Radiology Case Reports 16 (2021) 3969–3972

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case report

Posterior reversible encephalopathy syndrome revealing Takayasu's arteritis in a child ☆

Abdelhamid Jadib, Doctor; Radiology resident, Siham Salam, Radiology professor, Yassine Harmoumi, Doctor; Radiology resident, Lamiaa Chahidi El Ouazzani, Doctor; Radiology resident, Othmane Soussi, Doctor; Radiology resident, Dalale Laoudiyi, Radiology professor, Kamilia Chbani, Radiology professor, Lahcen Ouzidane, Radiology professor*

Pediatric Radiology Division, Ibn Rochd University Hospital, Faculty of Medicine and Pharmacy of Casablanca, Morocco, 1, quartiers des hôpitaux, Casablanca 20100, Morocco

ARTICLE INFO

Article history:
Received 1 July 2021
Revised 14 September 2021
Accepted 16 September 2021

Keywords:
Posterior reversible encephalopathy syndrome
Takayasu's arteritis
Imaging

ABSTRACT

Posterior Reversible Encephalopathy Syndrome (PRES) is a rare complication of Takayasu's Arteritis. We report the case of an 11-year-old girl who presented with a tonic-clonic seizure and loss of consciousness, without fever. Imaging revealed characteristic white matter edema of the occipital and parietal lobes, in keeping with PRES. Further imaging demonstrated right renal artery stenosis and wall thickening of the abdominal aorta. The combination of hypertension, the discrepancy of blood pressure recordings between upper limbs, and imaging abnormalities of the aorta and the left renal artery led to the diagnosis of PRES secondary to Takayasu's Arteritis. Treatment with oral corticosteroids, azathioprine, amlodipine, and propranolol resulted in the complete resolution of the patient's symptoms and imaging abnormalities.

Introduction

Posterior Reversible Encephalopathy Syndrome (PRES) is a rare complication of Takayasu's Arteritis (TA) [1]. It's revealed with symptoms of headaches, seizures, visual disturbances, confusion, or altered consciousness [1–3]. On imaging, it is characterized by vasogenic edema in the occipital, parietal, frontal and temporal lobes [3]. Early diagnosis and treatment of PRES are essential to prevent complications such as hemorrhage, hydrocephalus, brainstem compression, and death [4]. Prompt diagnosis and appropriate treatment of PRES can achieve complete resolution of symptoms and imaging abnormalities [3,5].

This case history outlines the presentation of an 11-year-old girl with signs and characteristic findings on imaging of PRES secondary left renal artery stenosis due to TA.

☆ Competing Interests: The authors have declared that no competing interests exist.

* Corresponding author. A. Jadib.
E-mail address: jadibabdelhamid@gmail.com (A. Jadib).

https://doi.org/10.1016/j.radcr.2021.09.034

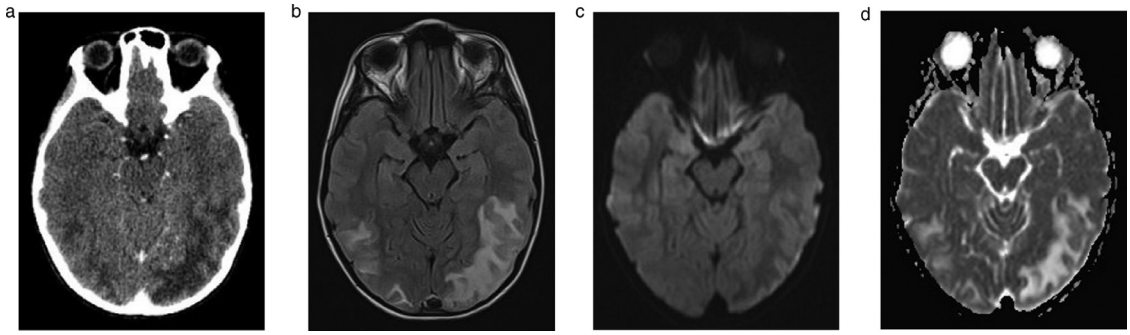

Fig. 1 – Brain CT scan (A) showing areas of ill-defined hypoattenuation involving predominantly the subcortical white matter in both parietal and occipital lobes, with no mass effect, contrast enhancement, or cranial hemorrhage. On brain MRI, the described areas on CT scan are generating high signal on T2 FLAIR images (B), low signal on DWI with high apparent diffusion coefficient (ADC) (C, D).

Case presentation

An 11-year-old girl with unremarkable medical history was admitted to the local pediatric emergency department after losing consciousness for 20 minutes following a generalized tonic-clonic seizure. She had a 3-day history of nausea, abdominal pain, headaches, and a more recent onset of blurred vision, without fever.

On admission, tonic-clonic seizure stopped after intrarectal diazepam, her body temperature was 37.4°C, heart rate was 121/min, blood pressure was 161/109 mm Hg, with discrepancy of blood pressure recordings between upper limbs (161/109 mm Hg at the right upper limb and 148/101mm Hg at the left upper limb), and glycemia was 6.1 mmol/L (109.91 mg/dl). Reference range was 2.5-5.3 mmol/L (45-96 mg/dL). She rapidly regained consciousness. The neurological examination was normal. The end of the physical examination and standard blood tests were normal.

Radiologic exams were done on the same day. Brain CT scan showed areas of ill-defined hypoattenuation involving predominantly the subcortical white-matter in both parietal and occipital lobes, with no mass effect, contrast enhancement, or cranial hemorrhage (Fig. 1A). On brain MRI, the described areas on CT scan were generating low signal on T1-weighted images and high signal on T2-weighted and T2 Fluid-Attenuated-Inversion-Recovery (FLAIR) images, high apparent diffusion coefficient with no hemorrhage (Fig. 1B, C and D). These findings, together with neurological manifestations and hypertension, suggested a PRES.

On abdominal ultrasound, the left kidney was small (8.5 cm of long axis), and the right kidney was hypertrophic (11cm of long axis).

Doppler examination of the left renal artery and its branches revealed a low resistance index (0.41) with flattened arterial curves, associated with left renal atrophy, suggestive for renal artery stenosis (Fig. 2A).

Multiphase CT scan of the renal tract demonstrated concentric mural thickening of the abdominal aorta and the proximal left renal artery causing its tight stenosis (Figs. 2B and C).

Tc-99m Dimercaptosuccinic Acid renal scintigraphy showed a non-functioning left kidney with a normal right kidney.

TA was diagnosed using the EULAR/PRINTO/PRES criteria, after excluding Antineutrophil Cytoplasmic Antibodies vasculitis, a renal disease, coarctation of the aorta, and endocrine diseases: onset under 40 years, a discrepancy of blood pressure recordings between upper limbs and narrowing of the left renal artery.

Continuous infusion of nicardipine was given. All neurological manifestations were resolved within 2 days, with the normalization of blood pressure.

Then, she was switched to long-acting oral medications, including amlodipine and propranolol.

The aetiological treatment of TA consisted of long-acting oral immunosuppressive medications, including azathioprine and corticosteroid. The left nephrectomy is foreseen since the left kidney is non-functioning.

Posttreatement MRI brain performed 11 months later, showed total resolution of PRES findings (Fig. 3).

Discussion

TA is a granulomatous large vessel vasculitis that predominantly involves the aorta and its major branches. It tends to affect younger patients, aged 10-30 years old, with a strong female predominance [1]. Clinical presentation is very variable depending on the territory of vascular involvement. PRES is a rare complication [6]. An extensive literature review revealed only 13 other cases of PRES secondary to TA.

The diagnosis of TA can be made when 3 of the following 6 criteria are present (EULAR/PRINTO/PRES criteria): onset under 40 years, claudication of an extremity, reduced peripheral pulse, a discrepancy of blood pressure recordings between upper limbs, bruit over the aorta or subclavian arteries and/or evidence of narrowing of the aorta or its primary branches [7].

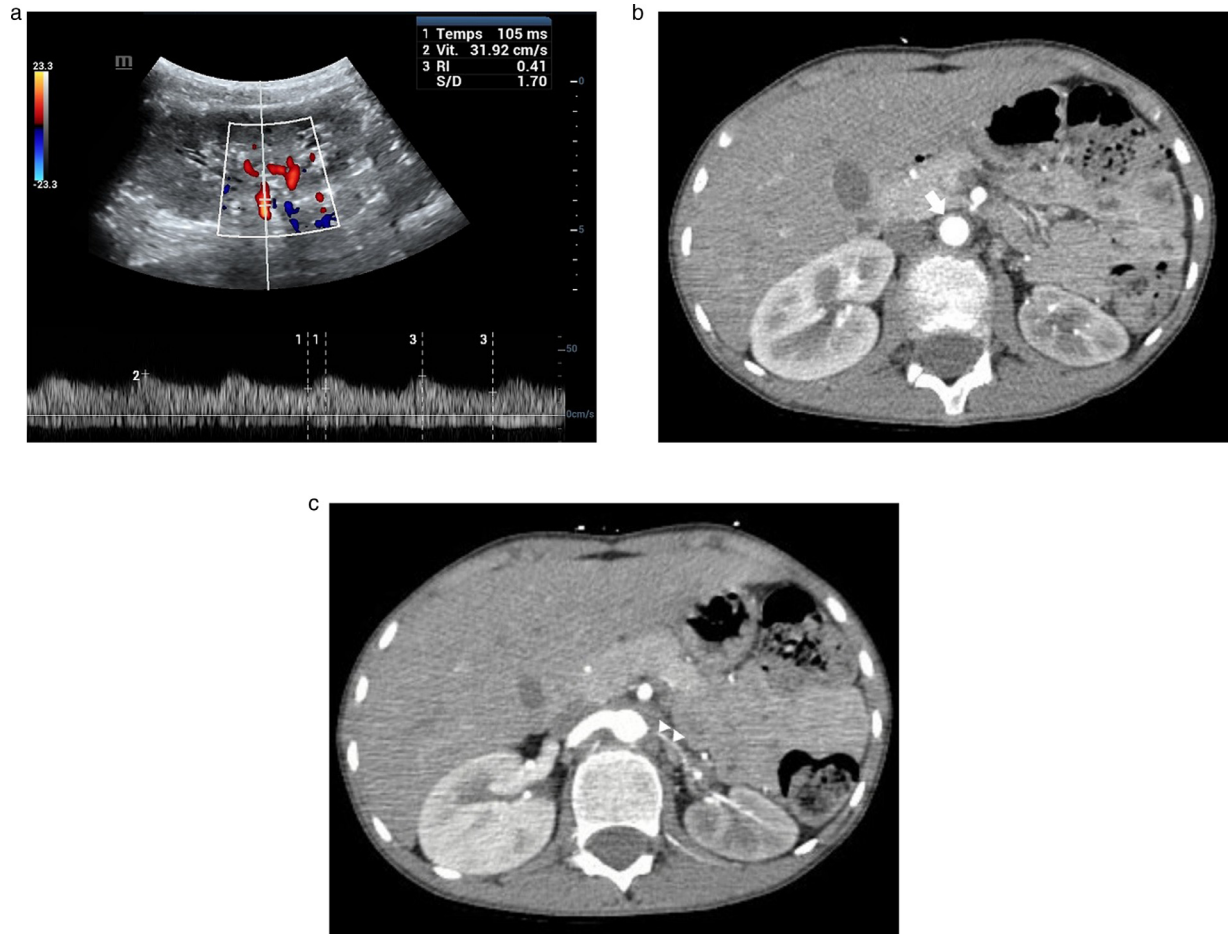

Fig. 2 – Renal Doppler examination revealing a low resistance index (0.41) with flattened arterial curves in the distal portion of the left renal and interlobar arteries (A), suggestive for renal artery stenosis. Multiphase CT scan of the renal tract demonstrating concentric mural thickening (arrow) of the abdominal aorta (B) and the proximal portion of the left renal artery (arrow heads) causing its tight stenosis (C).

Typical imaging features include mural thickening of the involved arteries associated with stenosis. Occlusion, ectasis, and aneurysms of the vessels are less common [8].

PRES is rare in childhood [9]. Clinical features include headaches, nausea, visual disturbances, seizures, and consciousness disorders, associated with neuro-imaging abnormalities predominating in the parieto-occipital lobes [10].

Various clinical settings can precipitate the syndrome, and hypertension is the most frequent cause. The other main conditions associated with PRES are hematological and autoimmune diseases, hemolytic–uremic syndrome, chronic renal failure, hepatitis C, HIV infection, blood transfusion, and immunosuppressive drug therapy [11,12].

The mechanism of PRES is not well understood. It is thought to be related to the blood-brain barrier disruption with fluid transudation due to high blood pressure, exceeding the cerebral vasculature's auto-regulatory capacity, which leads to focal vasodilatation vasoconstriction, particularly in arterial watershed zones [13].

The main imaging feature of PRES is subcortical and cortical vasogenic edema. It is best detected using T2 (FLAIR) sequences. Parieto-occipital involvement is classic and seen in the majority of cases [14].

Vasogenic edema is usually hypo- or iso-intense on Diffusion-Weighted Imaging (DWI) and hyperintense on corresponding apparent diffusion coefficient maps. Occasionally, the signal is slightly hyperintense on DWI due to T2 shine-through. Restricted diffusion is seen in 11%-26% of cases. It reflects cytotoxic edema and may indicate progression to infarction and irreversibility, associated with poor outcomes [15].

Hemorrhagic complications occur within the area of brain parenchyma affected by edema. They range from 15%-65% and include microhemorrhages, hematoma with masse effect, and or subarachnoid hemorrhage [15].

The presence of subcortical and cortical edema on T2 FLAIR in the appropriate clinical context is generally sufficient for the diagnosis. Contrast imaging is not mandatory and may show a gyriform or leptomeningeal pattern contrast enhancement [16].

Both the clinical and radiologic findings in PRES are mainly reversible, especially if diagnosed and treated promptly, which was the case for our patient. The features associated with

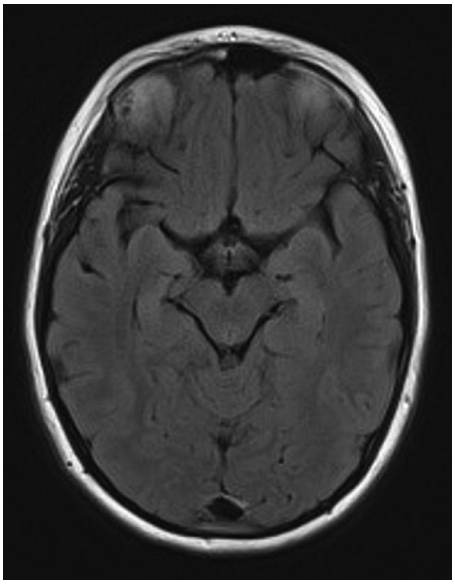

Fig. 3 – Posttreatement axial FLAIR MRI brain demonstrating a total resolution of the subcortical white matter signal abnormality.

worse clinical outcomes in PRES, impacting the reversibility of the radiologic findings, are extensive vasogenic edema, mass effect, diffusion restriction, and hemorrhage on initial imaging [17].

Nephrectomy is the best treatment for nephrogenic hypertension in children with a unilateral non-functioning kidney and a normal contralateral kidney, as foreseen in our patient [18].

Patient consent

Written and informed consent for publication of the case was obtained from the patient.

REFERENCES

[1] Zaki SA, Chavan V, Shanbag P. Unusual presentation of Takayasu's arteritis as posterior reversible encephalopathy syndrome. Ann Indian Acad Neurol 2011;14:214–16. doi:10.4103/0972-2327.85900.

[2] Camara-Lemarroy CR, Lara-Campos JG, Perez-Contreras E, Rodríguez-Gutiérrez R, DA Galarza-Delgado. Takayasu's arteritis and posterior reversible encephalopathy syndrome: a case-based review. Clin Rheumatol 2013;32:409–15. doi:10.1007/s10067-012-2151-9.

[3] Li K, Yang Y, Guo D, Sun D, Li C. Clinical and MRI features of posterior reversible encephalopathy syndrome with atypical regions: a descriptive study with a large sample size. Front Neurol 2020;11:194. doi:10.3389/fneur.2020.00194.

[4] Fugate JE, Rabinstein AA. Posterior reversible encephalopathy syndrome: clinical and radiological manifestations, pathophysiology, and outstanding questions. Lancet Neurol 2015;14:914–25. doi:10.1016/S1474-4422(15)00111-8.

[5] Shankar J, Banfield J. Posterior reversible encephalopathy syndrome: a review. Can Assoc Radiol J 2017;68:147–53. doi:10.1016/j.carj.2016.08.005.

[6] Wakida K, Morita H, Sakurai T, Nishida H. [A case of posterior reversible encephalopathy syndrome associated with Takayasu's arteritis]. Rinsho Shinkeigaku 2016;56:174–9. doi:10.5692/clinicalneurol.cn-000815.

[7] Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy SM, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum 1990;33:1129–34. doi:10.1002/art.1780330811.

[8] Zhu FP, Luo S, Wang ZJ, Jin ZY, Zhang LJ, Lu GM. Takayasu arteritis: imaging spectrum at multidetector CT angiography. Br J Radiol 2012;85:e1282–92. doi:10.1259/bjr/25536451.

[9] Endo A, Fuchigami T, Hasegawa M, Hashimoto K, Fujita Y, Inamo Y, et al. Posterior reversible encephalopathy syndrome in childhood: report of four cases and review of the literature. Pediatr Emerg Care 2012;28:153–7. doi:10.1097/PEC.0b013e3182442fe8.

[10] Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, et al. A reversible posterior leukoencephalopathy syndrome. N Engl J Med 1996;334:494–500. doi:10.1056/NEJM199602223340803.

[11] Nightingale S, Wood C, Ainsworth J. The posterior reversible encephalopathy syndrome in HIV infection. BMJ Case Rep 2012 2012. doi:10.1136/bcr.01.2012.5647.

[12] Abenza-Abildua MJ, Fuentes B, Diaz D, Royo A, Olea T, Aguilar-Amat MJ, et al. Cyclophosphamide-induced reversible posterior leukoencephalopathy syndrome. BMJ Case Rep 2009 2009. doi:10.1136/bcr.07.2008.0467.

[13] Dinsdale HB, Robertson DM, Haas RA. Cerebral blood flow in acute hypertension. Arch Neurol 1974;31:80–7. doi:10.1001/archneur.1974.00490380028002.

[14] Bartynski WS, Boardman JF. Distinct imaging patterns and lesion distribution in posterior reversible encephalopathy syndrome. AJNR Am J Neuroradiol 2007;28:1320–7. doi:10.3174/ajnr.A0549.

[15] Brady E, Parikh NS, Navi BB, Gupta A, Schweitzer AD. The imaging spectrum of posterior reversible encephalopathy syndrome: a pictorial review. Clin Imaging 2018;47:80–9. doi:10.1016/j.clinimag.2017.08.008.

[16] McKinney AM, Short J, Truwit CL, McKinney ZJ, Kozak OS, SantaCruz KS, et al. Posterior reversible encephalopathy syndrome: incidence of atypical regions of involvement and imaging findings. AJR Am J Roentgenol 2007;189:904–12. doi:10.2214/AJR.07.2024.

[17] Schweitzer AD, Parikh NS, Askin G, Nemade A, Lyo J, Karimi S, et al. Imaging characteristics associated with clinical outcomes in posterior reversible encephalopathy syndrome. Neuroradiology 2017;59:379–86. doi:10.1007/s00234-017-1815-1.

[18] Johal NS, Kraklau D, Cuckow PM. The role of unilateral nephrectomy in the treatment of nephrogenic hypertension in children. BJU Int 2005;95:140–2. doi:10.1111/j.1464-410X.2004.05266.x.