Cukurova Medical Journal
ÇUKUROVA ÜNİVERSİTESİ TIP FAKÜLTESİ

Cukurova Med J 2023;48(3):1187-1189
DOI: 10.17826/cumj.1315981

LETTER TO THE EDITOR

Concurrent medulloblastoma and Chiari Type-I deformity in a pediatric case

Pediatrik bir hastada eş zamanlı medulloblastoma ve Chiari Tip-I deformitesi

İlknur Sürücü Kara[1,2], Tayfun Çakır[3]

[1]Erzincan Binali Yildirim University Faculty of Medicine, Department of Child Health and Diseases, Erzincan, Turkey, [2]Ankara University, Faculty of Medicine, Pediatric Metabolism, Ankara, Turkey, [3]Department of Brain and Neurosurgery, Erzincan, Turkey

To The Editor,

Concurrence of medulloblastoma and Chiari type 1 deformity/malformation (C1D) in a pediatric patient is extremely rarely seen[1]. Any pathophysiological relationship between these two diseases has not been mentioned yet[1,2]. Medulloblastoma is frequently seen in the posterior fossa, and therefore, it can cause cerebellar dysfunction and increased intracranial pressure more quickly than other intra-cranial lesions[3].

C1D is the condition in which there is a caudal herniation of the cerebellar tonsils below the foramen magnum[1]. C1D puts pressure on parts of the brain and spinal cord and can cause mild to severe symptoms. In most cases, the problem is congenital. Also, associated abnormalities have been reported concurrent with CID [4,5]. When we diagnose any of these diseases in a patient, the coexistence of two conditions may occur in the same patient. And we may need to find the correct diagnoses.

In this letter, we report a case of a 10-year-old girl patient with concurrent C1D and medulloblastoma. She was admitted to the hospital for progressive neck pain, headache, nausea, and vomiting symptoms. She had a history of intracranial hemorrhage due to vitamin K deficiency when she was 45 days old. Therefore, she had severe complications in the cortex. She followed up annually in the related units due to developmental delay, epilepsy, hyperactivity, and attention deficit disorder. The patient hadn't signs of meningeal irritation, and neurological evaluation and measurement of blood pressure were in the normal range. Nevertheless, the headache duration experienced during the whole day with flarings lasted 7 to 8 days. Computed tomography (CT) scan indicated atrophy and encephalomalacia areas (in the cerebral hemisphere), asymmetric enlargement (in the lateral ventricles), and subcortical hypodense density changes (in the right frontal lobe). There was mild prominence in the third ventricle and the appearance of the left cerebellar tonsils down to the foramen magnum level. In addition, examining the patient for Chiari deformity (type 1) was suggested. The patient was hospitalized two days later because of severe abdominal pain and vomiting. Physical examination, laboratory parameters, and abdominal ultrasonography were unremarkable. The patient continued vomiting, and intense headaches increased. Cranial magnetic resonance imaging (MRI) was performed. Unfortunately, there was a lesion as a solid mass, about 60 mm sized in the level right cerebellar, a cerebellar tonsil hernia (11 mm), and ventricular dilatation in magnetic resonance imaging (Fig. 1, 2, 3). Emergency total tumor excision was performed, and posterior fossa decompression was applied to the patient. Histological diagnosis confirmed the lesion as a classic type of medulloblastoma. Post-operatively, the patient was administered chemo-radiation treatment according to institutional protocol. The patient died one year after the diagnosis of medulloblastoma.

Address for Correspondence: İlknur Sürücü Kara, Erzincan Binali Yildirim University Faculty of Medicine, Department of Child Health and Diseases, Erzincan, Turkey E-mail: drilknursurucu@gmail.com
Received: 23.06.2023 Accepted: 27.08.2023

As can be seen, the clinical findings of this patient were compatible with both posterior fossa tumors and C1D. Meningeal irritation, neck pain, and headaches are the most common presentations/ clinical findings of C1D and posterior fossa tumors [3, 6]. Medulloblastoma causes 90% of embryonal tumors in children. About 10% of all brain tumors are caused by medulloblastoma, and medulloblastoma can spread through leptomeningeal tissue. The peak of incidence is 5-9 years of age. Three-quarters of medulloblastoma cases originate from the cerebellar vermis and tend to progress towards the fourth ventricle, typically without extending to basal cisterns[7, 8].

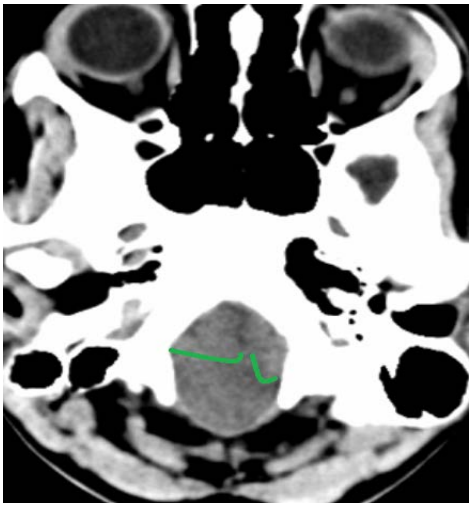

Figure 1. Cerebellar tonsils are seen at the level of foramen magnum on cranial CT.

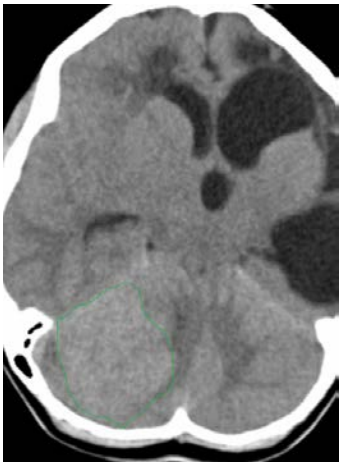

Figure 2. The hypodense footprint lesion was too faint to be noticed on cranial CT.

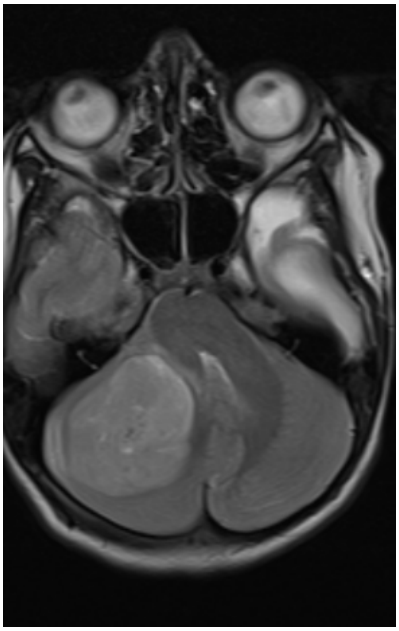

Figure 3. Cerebellar lesion that occupies a place compatible with medulloblastoma in the cranial MRI.

Less than 5 percent of cases are associated with hereditary cancer susceptibility[3]. Medulloblastomas are classified according to molecular subgroups; their prognosis varies with their molecular subgroups, risk groups, age, and response to treatment[3].

Medulloblastomas have different imaging features in both CT and MRI [3]. With the widespread usage of MRI, patients are diagnosed at an earlier age even if they do not have symptoms or there is a minor number of symptoms[9].

Cerebellar tonsils enlarge with age, so cerebellar tonsils may be considered normal if it is 6 mm below the foramen magnum in infants, borderline displacement of cerebellar tonsils in adults is ≥3 to <5 mm below the foramen magnum, other craniocervical joint anomalies or syringomyelia, which are associated with additional features of C1D, are considered pathological ones [9]. Chiari deformities have no diagnostic biomarkers. The diagnosis is based on neuroanatomy, and MRI is the best imaging evaluation method[9].

It has been supposed that local friction or changes in local pressure due to Chiari deformities may cause malignant transformation in cells and more severe cerebellar tonsil herniation as the tumor grows [10].

Therefore, early decompression is recommended for Chiari deformities[10].

The number of cases in which the association of posterior fossa medulloblastoma with Chiari type 1 deformity has been reported is low (in the literature, we found only 6 cases of children with Chiari type 1 deformity and medulloblastoma coexistence [1, 2, 10]. Headache is the most common symptom of Chiari 1 deformity. Cerebellar herniation may be due to the mass effect of posterior fossa tumors. A CT scan may be insufficient to reveal the presence of additional pathologies. MRI, which indicates the craniovertebral junction, is the first method to be selected for diagnosis, and it can be used singly.

Author Contributions: Concept/Design: İSK, TÇ; Data acquisition: -; Data analysis and interpretation: -; Drafting manuscript: İSK; Critical revision of the manuscript: İSK, TÇ; Final approval and accountability: İSK, TÇ; Technical or material support: -; Supervision: İSK, TÇ; Securing funding (if available): n/a.
Peer-review: Editorial review.
Conflict of Interest: Authors declared no conflict of interest.
Financial Disclosure: Authors declared no financial support

REFERENCES

1. Low SYY, Lian DWQ, Tang PH, Loh E, Seow WT, Low DCY. Concurrent pediatric medulloblastoma and Chiari I malformation with syringomyelia. Childs Nerv Sys. 2017;33:881-3.
2. El Hassani Y, Burkhardt K, Delavellle J, Vargas MI, Boex C, Rilliet B. Symptomatic syringomyelia occurring as a late complication of posterior fossa medulloblastoma removal in infancy in a boy also suffering from scaphocephaly. Childs Nerv Syst. 2009;25:1633-7.
3. Millard NE, De Braganca KC, Medulloblastoma. J Child Neurol. 2016;31:1341-53.
4. Fernández AA, Guerrero AI, Martínez MI, Vázquez ME, Fernández JB, Chesa i Octavio E et al. Malformations of the craniocervical junction (Chiari type I and syringomyelia: classification, diagnosis and treatment). BMC Musculoskelet Disord. 2009;10 Suppl 1:S1..
5. Aslan N, Sesli E, Pirgon Ö. A rare cause of tall stature: Sotos syndrome. Dicle Med J. 2014;41:760-2
6. Poretti A, Ashmawy R, Garzon-Muvdi T, Jallo GI, Huisman TA, Raybaud C. Chiari type 1 deformity in children: pathogenetic, clinical, neuroimaging, and management aspects. Neuropediatrics. 2016;47:293-307.
7. Koeller KK, Rushing EJ. From the archives of the AFIP: medulloblastoma: a comprehensive review with radiologic-pathologic correlation. Radiographics. 2003;23:1613-37.
8. Poretti A, Meoded A, Huisman TA. Neuroimaging of pediatric posterior fossa tumors including review of the literature. J Magn Reson Imaging. 2012;35:32-47.
9. Uptodate, Chiari malformations. Topic 13507 Version 24.0. https://www.uptodate.com/contents/chiari-malformations (Jul 2023).
10. Qi X, Yang S, Xu Z, Ma Z, Yan Q, Wang Y, Zhu Y. Simultaneous occurrence of Chiari type I malformation and classic medulloblastoma. Neurosurg Q. 2015;25:415-18.