

Primary Cerebellar Tuberculoma in Arnold-Chiari Malformation Mimicking Posterior Cranial Fossa Tumor: The First Report

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Global Spine J 2011;1:19–22

Abstract

Keywords

- cerebellar tuberculoma
- Arnold-Chiari malformation
- cranial fossa tumor

Chiari malformations are a congenital heterogeneous group of disorders characterized by anatomic anomalies of the cerebellum, brain stem, and craniocervical junction associated with downward displacement of the cerebellum, alone or with lower medulla, into the cervical spine canal. The patient was a 23-year-old woman, a known case of Arnold-Chiari malformation with peripheral neuropathy and muscular atrophy, who presented with headache, drowsiness, decreased vision, and severe gait dysfunction lasting for several years. Brain magnetic resonance imaging confirmed a hypointense signal mass in the left hemisphere of the cerebellum causing mass effects on the fourth ventricle, which shifted it, accompanied with dilation of third and lateral ventricles.

Chiari malformations are a congenital heterogeneous group of disorders characterized by anatomic anomalies of the cerebellum, brain stem, and craniocervical junction associated with downward displacement of the cerebellum, alone or with lower medulla, into the cervical spine canal.¹ These disorders are first described by John Cleland in 1883^{2,3} and were classified later by Hans Chiari in 1891 into four groups.^{1,4}

Arnold-Chiari malformation, or Chiari II malformation, is characterized by downward displacement of the cerebellar vermis and tonsils, a spinal myelomeningocele, and a brain stem malformation with beaked midbrain in neuroimaging. We report an unusual case of Arnold-Chiari malformation with primary cerebellar tuberculoma mimicking posterior cranial fossa tumors. To our knowledge, this is the first report of primary cerebellar tuberculoma in a patient with Arnold-Chiari malformation.

Case Report

The patient was a 23-year-old woman, a known case of Arnold-Chiari malformation with peripheral neuropathy

and muscular atrophy, who presented with headache, drowsiness, decreased vision, and severe gait dysfunction lasting for several years. The patient was hospitalized with the primary impression of cerebellar tonsil herniation. The neurological examination revealed upper motor neuron dysfunction with abnormal cerebellar function tests.

Brain magnetic resonance imaging (MRI) confirmed a hypointense-signal mass in the left hemisphere of the cerebellum causing mass effects on the fourth ventricle, which shifted it, accompanied with dilation of third and lateral ventricles. Hypertensive cerebrospinal fluid (CSF) form of hydrocephaly was seen in the supratentorial region, but the fourth ventricle was normal. Subependymal CSF diffusion around lateral ventricles was found. The possibility of aqueduct stenosis or obstruction was mentioned (► Fig. 1). Cervical spine MRI revealed a normal spinal column with herniation of the cerebellar tonsil (► Fig. 2).

With the impression of tonsillar herniation and cerebellar tumor in the sense of Arnold-Chiari syndrome, external ventricular drainage with external reservoir was performed with a right posterior lateral approach. Then the patient

received
September 13, 2011
accepted
October 24, 2011

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Tel: +1(212) 584-4662.

DOI <http://dx.doi.org/10.1055/s-0031-1296052>.
ISSN 2192-5682.



Figure 1 Brain magnetic resonance imaging. A hypointense signal mass in the left hemisphere of the cerebellum causing mass effects on the fourth ventricle, which shifted it, accompanied with dilation of third and lateral ventricles.

underwent microscopic craniotomy of posterior fossa. A cream-white tight mass was seen, which could not be extracted by suction and thus was totally resected. Simultaneously, regarding the Arnold-Chiari malformation and tonsillar herniation, the patient underwent suboccipital decompression and microscopic laminectomy of C2 vertebrae with excision of foramen magnum and duraplasty.

Surgical specimens of the cerebellar mass were submitted for histopathologic examination. Gross examination revealed multiple pieces of irregular cream tissue totally measuring $2.5 \times 2.0 \times 0.7$ cm. Microscopic assessment confirmed a granulomatous inflammation composed of aggregation of epithelioid histiocytes associated with giant cells and lymphocyte cuffing foci of caseating necrosis compatible with tuberculoma.

Postoperative spiral chest computed tomography (CT) scan showed a normal pulmonary parenchyma without evidence for pulmonary tuberculosis. Thus, the patient had a primary extrapulmonary cerebellar tuberculoma (→Fig. 3). The patient has been followed to now; the neurological symptoms were alleviated 6 months subsequent to the surgery.

Discussion

Chiari malformations are rare congenital anomalies with an estimated prevalence of 0.1 to 0.5%⁵; however, the true frequency is unknown. In most cases, due to the small posterior fossa, neural elements are crowded and impacted at the foramen magnum. The main pathogenesis of this malformation remains the subject of debate and involves patients presenting with a wide spectrum of clinical symptoms.¹

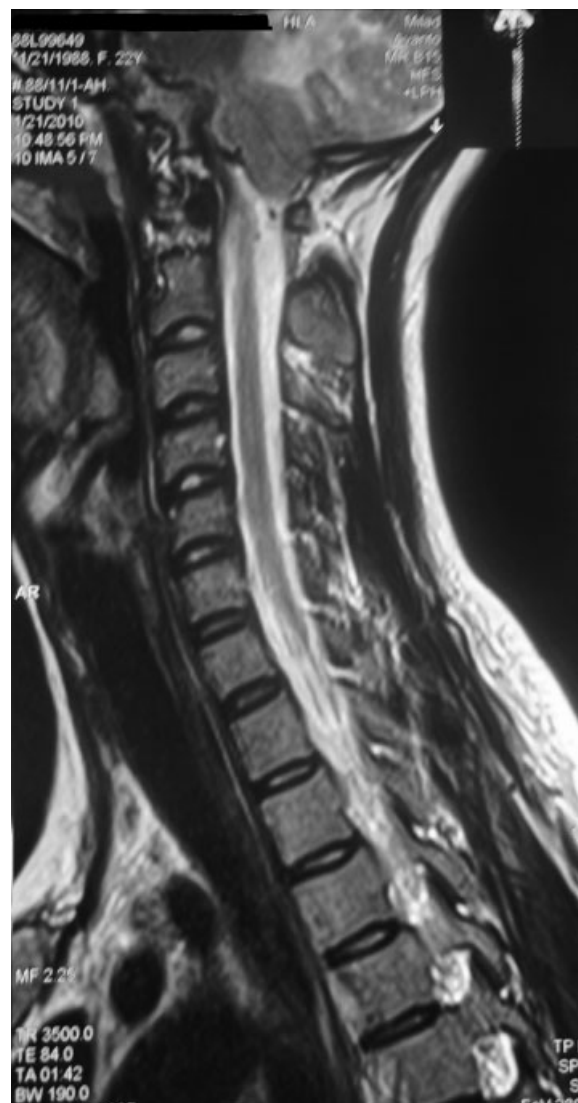


Figure 2 Cervical spine magnetic resonance imaging. Cervical cord has a normal diameter and signal without evidence for compression. Cervical vertebrae have normal height without fractures or pathological lesions. Cervical canal has a normal diameter without any congenital or acquired stenosis. The alignment of the cervical spine is normal without any evidence for disc herniation. Tonsillar herniation to the cervical spine is seen.

Arnold-Chiari syndrome is usually detected prenatally or at birth, as it is nearly always associated with lumbosacral or thoracic myelomeningocele.¹ Weakness, stridor, apneic spells, aspiration, and dysphagia are the common manifestations in infancy,⁶ followed by progressive hydrocephalus in childhood.¹ It may be associated with other syndromes like syringomyelia and scoliosis.^{7,8} Our case presented with headache, drowsiness, decreased vision, and severe gait dysfunction lasting for several years.

Arnold-Chiari syndrome should be considered in any fetus or newborn with clinical evidence of a spinal myelomeningocele. Neuroimaging plays the main role in confirming the diagnosis, and MRI is the best imaging modality for evaluation.⁹ MRI of the brain along with the entire spinal cord (cervical) is appropriate to demonstrate downward displacement of the

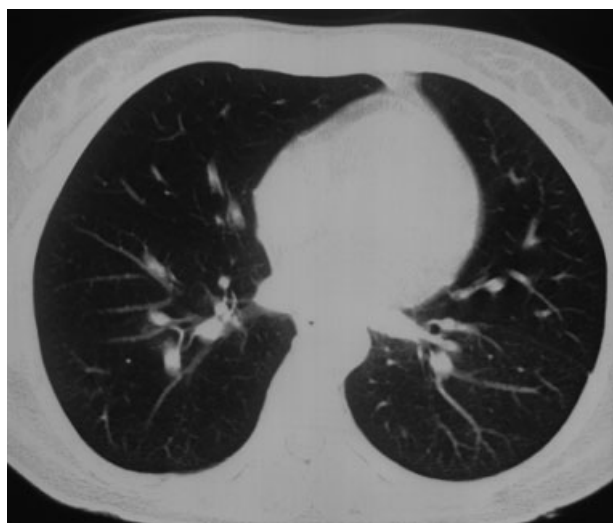


Figure 3 Spiral chest computed tomography. Pulmonary parenchyma and vessels are normal. There is no nodule or occupied lesion in the parenchyma. Mediastinum and pulmonary hila are normal. No pleural lesion or pleural effusion is seen. Pericardium is normal. Mediastinal vessels do not show any pathology. Lungs are relatively hyperinflated.

inferior cerebellar vermis and medulla through the foramen magnum into the upper cervical canal. Despite these typical findings, brain MRI in our case showed an unusual mass in the left cerebellar hemisphere causing mass effect.

The patient underwent the most common procedure for Arnold-Chiari malformations, which is posterior decompression via suboccipital craniotomy with duraplasty.^{10,11} Simultaneously, regarding the cerebellar mass with the primary impression of cerebellar tumor, posterior fossa craniotomy and resection of the mass were performed. Postoperative histopathologic evaluation showed that the mass was in fact a granulomatous tuberculoma. Because no pulmonary involvement was found in either clinical manifestation or pulmonary CT scan, it was diagnosed as a primary extrapulmonary cerebellar tuberculoma mimicking a cerebellar tumor.

Although tuberculosis is considered primarily a pulmonary disease, it can affect any organ system. However, central nervous system (CNS) involvement is so rare, it is associated with potentially devastating complications; it affects both immunocompetent and immunologically incompetent populations. The cerebellar involvement is even rarer. Up to now, only a few numbers of cases with cerebellar tuberculoma have been reported, and most of them are secondary to the pulmonary involvement¹² and occurred in immunocompromised patients.^{13,14} Primary cerebellar tuberculoma is rare in an immunologically incompetent patient like our case. To our knowledge, this is the first reported case of primary cerebellar tuberculoma mimicking a posterior fossa tumor in a patient with Arnold-Chiari malformation.

Depending upon the involved part of the CNS, various spectrums of clinical manifestations may occur. Intracranial

tuberculoma may present as either a solitary or multiple lesions in the brain parenchyma. A ring-enhanced area on CT scan or MRI is a characteristic appearance, but when there are no accompanied clinical manifestation or laboratory findings for tuberculosis, it would be so difficult to differentiate it from other CNS tumors.

Conclusion

Despite being rare, CNS involvement of tuberculosis always should be kept in mind in any patient with neurological complaints from regions with a high endemic rate of tuberculosis, either in those being immunocompetent or immunologically incompetent.

Acknowledgment

The authors would like to thank Farzan Institute for Research and Technology for technical assistance.

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