of the therapy.

DISCUSSION

Hemangiopericytoma (HPC) is rare in childhood and accounts for approximately 3% of all soft tissue sarcomas in pediatric age group.[1-5] The incidence of HPC of the central nervous system (CNS) is less than 1% of all CNS tumors, presenting most often in the third to fourth decade of life but also can occur at any age.[11] Additionaly, these tumors are even more rare in infantile age group. Previously nine infantile intracranial HPC cases have been reported.[6,7,10] Our patient is the tenth infantile intracranial HPC case.

Clinical presentation is usually relates to tumor mass effect or seizures, but in a few patients as in our patient, the initial course presents with intracranial haemorrhage.[12] Also projectile vomiting, focal neurological deficit can be other presentitions. Juvenil children shows headache usually.

These tumors are well demarcated masses which are reported to be heterogeneous and predominately isointense on T1W and T2W images, some of which demonstrates signal flow voids.[13,14] On nonenhanced CT they are slightly hyperdense and may show bony erosions but usually lack the calcifications and hyperostosis associated with meningiomas. Heterogenous contrast enhancement is usually demonstrated on both CT and MRI.[13,14] Our patient was urgently operated because of clinical emergency after observation of the huge mass on CT. The tumor was heterogeneous, hyperdense and well demarcated as reported in literature.

Metastases, although rare, are usually to bone, liver, lungs and kidneys[7,10,15] and our patient didn't show any metastasis neither.

HPC have a propensity for recurring locally. Therefore, every attempt should be made to achieve complete resection of the tumor at the time of initial surgery. Soyuer et al.[10] reported, the 5-year local control rates for patients treated with total tumor resection and subtotal tumor resection were 84% and 38%, respectively. Similarly Kim et al.[15] reported, the 5-year disease recurrence free rates were 73% and 21% for patients with complete and incomplete excision, respectively. Unfortunately complete excision was possible in only 50-67% of the cases in different series.[16] Either total excision wasn't possible in our patient and this process affected prognosis negatively.

Temozolomide is an alkylating agent with excellent oral bioavailability, good penetration across the blood brain barrier, and low toxicity profile. It has been demonstrated to be well tolerated in adults and children with cancer.[17,18] Temozolomide has been studied in more than 1000 patients with primary brain tumors. Various regimens and dosages have been tested in first and second line chemotherapy either monotherapy or in combination with other compounds.[19] Since the introduction of temozolomide, numerous studies have evaluated the efficacy of the compound in the treatment of primary brain tumors as well as brain metastases.[20]

On our previous study at during years of 2003 – 2007 we had administrated temozolomide (200 mg/m^2/day 1-5 days), etoposide (100 mg/m^2/day 1, 2. days), carboplatin (250 mg/m^2/day 1, 2. days) to 34 cases of brain tumor (32 primary, 2 metastatic). The median age of patients were 8.6 ± 4.3 (range 1-17) years. The 3 and 5 years OS rates were 68% and 36% respectively.[21]

We perefered temozolomide based chemotherapy in our case because it was a primary brain tumor and temozolomide has excellent oral bioavailability, good penetration across the blood brain barrier, and low toxicity profile. But there is no report about temozolomide based chemotherapy on intracraniyal HPC cases in the literature.

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