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Review article

Isolated pons involvement in Posterior Reversible Encephalopathy Syndrome: Case report and review of the literature

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ABSTRACT

Background: Posterior Reversible Encephalopathy Syndrome (PRES) is a clinical-radiological syndrome, usually reversible and with a favorable prognosis, which recognizes a variety of etiologies and clinical patterns and is likely due to an impairment in cerebral blood flow autoregulation. It is typically characterized by subcortical, predominantly parieto-occipital, vasogenic brain oedema in patients with acute-subacute neurological symptoms. Infratentorial oedema on neuroimaging has been mostly described in association with the typical supratentorial pattern and seldom as isolated.
Case report: We report a case of PRES with isolated pons involvement on MRI. A woman affected by Turner syndrome, epilepsy, slight mental deficiency, obesity and hypothyroidism, experienced a progressive gait and standing impairment, worsening in the last 2 weeks. At admission blood pressure was 220/110 mmHg. Brain MRI showed a wide FLAIR signal hyperintensity on T2-weighted sequences affecting the entire pons, without contrast enhancement. Clonidine, doxazosine, furosemide and telmisartan were effective in restoring normal blood pressure. Pons hyperintensity completely resolved on MRI 3 weeks later, together with return to normal neurological examination.
Conclusions: Though isolated infratentorial involvement in PRES recognizes several causes, hypertension, which is a common feature in Turner syndrome, would have played a key role in our case with solely pons MRI T2-hyperintensity.

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1. Introduction

Posterior Reversible Encephalopathy Syndrome (PRES) is a clinical condition likely sustained by an impaired neurovascular unit autoregulation of the cerebral blood flow which, in turn, leads to endothelial dysfunction and vasogenic brain oedema. It recognizes different etiologies. Clinical presentation varies widely for both features and severity across reported case series and, at times, does not match either with the pattern or the amount of brain oedema on MRI [1]. Bartynski WS et al. [2] reported three major patterns consistent for typical PRES imaging: holohemispheric watershed (linear vasogenic oedema at anastomotic border zone spanning the frontal, parietal and occipital lobes with lesser temporal lobes involvement), superior frontal sulcal (involvement of the frontal lobe along the mid to posterior aspect of the

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superior frontal sulcus with no frontal pole extension) and dominant parietal-occipital (vasogenic oedema of the parietal and occipital cortex and white matter, variably extended to the temporal lobes). Partial or asymmetric expressions of the primary patterns (with, respectively, bilateral or unilateral absence of signal hyperintensity in either the parietal or occipital lobes) are possible. Brainstem involvement is infrequent and usually associated with the typical subcortical parietal-occipital bilateral sites of T2 hyperintensity [3].

2. Case description

A 28 years-old woman, affected by Turner syndrome, epilepsy treated with oxcarbazepine 600 mg/day, slight mental deficiency, obesity and hypothyroidism in therapy with levothyroxine 125 μgr/day, was admitted with a progressive gait and standing impairment, worsening in the last 2 weeks. She had experienced episodes of tachycardia and acute hypertensive crisis in the past years, although blood pressure monitoring and 24-hour electrocardiogram were normal at that time. She was in ovarian hormone replacement treatment (gestodene 0.075 mg + ethinyl estradiol 0.03 mg/day). At admission blood pressure was 220/110 mmHg with heart rate of 120 bpm. Neurological examination showed inability at standing and walking, paraparesis, brisk reflexes at four limbs, and bilateral Babinski sign. Sensitivities were intact. The patient was confused but alert. Furosemide 25 mg was rapidly administered by infusion and transdermal patch clonidine 2.5 mg/week was prescribed. She underwent full blood examination (complete blood count, electrolytes, liver and kidney function, homocisteine, folate, B12 vitamin, vanilmandelic acid and chromogranine A levels, serum and urinary catecholamines and metanephrines, antiviral, bacterial, thrombophilic and autoimmune screenings), CSF examination, abdominal, renal arteries and cardiac ultrasound examinations (searching for causes of secondary arterial hypertension), thoracic aorta MRI (searching for aortic bicuspid valve or aortic arch abnormalities, typical in Turner syndrome, or possible dissecation). All these examinations were normal. Oxcarbazepine dosage and thyroid hormones were in range. 24-hour electrocardiogram showed sinusal tachycardia. A brain MRI at admission showed a wide FLAIR hyperintensity on T2-weighted sequences affecting the entire pons, without contrast enhancement (Fig. 1).

Ovarian hormone replacement treatment was suspended. Clonidine transdermic patch 2.5 mg/week, doxazosine (tapering slowly up to 6 mg/day), furosemide (up to 50 mg/day) and telmisartan (80 mg/day) were effective in restoring normal blood pressure within 10 days together with concomitant return to normal neurological examination. Notably, at present, neither specific antihypertensive drug has been formally studied for the treatment nor randomized clinical trials assessing therapeutic interventions in PRES have been undertaken [1]. The selection of drugs is left to the discretion of the physician and our initial therapeutic goal was to reduce blood pressure by 25% within the first few hours, as recommended [4]. Beta-blockers for sinus tachycardia were initially delayed whereas possible neoplasm of chromaffin tissue was investigated. Repeated serum and urinary dosages of catecholamines, metanephrines, and vanilmandelic acid were normal 2 weeks after admission. A metaiodobenzylguanidine scintigraphy was performed to exclude a paraganglioma-pheochromocytoma and resulted normal. Then atenolol 50 mg/day was introduced successfully.

Pons hyperintensity was completely resolved on MRI performed 3 weeks after admission (Fig. 2).

3. Discussion

PRES with isolated pons involvement has been rarely described [5–10] and only small series in the form of isolated pontine T2 hyperintensity on MRI have been reported [7]. Severe arterial hypertension alone [5] or in association with acute renal failure [7], highly active antiretroviral therapy [6,10] and oxaliplatinum chemotherapy [9] have been described as precipitating factors in clinical settings of chronic renal failure [7,8], arterial hypertension [5], AIDS [6,10] and cancer [9]. To date, a review of seven brainstem PRES by Gao et al. [7] listed only one case with isolated pons lesion and spared supratentorial involvement. In this case dizziness and weakness were reported as clinical features in the context of arterial hypertension and chronic renal failure. The remaining six brainstem PRES cases in Gao and coll. showed a relatively mild clinical presentation wherein, apart from a case with coma, dizziness and instability were most commonly described. To our knowledge, only seven cases of pontine PRES has been published so far, as reported in Table 1.

Overall, they show a dissociation between the mild clinical features, mostly not referable to the brainstem, and the severity of the MRI images [11–14]. This pattern of *clinico-radiological dissociation* may be considered a key feature of brainstem PRES, and represents a clue for

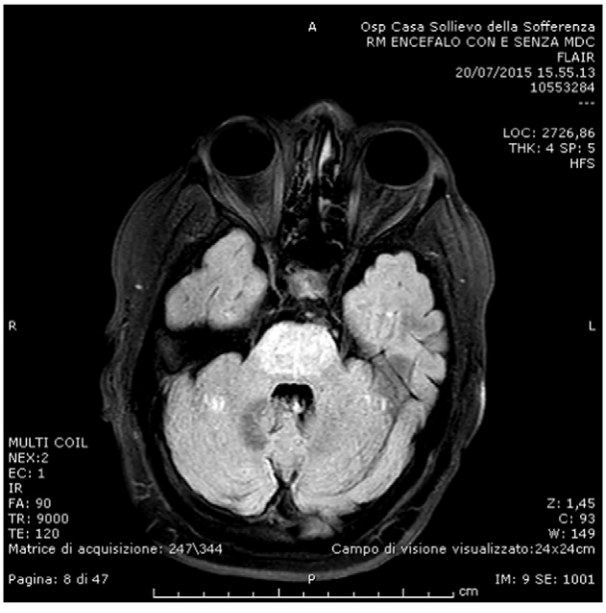

Fig. 1. Isolated involvement of the entire pons in FLAIR sequences at entry MRI.

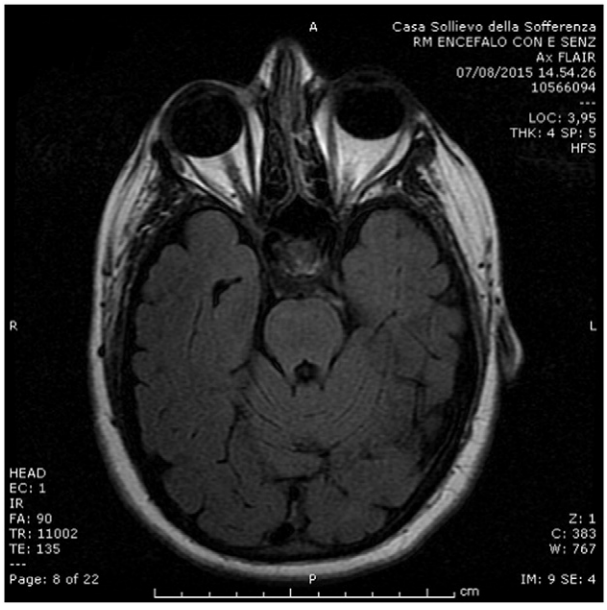

Fig. 2. Complete resolution of pontineT2-hyperintensity at control MRI, after antihypertensive treatment and neurological recovery.