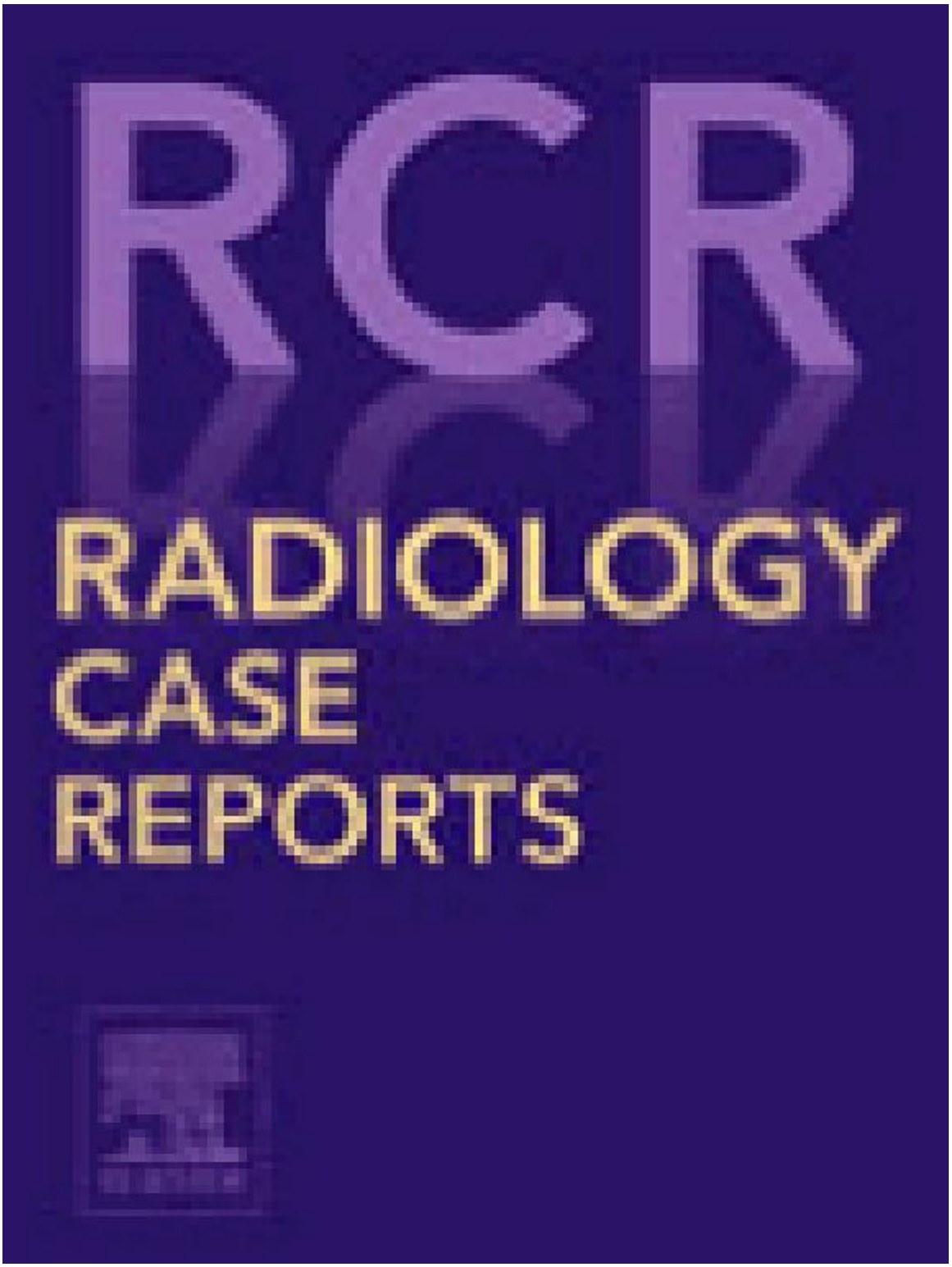
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Chiari malformation type III: Case report and review of the literature

Christina Andica, MD, and Ristaniah D. Soetikno, MD

Chiari malformation type III is an extremely rare anomaly characterized by a small posterior fossa and a low occipital/high cervical encephalocele with herniation of the posterior fossa contents (that is, the cerebellum and/or the brainstem, occipital lobe, and fourth ventricle). We report a case of Chiari malformation type III in a neonate, discuss the etiopathogenetic and radiological features, and review the pertinent literature.

Introduction

Hans Chiari, Professor of Pathology at the German University in Prague, published a series on hindbrain herniations based on autopsy findings in 1891 (1). He described three classes of hindbrain anomalies, including Chiari malformation type III; he found this exclusively in patients with occipital and/or high cervical encephalocele, with herniated dysplastic posterior fossa contents, and other associated anomalies (2, 3)

Chiari malformation type III is the rarest of the Chiari malformations, and it is usually associated with a dismal prognosis--early death or severe disability in long-term survivors (4).

Case report

A two-day-old male child presented to our hospital emergency department with a ruptured occipital encephalocele. He was a full-term normal delivery, born of nonconsanguineous parents. There was neither any history of any medicinal intake nor any evidence of any maternal infection during pregnancy. The mother's ultrasound examination, during the eighth month of pregnancy, was normal. On examination, the child weighed 2.7 kg. There was no retrocollis or any other postural deformity. The child had a normal cry, with no episodes of apnea. There was no cranial nerve palsy, and eye movements were normal without any nystagmus. The tones of the limbs as well as reflexes were normal. On auscultation, the chest was clear, with normal vesicular breath sounds. On local examination, there was a soft multilobulated mass in the occipital region that was not covered by skin, but there was no cerebrospinal fluid leakage (Fig. 1). The hematological and biochemical parameters of the child were normal. Skull x-ray and

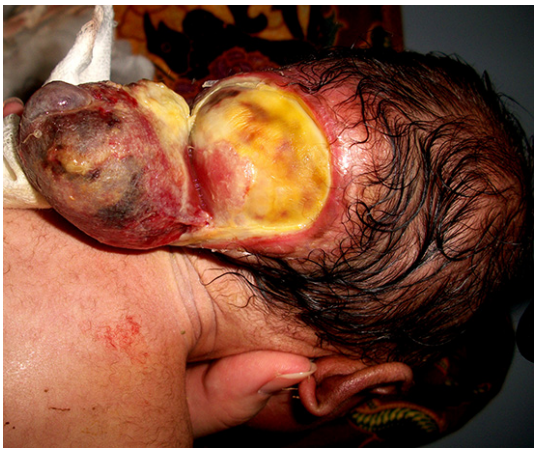

Figure 1. Neonate with Chiari malformation type III. 3D reformatted CT scan shows lacunar skull and a large defect in the occipital regional striae.

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