Section: Paediatrics

Case Report

HYPONATREMIC HYPERTENSIVE SYNDROME WITH RENAL ISCHEMIA SECONDARY TO TAKAYASU VASCULITIS: A RARE PEDIATRIC CASE

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ABSTRACT

Takayasu arteritis is a chronic inflammatory large-vessel vasculitis predominantly affecting the aorta and its major branches, commonly seen in young females and frequently involving the renal arteries in pediatric patients from India and the Far East. Hyponatremic hypertensive syndrome is a rare but important clinical entity characterized by severe hypertension, hyponatremia, and polyuria, typically driven by unilateral renal ischemia with excessive renin secretion and pressure natriuresis from the contralateral kidney; early recognition is essential to prevent complications. This report describes a 4-year-old girl admitted to the pediatric intensive care unit with multiple afebrile convulsions and a history of polyuria and polydipsia, found to have absent bilateral brachial and radial pulses, higher lower-limb than upper-limb blood pressure, and grade 1 hypertensive retinopathy. Laboratory evaluation demonstrated persistent hyponatremia with polyuria and normal serum creatinine. Brain MRI showed multiple hypodensities, while CT angiography revealed multiple stenotic lesions of the right and left subclavian arteries and left vertebral artery, along with significant right renal artery stenosis and a small right kidney; echocardiography demonstrated left ventricular hypertrophy consistent with chronic hypertension. These findings supported a diagnosis of Takayasu arteritis complicated by renal artery stenosis and hyponatremic hypertensive syndrome, likely mediated by RAAS activation, contralateral pressure diuresis, and ADH-related volume effects exacerbating hyponatremia. Treatment with corticosteroids and methotrexate led to clinical improvement with normalization of blood pressure and serum sodium; however, repeat CT angiography showed progression of renal artery stenosis, prompting planning for renal angioplasty. The case underscores that refractory stage 2 hypertension with persistent hyponatremia in children should prompt evaluation for renal artery stenosis and vasculitis, with careful pulse examination and early imaging-based diagnosis to enable timely immunosuppression and, when needed, vascular intervention to reduce life-threatening complications and long-term morbidity.

Keywords: Hyponatremic hypertensive syndrome (HHS), Takayasu arteritis (TA), renal artery stenosis, pediatric hypertension, EULAR.

INTRODUCTION

Takayasu arteritis is a chronic inflammatory vasculitis involving large vessels, predominantly the aorta and its branches.[1] This disorder affects medium- and large-sized arteries and their branches. It is most commonly observed in young women, particularly in Asian populations. The disease primarily targets the aorta and its major branches—such as the renal, carotid, and subclavian arteries—resulting in narrowing (stenosis), blockage (occlusion), or sometimes abnormal dilation (aneurysm) of these vessels.[1] Renal artery involvement is seen in approximately 60% of

pediatric cases in India and the Far East.[2] Hyponatremic hypertensive syndrome (HHS) is a rare clinical entity characterized by severe hypertension, hyponatremia, and polyuria, typically resulting from unilateral renal ischemia. It is mediated by excessive renin secretion and pressure natriuresis from the contralateral kidney.[3] Early recognition is essential to prevent complications.

CASE PRESENTATION

A 4-year-old girl was admitted to the Pediatric Intensive Care Unit (PICU) with multiple episodes of afebrile convulsions. The patient had no significant medical history except for polyuria and polydipsia.

On examination, bilateral brachial and radial pulses were absent in the patient. Blood pressure measurements revealed hypertension, with lower limb blood pressure higher than upper limb blood pressure. Fundoscopic examination revealed grade 1 hypertensive retinopathy. Other systemic examinations were unremarkable.

Investigations

Laboratory findings revealed persistent hyponatremia and polyuria. The serum creatinine level was within the normal range.

Neuroimaging (brain MRI) revealed multiple hypodensities. CT angiography revealed multiple stenotic lesions involving the right and left subclavian arteries and the left vertebral artery, along with significant stenosis of the right renal artery and a small right kidney.

Echocardiography revealed left ventricular hypertrophy (LVH), likely secondary to chronic hypertension.

Diagnosis

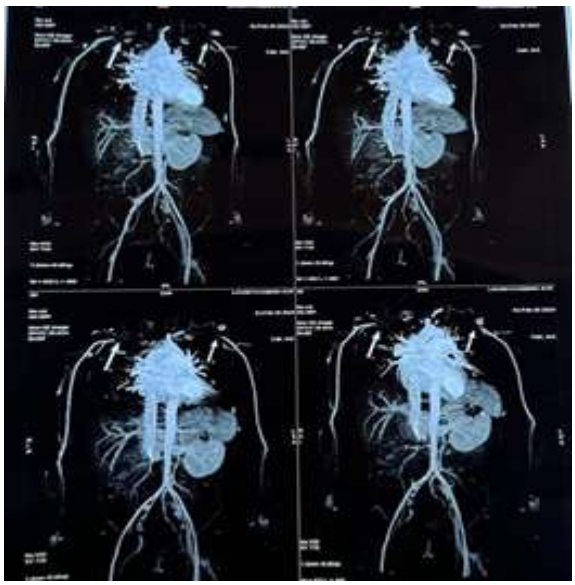
Based on clinical findings and imaging, a diagnosis of Takayasu arteritis with hyponatremic hypertensive syndrome was established according to EULAR criteria.

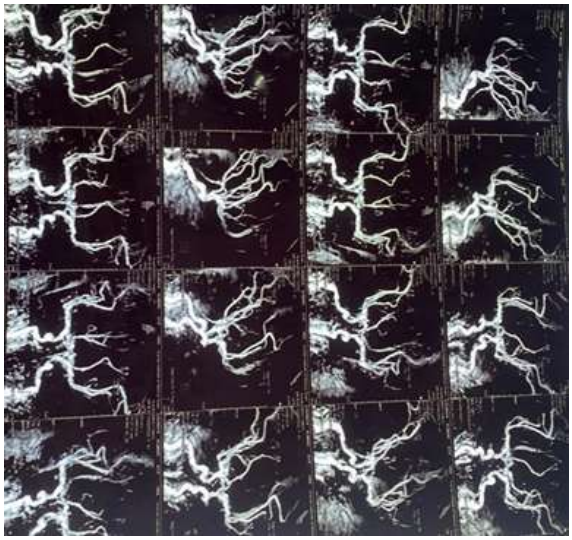
Management and Outcome

Based on clinical findings and imaging, a diagnosis of Takayasu arteritis with hyponatremic

The patient was started on corticosteroids and methotrexate, as per the established treatment guidelines. On follow-up, the patient showed significant improvement, with normalization of blood pressure and serum sodium levels.

However, repeat CT angiography revealed the progression of renal artery stenosis. Therefore, renal angioplasty was planned as part of the further management.





DISCUSSION

Child with renal artery stenosis with persistent stage2 hypertension, vasculitis should be one of the differential diagnosis and work up should be done to rule out causes of vasculitis.[4,5] Takayasu arteritis with refractory hypertension and hyponatremia is a rare complication. The pathophysiology behind it is the involvement of RAAS leading to volume overload, which causes pressure diuresis through the contralateral kidney, leading to persistent hyponatremia. ADH also causes volume overload in this condition, leading to worsening hyponatremia. Takayasu arteritis should be suspected in children presenting with absent pulses, blood pressure discrepancies, and vascular bruits.[6] Imaging modalities such as CT angiography play a crucial role in diagnosis.[7] Early immunosuppressive therapy can control disease progression, while interventional procedures like angioplasty may be required in progressive vascular stenosis.[8]

CONCLUSION

Refractory hypertension with persistent hyponatremia is a very unusual presentation in children, and a thorough evaluation should be performed to rule out renal artery stenosis. General physical examination, particularly pulse examination, should be performed in all suspected cases of Takayasu arteritis. Early diagnosis is crucial to initiate medical and surgical interventions to prevent life-threatening complications and long-term morbidity.

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