



Chiari Type Iii Malformation: Clinical Image

Feyza Karagoz Guzey*, Burak Eren and Ilker Gulec

Department of Neurosurgery, Health Sciences University, Bagcilar Training and Research Hospital, Neurosurgery Clinic, Istanbul, Turkey

Clinical Image

Chiari Malformation Type III (CMIII) is a very rare anomaly characterized by herniation of the infra- and sometimes supratentorial structures into a sub-occipital/upper cervical encephalocele [1]. The presentation spectrum ranges from asymptomatic newborn to severe deficits [2]. In Figure 1, magnetic resonance imaging (MRI) of a 6 month old baby was shown. There was only microcephaly and a small suboccipital sac covered by skin when she was born. However, there were serious brain anomalies on her radiological investigations. Surgery without necessary precautions may be catastrophic because of concomitant venous sinus anomalies in these babies. In our patient, although there was only a small sac on physical examination, the lateral sinuses encircled the suboccipital bony defect probably because of tentorial agenesis (Figure 2). Meticulous evaluation of radiological investigations is mandatory in patients with occipital encephaloceles during surgical planning to avoid catastrophic consequences.

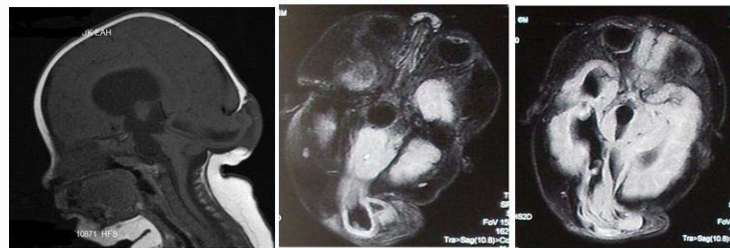


Figure 1: The MRI examination of the patient when she was 8 month-old. a-Sagittal, b-axial infratentorial and c-axial supratentorial sections showing extensive cerebellar and partly right occipital herniation into the sac (a), and tentorial agenesis (a).

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*Correspondence:

Feyza Karagoz Guzey, Department of Neurosurgery, Health Sciences University, Bagcilar Training and Research Hospital, Neurosurgery Clinic, Istanbul, Topkapi M Kahal Basi S 46/2 34093 Istanbul, Turkey, Tel: 90-212-4404000; Fax: 90-212-5312617; E-mail: fkarag@yahoo.com

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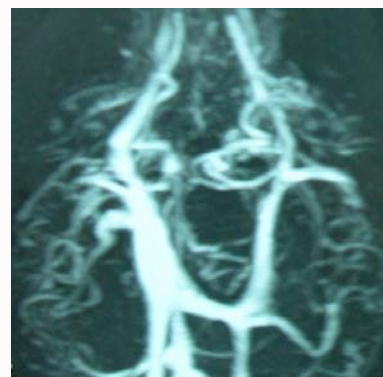


Figure 2: Brain MR Angiography investigation of the patient showing that both lateral sinuses encircled the suboccipital bony defect.

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