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Abnormal Brain MRI in a Case of Acute Ataxia as the Only Sign of Abdominal Neuroblastoma

Ataxia is a movement disorder that may manifest an acute, intermittent, non progressive or chronic progressive course. Ataxia alone is rare as a paraneoplastic sign, especially if it is due to neuroblastoma (abdominal or chest).
We report an abdominal neuroblastoma in a two-year-old girl presenting with only acute ataxia and abnormal neuroimaging. Brain MRI showed abnormal signal finding in the medulla, pons, corticospinal tract and the periventricular space. In the abdominal CT, a mass was detected in the right adrenal gland with calcification and the histopathologic examination re-vealed neuroblastoma.
We suggest in children with acute ataxia, with or without opsoclonus-myoclonus, neuroblastoma should be considered.

Keywords: Neuroblastoma, Abnormal Neuroimaging Finding, Acute Ataxia

Introduction

Neuroblastoma is a common pediatric solid tumor accounting for approximately 8% of childhood malignancies. This cancer is the third most common cancer in the pediatric population.[1] As it is able to simulate many other disorders, the diagnosis may be difficult.

Neuroblastoma may present as a paraneoplastic syndrome of autoimmune origin, manifesting as ataxia or opsoclonus (dancing eyes and dancing feet). In such cases, the primary tumor is located in the chest or abdomen and the brain is not involved directly by the tumor.

Infantile polymyoclonus or opsoclonus-myoclonus syndrome (OMS) or Kinsbourne syndrome is an uncommon acquired disorder of late infancy. The clinical features of the syndrome include acute or subacute onset of rapid dancing eye movements and myoclonic jerks of the limbs and the trunk and ataxia. OMS has several potential causes. There is an important association between OMS and neuroblastoma.[1]

In a previous study, the mean delay mentioned for diagnosis was 11 weeks, and treatment was initiated after 17 weeks.[2] Neuroblastoma is detected in approximately one-half of cases with OMS and in some studies, all patients have been reported as neuroblastoma. It is important to exclude OMS, even when neither opsoclonus nor myoclonus is present; consequently, acute ataxia, as the only sign should be considered.[3]

OMS particularly originates from the middle part of the cerebellum and single photon emission computed tomography (SPECT) is the best way to detect functional deterioration in these patients.[4] Myoclonous may cease and opsoclonus may persist during sleep in these patients.[5] In OMS patients, immunologic abnormalities in-cluding T-cell abnormalities, decrease of CD4+, T-cell subset and the CD4/CD8 ratio have been detected.[6]

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Initially, the syndrome was attributed either to the presence of occult neuroblastoma or to an unknown cause. With improved imaging techniques, approximately all cases appear to be related to neuroblastoma.[7]

Case Presentation

A two-year-old girl was admitted to Mofid's Children Hospital in Tehran with acute ataxia. She had this complaint from 2 weeks ago, and there was no history of previous infections, drug ingestion, trauma, seizure, vomiting or positive family history of ataxia. Neurodevelopmental milestones were adequate. Physical examination of the chest, abdomen and musculoskeletal system were normal. Neurological examination revealed intact cranial nerves, truncal and limb ataxia, intention tremor, wide base gait, decreased deep tendon reflex (DTR) and dysarthria. Nystagmus, opsoclonus and myoclonus were negative and the mental status was normal. Routine laboratory tests were normal (Table 1).

Brain MRI showed several hyperintensities in the medulla, pons and the corticospinal tract mimicking demyelinating lesions (Figs. 1 & 2).

Based on the above mentioned findings, at first our impression was ADEM (Acute Disseminated Encephalo Myelitis), but to rule out other possibilities, abdominal and chest CT were performed. Chest CT was normal but in the abdominal CT, there was a mass in the right adrenal gland crossing the midline and calcifications were detected (Fig. 3).

Neuroblastoma was the first diagnosis, which was confirmed by histopathological examination (Fig. 4).

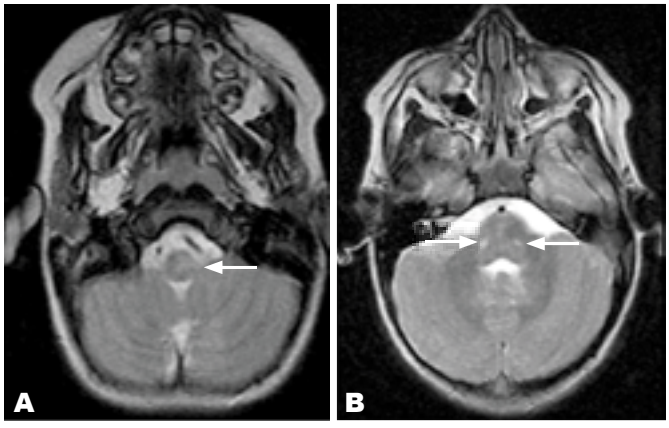

Fig. 1. A two-year-old girl with abdominal neuroblastoma. Two sections of axial T2-Weighted images.
A. Hyper intensity in the left side of the medulla (inferior olive).
B. Hyperintensities in the bilateral corticospinal tracts.

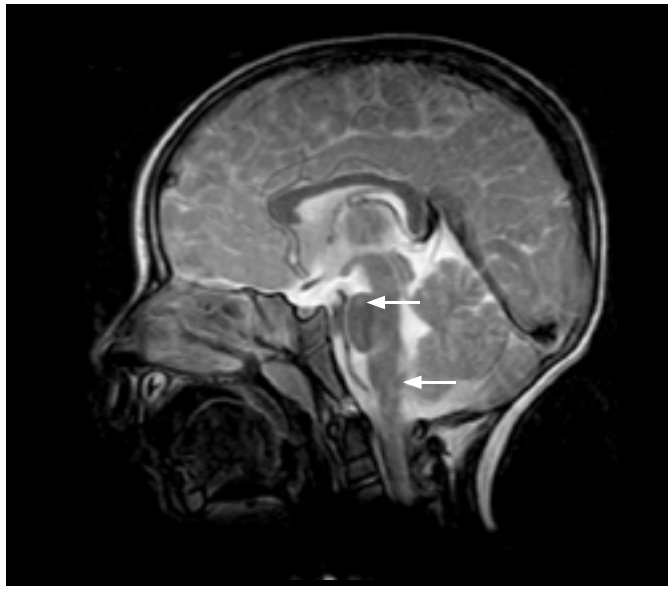

Fig. 2. Sagittal T2-Weighted image reveals patchy hyperintensities in the medulla and pons (arrows).

Discussion

Acute ataxia has many etiologies. ADEM and Kinsbourne syndrome are two rare causes of acute ataxia. Abnormal neuroimaging is usual in ADEM, but rare in Kinsbourne syndrome. To the best of our knowledge, two adult cases with abnormal neuroimaging and Kinsboure syndrome have been reported. Differential diagnoses of acute ataxia and abnormal neuroimaging are brain tumor, cerebellar tumor, ADEM, stroke and Multiple Sclerosis (MS).

In a case reported by the Chinese University of Hong Kong in 2003 of a 26-month-old girl who presented with manifestations similar to encephalitis including ataxia, seizure, decreased consciousness and involuntary movements, apart from late demyelinating changes seen on MRI, brain and spine MRI, she had no problems two weeks after presentation. She was a case of occult neuroblastoma and the above mentioned findings confirmed an immunological explanation

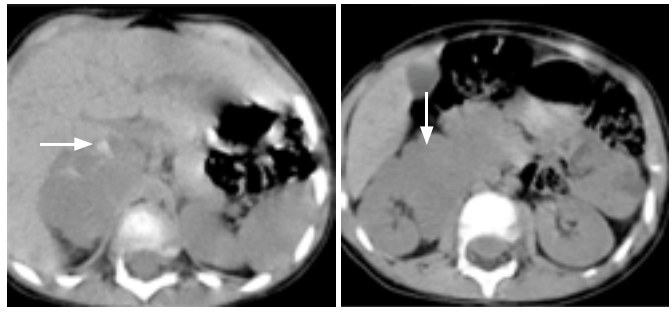

Fig. 3. Axial abdominal CT scan without contrast revealing calcified soft tissue mass over the right kidney in the region of the right adrenal gland which was proved to be neuroblastoma after surgery.

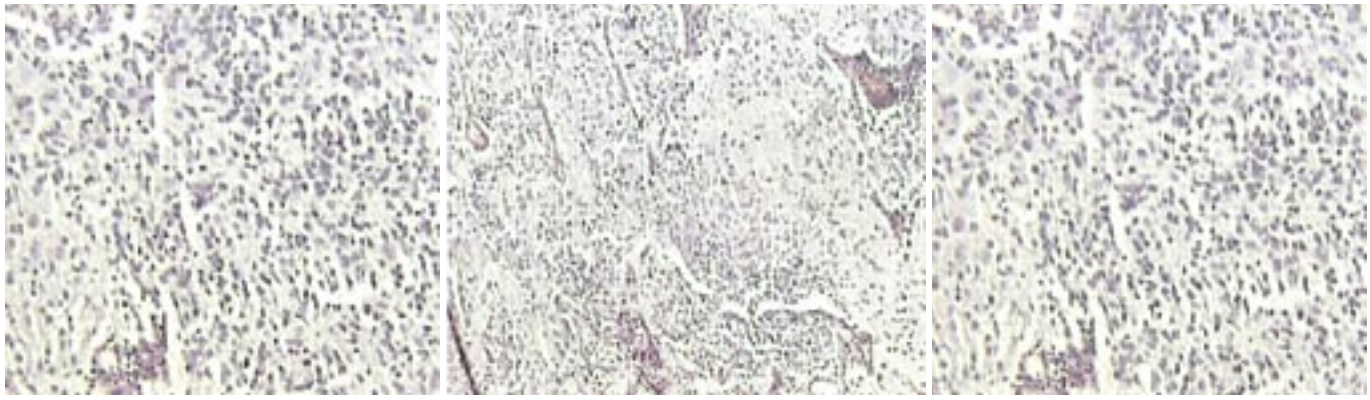

Fig. 4. Histopathology of neuroblastoma in this patient.

for the paraneoplastic demonstration of occult neuroblastoma.[8]

Some studies reported an association between OMS (without neuroblastoma) and focal lesions in the brain stem (especially pontine lesions) in MRI. In the review of literature, two patients with OMS whose MRIs showed brain stem lesions and developed OMS after an upper respiratory tract illness were reported. In one case, the lesion was in the pons at the junction of basis and tegmentum and in another case, a focal lesion was located in the upper pontine tegmentum.[9]

Dropcho et al. assumed that the clinical appearance of opsoclonus may be caused by functional or structural disturbance of several locations in the CNS; whereas, Hattori et al. approved that the pontine tegmentum lesion accomplishes a specific key role in its development.[9]

In our case, the pontine tegmentum was intact and absence of opsoclonus-myoclonus was described with the location of the lesion. Therefore, our patient is the first case of neuroblastoma and its paraneoplastic syndrome, manifesting only with acute ataxia and abnormal neuroimaging finding in the medulla and pons.

We recommend that every child with acute ataxia, with or without abnormal neuroimaging should be examined for occult neuroblastoma.

Table 1. The Patient's Laboratory Test Results

Lab.Tests	Result
LFT*	NL*
Biochemistry	NL
CBC*	NL
ESR*	16
CRP*	Neg.
VMA*	10/4[NL(<5/2)]

*NL=normal, LFT=liver function test, CBC=complete blood count, ESR=erythrocyte sedimentation rate, CRP=C reactive protein, VMA=vanillylmandelic acid.

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