Post-surgical Reversible Cerebral Vasoconstriction Syndrome in a pediatric patient with Chiari I malformation

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Introduction

Reversible cerebral vasoconstriction syndrome (RCVS) is a term used to describe a group of disorders sharing angiographic and clinical features, namely a reversible segmental and multifocal vasoconstriction of cerebral arteries and severe headache, with or without focal neurological deficits or seizures (1). The known coexistence of RCVS with posterior reversible encephalopathy syndrome (PRES) suggests a common pathogenetic mechanism, with disturbance of vascular tone (2).

Case Report

A 15 year old girl was admitted a week after a surgical procedure of occipito-cervical osteodural decompression for Chiari I malformation **(fig 1)**, with severe intermittent headache, photofobia, vomiting and blurred vision. The symptoms gradually worsened with left-sided paresthesias and motor impairment, agitated behavior, decreased consciousness and recurrent left hemiclonic seizures requiring admission in intensive care unit (ICU). EEG recordings documented intermittent diffuse rhytmic slow waves with anterior predominance **(fig 2)**; brain magnetic resonance (MR) revealed left parieto-occipital and right parasagittal, right frontal and right periventricular FLAIR hyperintense non-enhancing lesions **(fig 3,)**, hyperintense in diffusion weighted imaging (DWI) **(fig 3)**. Brain MR angiography documented normal venous vessels and segmental narrowing in bilateral middle cerebral (MCA) and posterior cerebral arteries (PCA); cerebral angiography demonstrated the characteristic "string of beads" pattern in the same arterial vessels **(fig 4**.) CSF analysis revealed cellular pleiocytosis (300 lymphocytes), normal glucose and protein levels; coltural examination and PCR for viral nucleic acids on CSF were negative. Serum autoimmunity was negative. Echocardiogram was normal. Supportive measures and levetiracetam were administered with a good control of the seizures; the patient was discharged from ICU with residual mild left-sided hemiparesis and executive cognitive problems. Follow-up MRI at six months showed persistent T2 hyperintense lesions in the right frontal lobe, DWI negative **(fig 3,)**, and resolution of arterial narrowing on cerebral angiography. EEG normalized **(fig 2)**.

Results

The multidistrectual monophasic ischemic injury in watershed regions based on vasogenic edema and vasospasm with the typical angiographic reversible vasospasm pattern and the benign clinical evolution supported the diagnosis of RCVS. Differential diagnosis with paradoxical embolism and particularly with primary or secondary SNC angiitis have been ruled out. Rare cases of children with RVCS and thunderclap headache and few pediatric cases of PRES after posterior fossa surgery have been reported (3); the manipulation of brainstem areas during neurosurgical procedures and the increase of circulating cytokines and inflammatory molecules may contribute to the pathogenesis of RCVS and PRES.

Conclusions

The diagnosis of RCVS should be considered in patients with recurrent thunderclap headache followed by focal neurological deficit and/or seizures; this case may contribute to the post surgical pediatric case-series of RCVS .

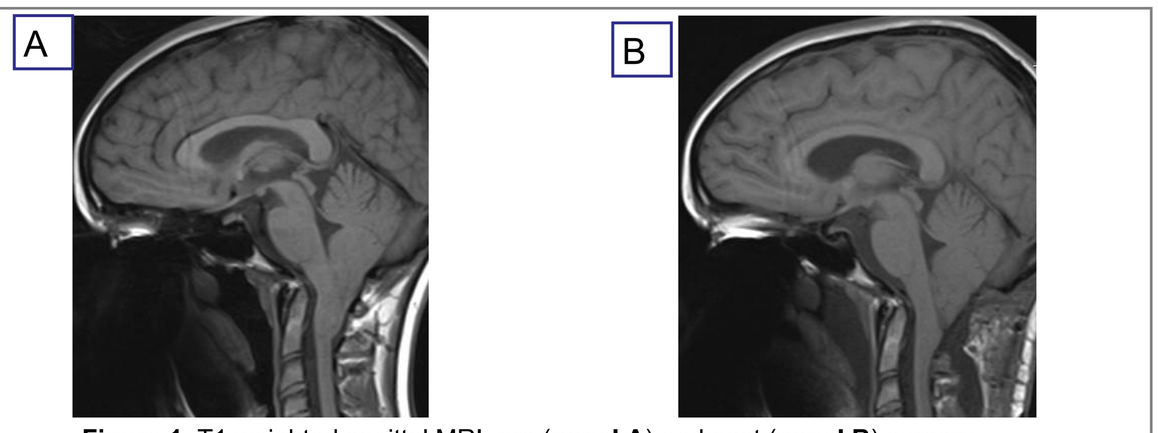

Figure 1. T1 weighted sagittal MRI, pre (**panel A**) and post (**panel B**) surgery

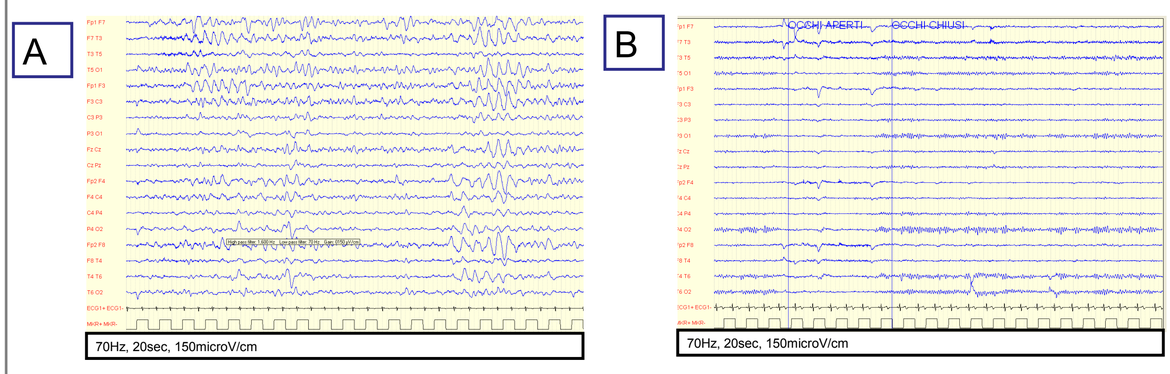

Figure 2. EEG recordings at presentation (**panel A**) and at six month follow-up (**B**)

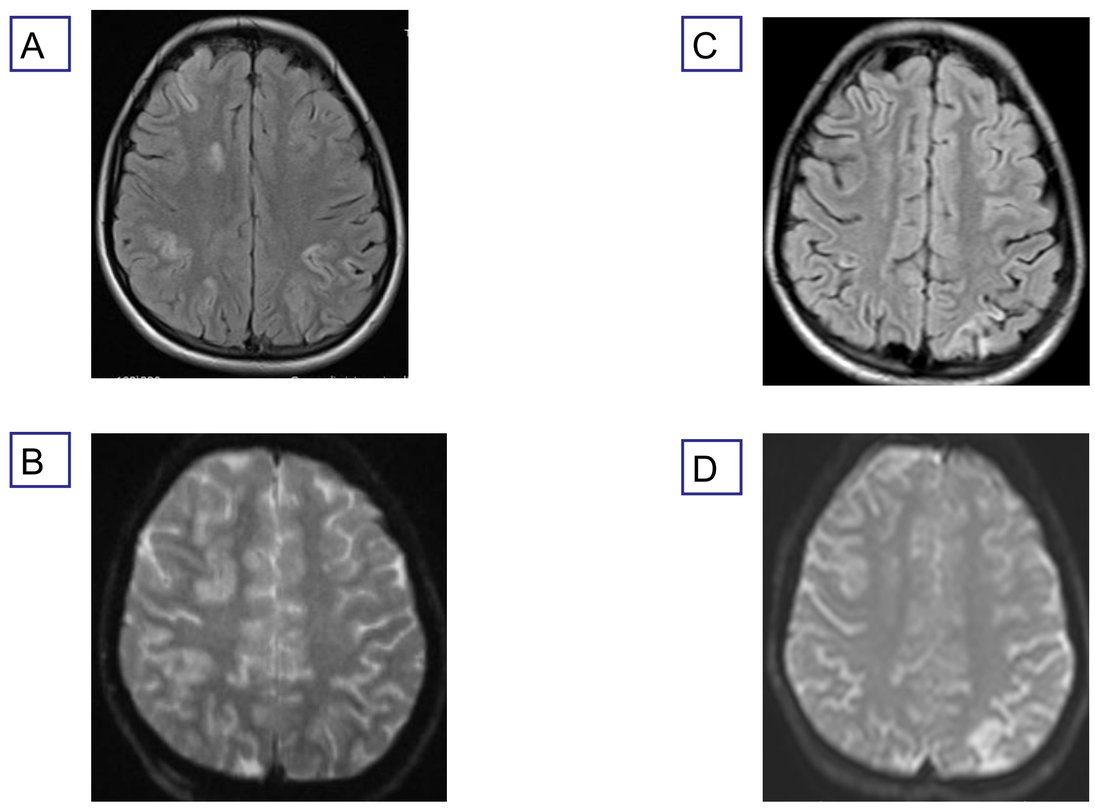

Figure 3. Panel A : FLAIR weighted coronal Brain MRI at presentation**. Panel B**: : DWI weighted coronal Brain MRI at presentation. **. Panel C :** FLAIR weighted coronal Brain MRI at six month follow-up. **Panel D:** DWI weighted coronal Brain MRI at six month follow-up

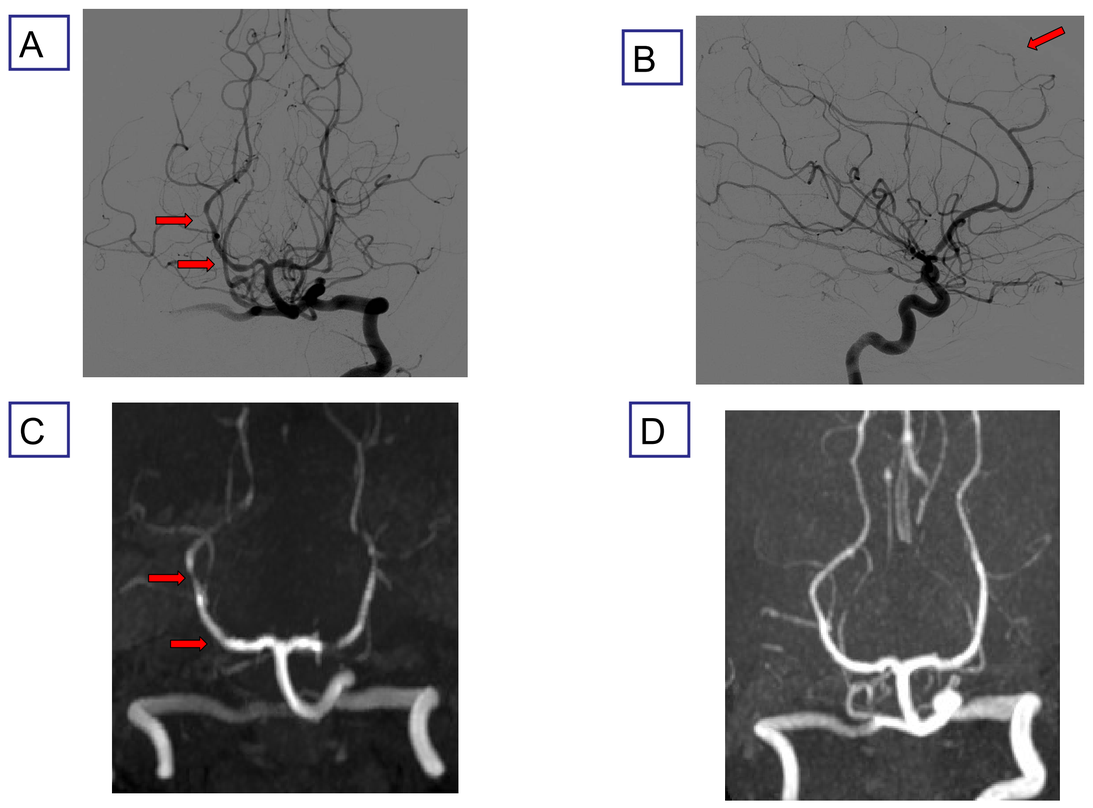

Figure 4: Panel A-B: : cerebral angiography at presentation. **Panel C:** : angio-MRI at presentation **Panel D:** : angio-MRI at six month follow-up

References

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