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Radiology

Posterior Nutcracker Syndrome Associated with Azygos Continuation of the Inferior Vena Cava in a Child: Case Report

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Abstract | **Case Report**

Posterior nutcracker syndrome (PNCS) is a rare vascular compression disorder caused by compression of the left renal vein between the aorta and vertebral column. It is infrequent in pediatric populations and can present with nonspecific symptoms such as chronic abdominal pain. The association with azygos continuation of the inferior vena cava (IVC) is exceedingly rare. We report a case of a 10-year-old boy presenting with abdominal pain, diagnosed with posterior nutcracker syndrome associated with azygos continuation of the IVC. This diagnosis was established through CT imaging and conservative management led to a favorable clinical outcome.

Keywords: Posterior Nutcracker Syndrome, Azygos Continuation, Inferior Vena Cava Anomaly, Pediatric, Computed Tomography.

INTRODUCTION

Posterior nutcracker syndrome (PNCS) is a rare vascular compression disorder caused by entrapment of the retroaortic left renal vein between the abdominal aorta and the vertebral column. While anterior nutcracker syndrome is more frequently encountered, the posterior variant is far less common and particularly rare in children. Clinical manifestations are variable and nonspecific, making diagnosis challenging in the pediatric population.

Azygos continuation of the inferior vena cava (IVC), a rare congenital vascular anomaly, results from developmental failure of the hepatic segment of the IVC. This anomaly is generally asymptomatic but may alter normal venous return and complicate clinical scenarios.

The coexistence of PNCS with azygos continuation of the IVC is exceptionally rare and has not been widely documented in pediatric patients. Its recognition is important due to potential implications for renal perfusion and venous drainage.

This case report aims to highlight this rare association, underlining the role of imaging—particularly contrast-enhanced CT—in establishing the diagnosis, and to discuss the rationale for conservative management in such cases.

CASE REPORT

A 10-year-old male patient with no significant past medical history presented with chronic, intermittent abdominal pain evolving over several months. The pain was localized to the left flank and epigastric regions, without specific triggering factors. There was no history of hematuria, urinary tract infections, weight loss, or systemic symptoms.

Physical examination was unremarkable. Laboratory investigations, including complete blood count, renal function tests, and urinalysis, were within normal limits.

An initial abdominal ultrasound was inconclusive, leading to a contrast-enhanced abdominal CT scan. Imaging revealed:

- Azygos continuation of the inferior vena cava, with absence of the hepatic segment of the IVC and drainage of the lower body venous return via a dilated azygos vein.
- Compression of the retroaortic left renal vein between the abdominal aorta and the vertebral body, consistent with posterior nutcracker syndrome.
- No evidence of thrombus, renal vein aneurysm, or collateral venous dilatation.

Given the absence of significant hematuria, renal dysfunction, or severe symptoms, a conservative symptomatic management strategy was adopted. The patient was treated with analgesics and advised regular follow-up.

At 6-month follow-up, the patient reported significant clinical improvement, with a decrease in the frequency and intensity of abdominal pain. No new symptoms developed.

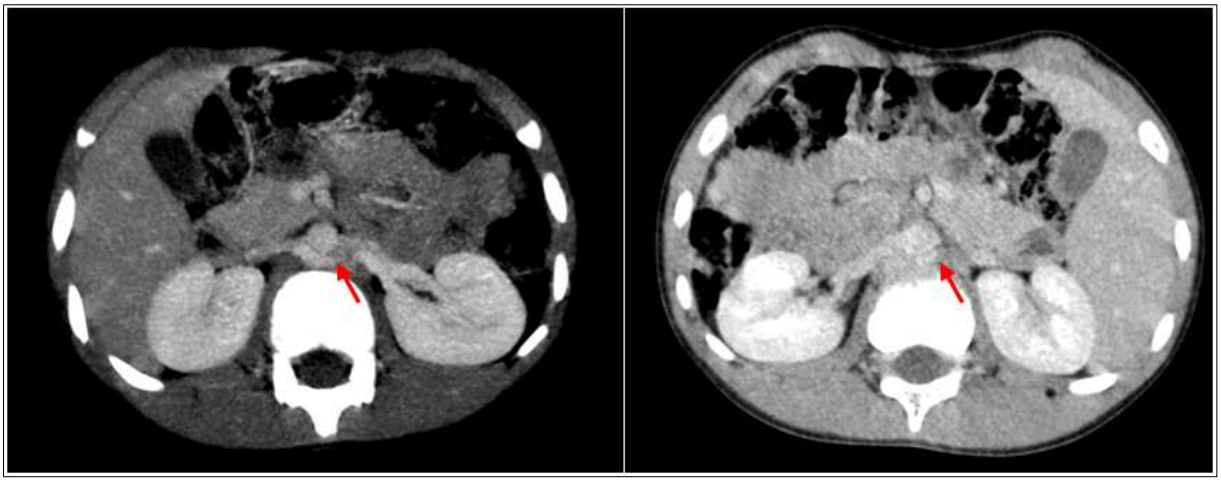

Figure 1: Axial arterial (right) and portal (left)CT images showing a retro aortic left renal vein (red arrow) which is compressed between the abdominal aorta and the third lumbar vertebral body

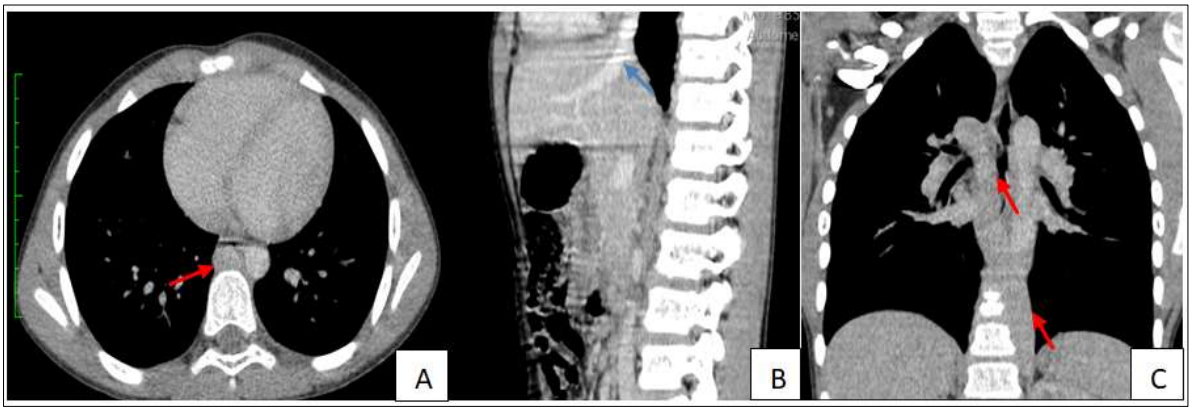

Figure 2: Axial (A), sagittal (B) and coronal (C) with contrast CT images showing an azygos continuation of the IVC with absence of the hepatic segment of the IVC (blue arrow) and drainage of the lower body venous return via a dilated azygos vein (red arrow)

DISCUSSION

Posterior nutcracker syndrome (PNCS) results from compression of the retroaortic LRV between the aorta and the vertebral column [1]. Unlike anterior NCS, which is relatively more common, PNCS is rare and even more so in the pediatric population. Clinical manifestations are variable and can include hematuria, abdominal or flank pain, varicocele, or be entirely asymptomatic [2].

Azygos continuation of the IVC is a rare congenital anomaly with an incidence estimated at 0.6% of the population [3]. It results from embryological failure of the normal development of the IVC, with blood from the lower body rerouted via the azygos vein into the superior vena cava. Although often asymptomatic, it may have implications for venous return dynamics and complicate central venous access or surgical procedures [4].

The association of PNCS with azygos continuation of the IVC is extremely rare and may increase the susceptibility to venous congestion in the renal circulation, potentially exacerbating symptoms [5]. Diagnosis is primarily imaging-based. Doppler ultrasound can suggest the diagnosis by demonstrating altered flow patterns, but CT or MR angiography provides definitive anatomical information [6, 7].

Management strategies depend on symptom severity. Conservative management is favored in cases with mild symptoms, particularly in children, due to the potential for growth-related anatomical changes to alleviate the compression [8]. Surgical or endovascular interventions are reserved for cases with severe

hematuria, significant renal dysfunction, or debilitating symptoms [9, 10].

Our patient benefited from conservative management with symptomatic treatment and showed favorable clinical evolution, supporting the appropriateness of a non-invasive approach in selected pediatric cases.

CONCLUSION

Posterior nutcracker syndrome, particularly when associated with azygos continuation of the IVC, is an exceedingly rare but important consideration in the differential diagnosis of chronic abdominal pain in children. Contrast-enhanced CT imaging is instrumental in establishing the diagnosis. In the absence of severe complications, conservative management with close clinical follow-up is a reasonable and effective therapeutic strategy.

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