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A case of medulloblastoma mimicking astrocytoma located in cerebello-pontine angle

Serebellopontin açı yerleşimli ve astrositomu taklit eden bir medullablastom olgusu

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Abstract

Medulloblastoma is the most common malignant and uncontrollable childhood brain tumor. This tumor is observed in patients of all ages, reaches a peak in children between 3 and 9 years old, and it is also the most common malignant cerebellar tumor in infants. It accounts for 1% to 2% of all brain tumors in adults. By definition, medulloblastomas arise in the posterior fossa, but histologically similar tumors originating from the pineal region or the pontocerebellar region are no longer classified as medulloblastomas. The traditional treatment for this disease consists of posterior fossa surgery followed by craniospinal radiation therapy plus a high dose boost to the entire posterior fossa. In this report, we present a 13 year-old girl admitted with headache and dizziness symptoms and had medulloblastoma that is located in cerebellopontine angle and mimicking astrocytoma.

Keywords: Cerebellopontine angle, childhood, medulloblastoma.

Öz

Medulloblastom en sık görülen malign ve kontrol edilemeyen çocukluk çağı beyin tümörüdür. Tümör her yaştaki hastalarda görülür, 3 ve 9 yaş arasındaki çocuklarda bir zirveye ulaşır ve aynı zamanda çocukluk çağında en sık görülen kötü huylu beyincik tümörüdür. Yetişkinlerde beyin tümörlerinin %1 ile %2'sini oluşturur. Tanım olarak, medulloblastomlar genellikle posterior fossa, bazen pineal bölge veya pontoserebellar bölgeden kaynaklanan tümörler histolojik olarak benzer şekilde sınıflandırılır. Bu hastalığın geleneksel tedavisi, posterior fossa cerrahisi, ardından posterior fossaya yüksek doz olmak kaydıyla kraniyospinal radyasyon tedavisi şeklindedir. Bu yazıda, 13 yaşında bir kız çocuğunda baş ağrısı ve baş dönmesi belirtileri ile ortaya çıkan, astrositomu taklit eden bir posterior fossa medülloblastomu olgusunu sunuyoruz.

Anahtar Sözcükler: *Serebellopontin açı, çocukluk, medulloblastom.*

Introduction

Mass lesions of the posterior fossa in children usually present a diagnostic challenge despite their frequent occurrence and a number of differential diagnostic possibilities. Medulloblastoma requires an aggressive surgical approach. Preoperative imaging of the entire neuro-axis is critical which shows the presence of drop metastases (1).

Medulloblastoma is highly malignant neuroepithelial tumor of the posterior fossa and also being more important in children. It is tought to arise from primitive, undifferentiated, small, round cells which are located in the superior medullary velum at the roof of the fourth ventricle (2).

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Cerebellar mass is most commonly arise in midline. Obstructive hydrocephalus exists in 95% of cases at presentation. Spinal magnetic resonance imaging (MRI) is necessary to examine leptomeningeal spread of the tumor. The normal expansion of the choroid plexus in the foramina of Luschka from primary cerebrospinal fluid (CSF) seeding of medulloblastoma could be distinguished by MRI (3).

Case Report

A 13-year-old female child admitted to emergency room with complaints of headache and dizziness. The neurological examination of the child revealed disequilibrum. The physical examination and laboratory evaluations were normal. Because of the continuing headache, cranial computed tomography (CT) was performed in which an infratentorial left posterior fossa mass was detected. MRI was planned after the hospitalization of patient at the neurosurgery clinic.

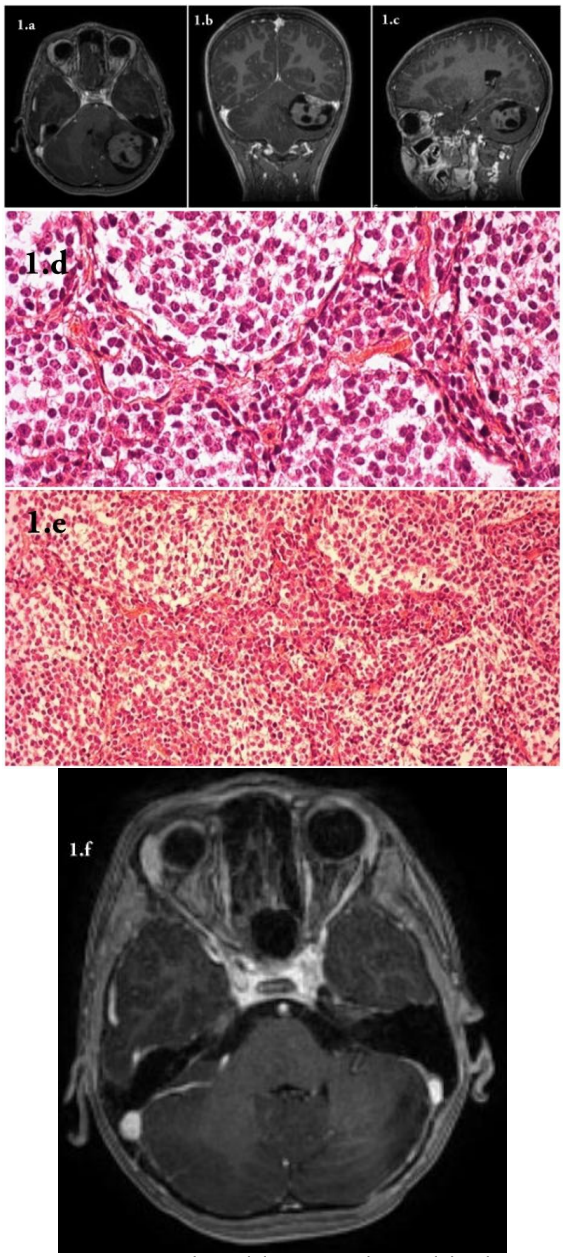

Figure-1.a. Pre-operative axial contrasted T1 weighted MRI scan showed a mural nodule with cystic lesion mass which caused edema and shift in the left posterior fossa region. **b.** Pre-operative coronal contrasted T1 weighted MRI scan showed that left infratentorial-cerebellopontine angle localized mass solid tumor's diameter was approximately 4 cm. **c.** Pre-operative sagittal contrasted T1 weighted MRI scan showed the in the infratentorial cerebellar posterior fossa zone. **d,e.** Specimens showed medulloblastoma (H&E X200-X400). **f.** Post-operative 7.month axial contrasted T1 weighted MRI scan showed that tumoral lesion was totally removed.

MRI showed a solid tumor approximately 4 cm in diameter in the left posterior fossa region which caused edema and the obvious shift effect. It was a mural nodule consisting cystic components (Figure-1a,b,c).

Left posterior fossa surgery was performed into the patient in prone position. Primarily left suboccipital craniotomy and then cerebellomedullary region was seen and by using the tools of microneurosurgery, pontocerebellar angle mass was near totally excised. After the control of bleeding, dura mater was sutured watertight with a 4/0 silk. Histopathological results of the patient showed medulloblastoma (Figure-1d, e, f).

Discussion

Adult medulloblastomas are very rare lesions and make up only 2% of adult central nervous system tumors. The majority of medulloblastomas occur within the first decade of life, and these tumors usually reveal with signs and symptoms of increased intracranial pressure. The tumors originates from primitive neuroectodermal cells, which are originally present in the roof of the fourth ventricle. Radiologically, medulloblastomas are seen as solid, homogeneously enhancing, midline masses of the posterior fossa, frequently arising from the fastigium (4).

Acoustic neuromas and meningiomas form the majority of cerebellopontine angle tumors, however a large spectrum of many unusual lesions form the remaining group of cerebellopontine angle lesions (5).

Cerebellopontine angle astrocytomas are also infrequently seen children. The lesion generally arises in the vermis or cerebellar hemispheres; as a result, cranial nerve involvement is uncommon. Only when originating in the mid cerebellar peduncles, the tumor resides in the cerebellopontine angle. The slow growth of these neoplasms, may lead delay in diagnosis (6).

Medulloblastomas account for less than 2% of all central nervous system tumors in adults. In contrast to adolescents and adults, in whom these tumors are most commonly found laterally in the cerebellar hemispheres, approximately two-thirds of medulloblastomas in children are located and settled in the vermis. Medulloblastomas have relatively rapid growth rates and can intrude on near or next neural tissue and metastasize along CSF pathways (7).

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