Case report
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Reversible splenial lesion syndrome associated with severe pediatric idiopathic intracranial hypertension: a case report

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Introduction. Reversible lesions in the splenium of the corpus callosum are a recognized radio-clinical entity, which is rarely associated with idiopathic intracranial hypertension, especially in children. We present a case.

Case report. We describe the case of an 11-year-old prepubescent female patient with no significant medical history. She presented with idiopathic intracranial hypertension syndrome and exhibited a nodular lesion in the splenium of the corpus callosum on the MRI scan. Initial medical treatment with acetazolamide and a subtractive lumbar puncture failed to improve the patient's condition. Consequently, an emergency ventriculoperitoneal shunt procedure was performed, resulting in a favorable clinical outcome. A follow-up MRI was conducted six months later, revealing normalization of the imaging results.

Conclusion. Further studies are needed to understand the pathophysiology of reversible lesions in the splenium of the corpus callosum during idiopathic intracranial hypertension.

Keywords: idiopathic intracranial hypertension, reversible splenial lesion syndrome, lumbar puncture

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Syndrome de lésion spléniale réversible associé à une hypertension intracrânienne idiopathique pédiatrique sévère : étude de cas

Introduction. La lésion réversible du splénium du corps calleux est une entité radio-clinique connue, rarement associée à une hypertension intracrânienne idiopathique notamment chez l'enfant. Nous présentons un cas.

Étude de cas. Nous rapportons le cas d'une patiente prépubère de 11 ans, sans antécédents notables, qui présentait un syndrome d'hypertension intracrânienne idiopathique avec une lésion nodulaire du splénium du corps calleux à l'IRM. Un traitement médical à base d'acétazolamide et une ponction lombaire soustractive n'ayant pas amélioré l'état de la patiente, une dérivation ventriculo-péritonéale a été réalisée en urgence. Une évolution clinique favorable s'en est suivie. Une IRM de suivi a été réalisée six mois plus tard, avec une normalisation des résultats de l'imagerie.

Conclusion. Des études complémentaires sont nécessaires pour comprendre la physiopathologie des lésions réversibles du splénium du corps calleux au cours de l'hypertension intracrânienne idiopathique.

Mots-clés : hypertension intracrânienne idiopathique, syndrome de lésion spléniale réversible, ponction lombaire

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Abbreviations

CSF: cerebrospinal fluid
DWI: diffusion-weighted imaging
IIH: idiopathic intracranial hypertension
LP: lumbar puncture
MRI: magnetic resonance imaging
RESLES: reversible splenial lesion syndrome
SCC: splenium of the corpus callosum
SWI: susceptibility weighted imaging

1. Introduction

Reversible lesions in the splenium of the corpus callosum (SCC) identified through magnetic resonance imaging (MRI) represent a unique radio-clinical scenario with various causes but rarely associated with idiopathic intracranial hypertension (IIH) in children [1]. We report a case of IIH with a reversible splenial lesion syndrome (RESLES) in an 11-year-old patient.

2. Case report

In April 2019, an 11-year-old prepubertal patient with no previous history of any kind was admitted with an acute headache that had been evolving for 15 days prior to hospitalization, associated with binocular diplopia and projective vomiting.
Clinical examination revealed a body mass index of 14.7 Kg/m² and bilateral limitation of ocular abduction, without neck stiffness or other signs of focalization. Ophthalmological examination revealed a visual acuity of 0.40 and a bilateral stage 3 papilledema. Brain MRI showed indirect signs of IIH, including optic nerve tortuosity, optic disc protrusion, and an arachnoidocele. Additionally, a 9 mm nodular lesion was observed in the splenium of the corpus callosum (SCC), which appeared hypointense on T1, hyperintense on T2 and SWI sequences, with apparent diffusion coefficient restriction (Figure 1). Lumbar puncture (LP) with measurement of intracranial pressure (ICP) showed an elevated opening pressure of 60 cm H_2O. The study of cerebrospinal fluid (CSF) was normal, including protein (35 mg/dL), glucose (56 mg/dL), cell count (no white and red blood cells). An exhaustive biological work-up, including hormonal assessment (growth hormone, TSH, FSH, LH and prolactinemia), vitamin levels, renal and hepatic assessment, and electrolytic assessment, was unremarkable. The diagnosis of IIH was made according to the modified Dandy 2013 criteria [2]. The treatment consisted of a therapeutic LP, removing 30 mL of CSF, along with acetazolamide. There was minimal improvement in the first 48 hours, followed by worsening of headache intensity, necessitating a second LP. This second LP found a very high opening pressure of 130 cm H_2O, indicating the need for an emergency ventriculoperitoneal shunt to preserve visual function.
The surgery allowed for a complete regression of the symptoms, with normalization of the fundus after 15 days, as well as regression of bilateral limitation of ocular adduction and normal visual acuity. Follow-up MRI angiography at six months revealed no evidence of the SCC lesion, thus confirming a reversible lesion of the SCC.

3. Discussion

IIH is a rare neurological disorder in children, and the exact pathophysiological mechanism remains unknown. Its prevalence in the pediatric population increases with age and affects twice as many children aged 12 to 17 years as those aged 2 to 12 years [3]. Obesity is an important risk factor in IIH, as highlighted by some studies where BMI was significantly higher in the post-pubertal group [4]. Other studies have argued that this is not the case in prepubertal patients [5]. RESLES is a radioclinical entity characterized by the presence of a reversible lesion involving the SCC, with a decrease in the ADC of the lesion on brain MRI. Its etiologies are numerous, including head trauma, acute disseminated encephalomyelitis, alcoholism, seizures, and some drugs. Due to this etiological heterogeneity, no common pathophysiological mechanism has been identified yet [6]. Several hypotheses exist, including intramyelin edema, inflammatory infiltrates, oxidative stress, neuroaxonal injury, and cytotoxic edema [7].
The mean age of the populations in the RESLES studies is estimated to be 25.6 years, suggesting the rarity of this syndrome in the pediatric population [6]. Only one case has been reported of an 11-year-old boy with IIH associated with hypothyroidism, which regressed after 6 weeks following a therapeutic lumbar puncture and treatment with acetazolamide [3]. Most studies report a time to clearance of the reversible SCC lesion of 10 to 32 days, and in our patient a follow-up brain MRI performed six months later was normal [6]. A therapeutic LP, along with medical treatment with acetazolamide, did not resolve the symptoms. Ventriculoperitoneal shunting was the only way to preserve the optic nerves, especially in the absence of other techniques, such as fenestration of the optic nerves or venous sinus stenting. The rarity of the association between ICH and RESLES does not allow for the establishment of a causal relationship between the severity of the clinical picture and the presence of RESLES. Further studies are needed to understand this phenomenon in IIC.

4. Conclusion

The presence of reversible lesions of the splenium of the corpus callosum during IIC is a rare situation requiring further studies to understand its physiopathology and to evaluate the potential of these lesions as a radiological indicator of severity.

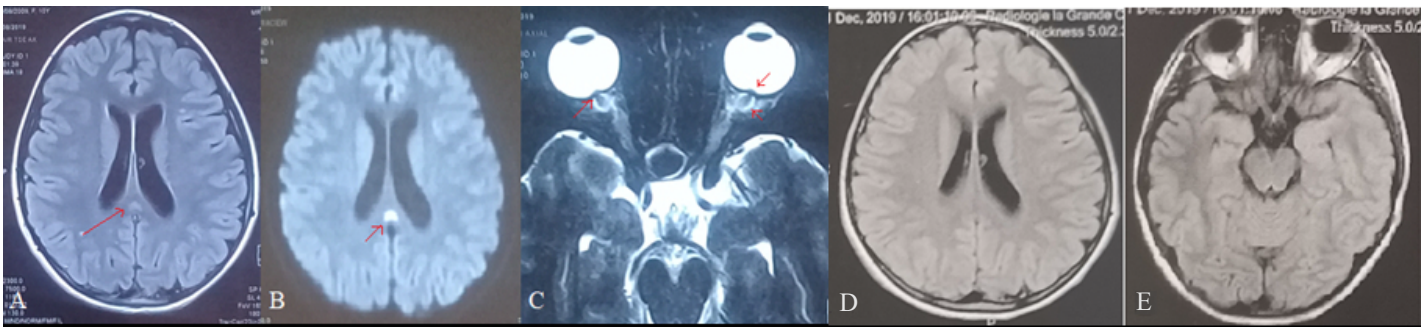

Figure 1. Brain magnetic resonance imaging at diagnosis and 6 months follow-up. (A) FLAIR sequence: 9 mm nodular hypersignal of the splenium of the corpus callosum (arrow). (B) Diffusion-weighted imaging (DWI): hyperintense signal (arrow). (C) T2 thin section sequence through the optic nerves showing the indirect signs of idiopathic intracranial hypertension: protrusion of the optic nerve papilla, tortuosity of the optic nerves with dilatation of their sheaths, and flattening of the posterior pole of the eyeball (arrows). (D), (E) 6-month follow-up brain MRI shows complete regression of the splenium lesion of the corpus callosum, as well as the indirect signs of idiopathic intracranial hypertension.

Statements

Ethics statement. The authors confirm that this work complies to the journal's guidelines on issues involved in ethical publication, which state that written informed consent was obtained from individual participants involved in case studies or through a surrogate where appropriate.

Declaration of interest. The authors report no conflicts of interest.

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