Case 1057

Eurorad

Chiari III malformation

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Section: Neuroradiology
Imaging Technique: MR
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Case Type: Clinical Cases
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Patient: 16 months, male

Clinical History:

A 16-month-old male patient with swelling at the back of the craniocervical junction and hydrocephalus.
Imaging Findings:

A 16-month-old male patient was referred to our hospital with the history of swelling at the back of the craniocervical junction and hydrocephalus at birth, upon which a ventriculoperitoneal shunt operation was performed with no diagnosis by any medical imaging modality. Since the swelling still persisted, he was referred for MRI, and SE T1 and FSE T2 weighted sequences on three planes were performed with 1.5 Tesla MR scanner. A small posterior fossa volume with low lying transverse sinuses, fenestrated falx cerebri, heart shaped incisura with hypoplastic tentorium cerebelli, concave clivus and petrous bone, tectal beaking, creeping of the cerebellar hemispheres around brain stem, triventricular hydrocephaly, elongated and tube-like narrowed inferiorly located fourth ventricle, colpocephaly, the herniation of cerebellovermian formations through an osseous defect at the level of inferior occipital bone and C1 and C2 cervical vertebra posterior elements were the findings.
Discussion:

In his initial report of 1891, Chiari described a single case of caudal medullary displacement and cerebellar herniation, initially through the foramen magnum, then dorsally through a cervical spina bifida, resulting in a cervical encephalocele. Since that time, the definition of Chiari III malformation has been expanded by some authors to include cases of hindbrain herniation into cephaloceles which encompass both the low occipital and upper cervical regions. Chiari III malformation is characterized by the herniation of posterior fossa contents (cerebellum, and sometimes brainstem and fourth ventricle) through a low occipital or upper cervical osseous defect, thus could be described as occipitocervical cephalocele or rhombencephalocele. Chiari III malformation is a rare abnormality as reported in the literature, and it is expected to show findings related to Chiari II malformation (Arnold-Chiari malformation) as well. Those findings are as follows: small posterior fossa volume with low lying transverse sinuses, fenestrated falx cerebri, heart shaped incisura with hypoplastic tentorium cerebelli, cerebellar hypertrophy, herniation of cerebellar tonsils into the foramen magnum, concave clivus and petrous bone, deformed mesencephalon and tectal beaking, creeping of the cerebellar hemispheres around brain stem, triventricular hydrocephaly, elongated and tube-like narrowed inferiorly located fourth ventricle, corpus callosum dysgenesis, agenesis of posterior cervical vertebra elements, syringomyelia, aberrant draining veins and ectopic venous sinuses. Histopathologic examination of occipitocervical cephaloceles reveals many associated anomalies such as necrosis, gliosis, heterotopias, meningeal fibrosis, most of which couldnot be evaluated by medical imaging modalities.

Differential Diagnosis List: Chiari III malformation

Final Diagnosis: Chiari III malformation

References:

Chiari H. ?ber Veranderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns. Dtsch Med Wochenschr 1891; 17:1172-1175.
Chiari H. ?ber Veranderungen des Kleinhirns, des Pons unde der Medulla
Oblongata infolge von congenitaler Hydrocephalie des Grosshirns. Denkschriften der Kais Akad Wiss math-naturw 1896;63:71-116.
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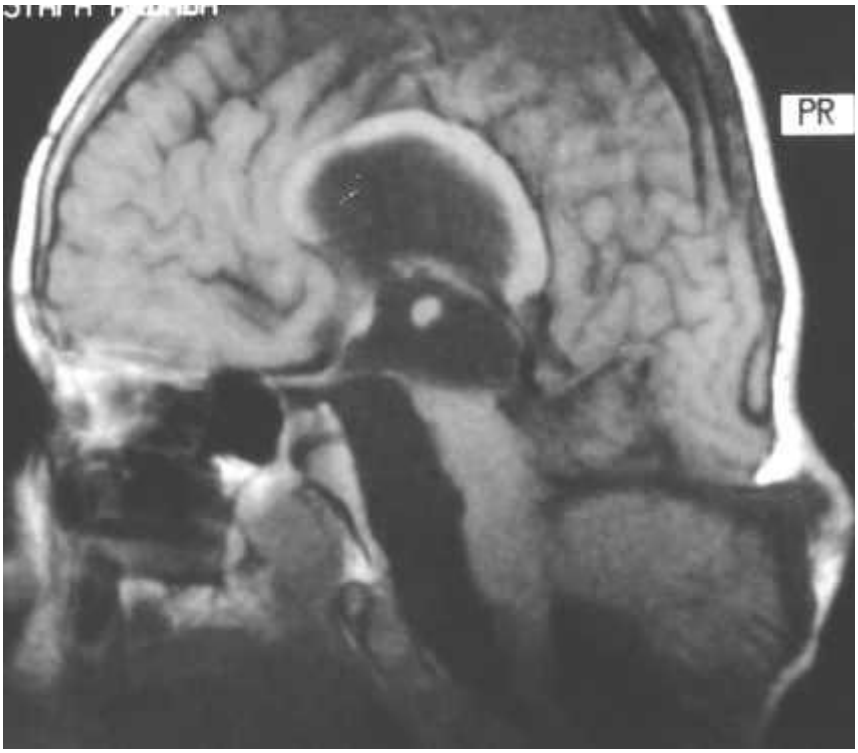
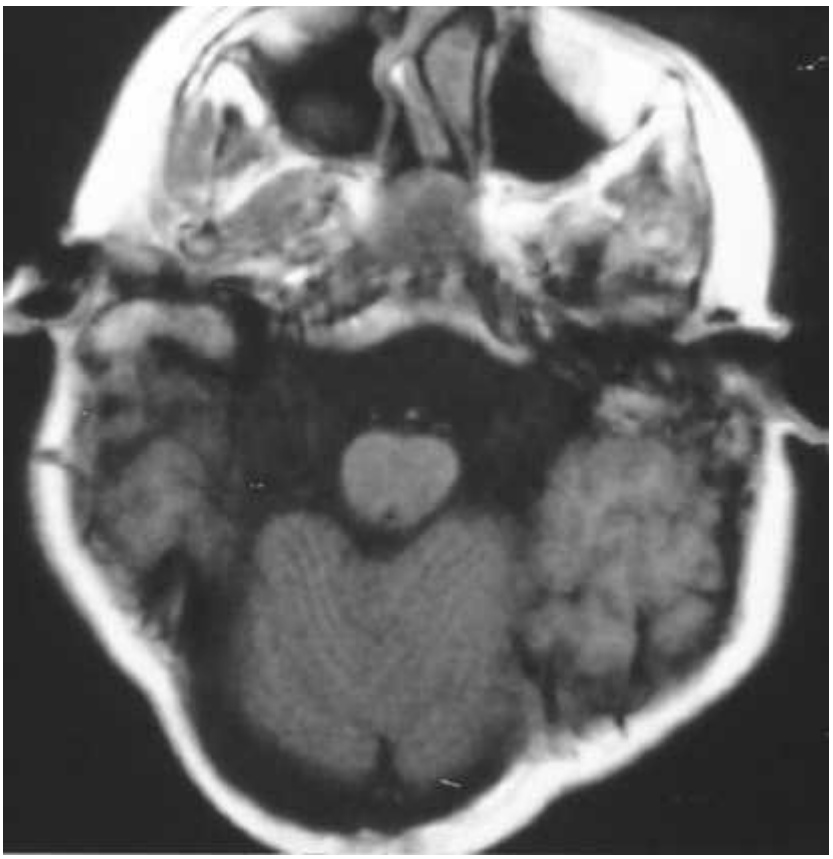
Figure 1

a



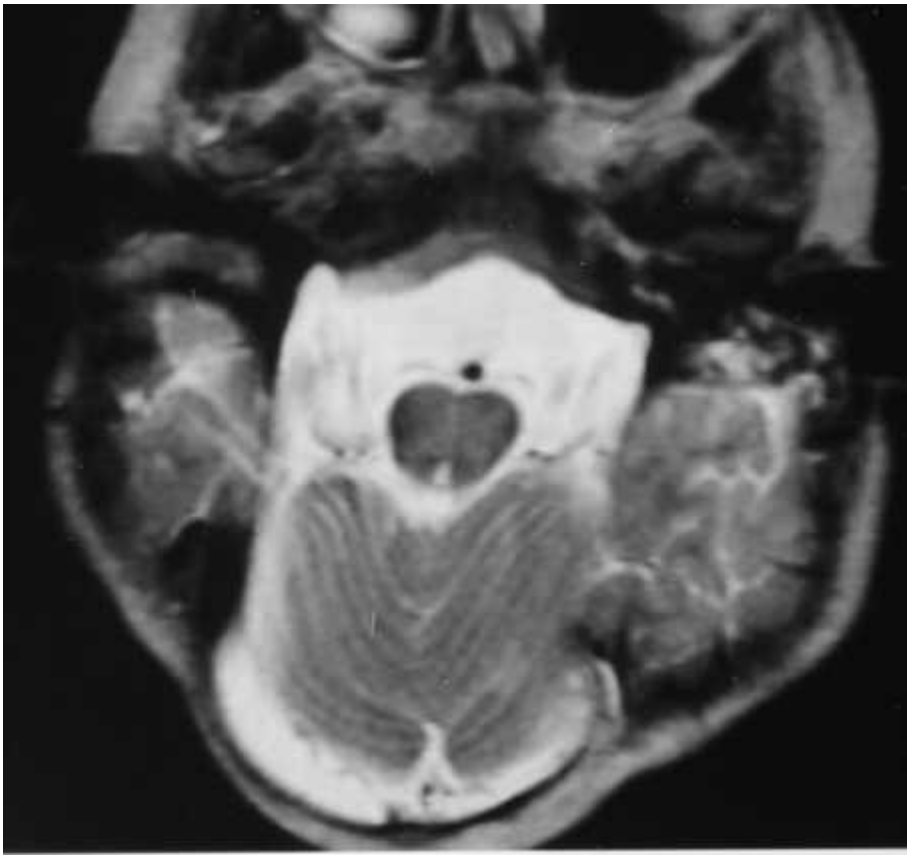
Description: Sagittal T1-weighted MR image demonstrates herniation of cerebellar hemispheres through a large occipitocervical osseous defect, flattening of anterior border of brainstem, narrowing of fourth ventricle, tectal beaking, low lying confluens sinuum. **Origin:**

b



Description: Axial T1-weighted MR image shows herniation of cerebellar hemispheres through a large occipitocervical osseous defect. **Origin:**

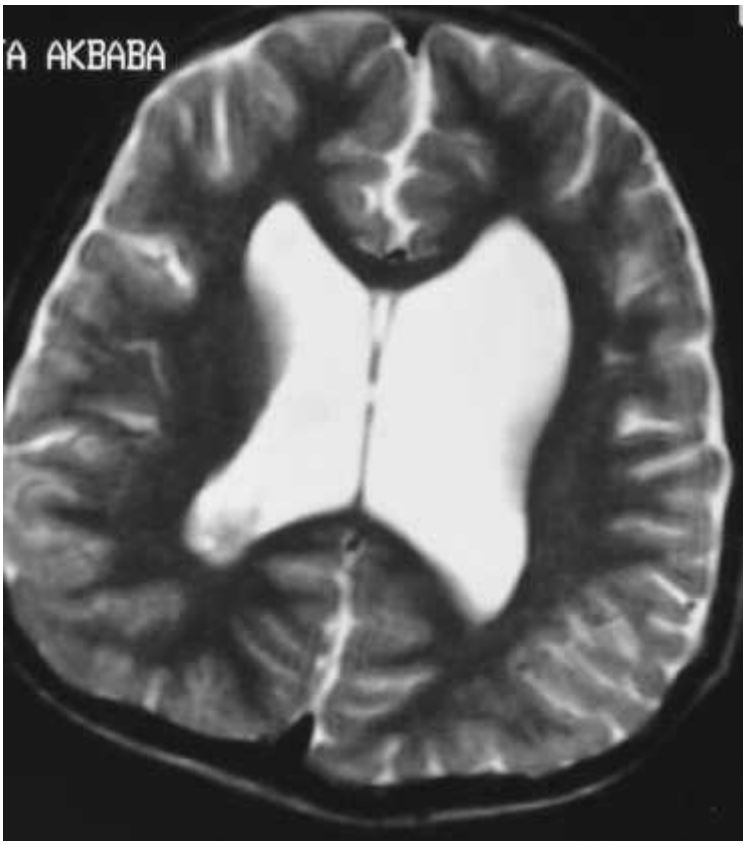
c



Description: Axial T2-weighted MR image shows herniation of cerebellar hemispheres through a large occipitocervical osseous defect. **Origin:**

Figure 2

a



Description: Axial T2–weighted MRI image demonstrates hydrocephalic enlargement of lateral ventricles. **Origin:**