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NUTCRACKER SYNDROME: A REPORT OF TWO CASES

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ABSTRACT

Introduction: **Nutcracker syndrome is a pathology that could pose a diagnostic conundrum to the physician as the patient could present with vague symptoms. This report brings to limelight the pathology of Nutcracker syndrome and the need for clinicians to consider this as a differential diagnosis when managing patients with non-specific abdominal pain.**

Case presentation: **This is a case series report of two patients that presented at the emergency unit with acute exacerbation of insidious onset of upper abdominal pain. Radiological evaluation with computerized tomography proved the presence of this pathology in both patients with reduction in aortomesenteric angles and distances. The management of this clinical entity is largely conservative with surgical intervention rarely needed.**

Conclusion: **Radiological evaluation is the bedrock of diagnosis of Nutcracker syndrome. The clinician should endeavor to consider this as a differential diagnosis in patients with recurrent and undefined upper abdominal symptoms.**

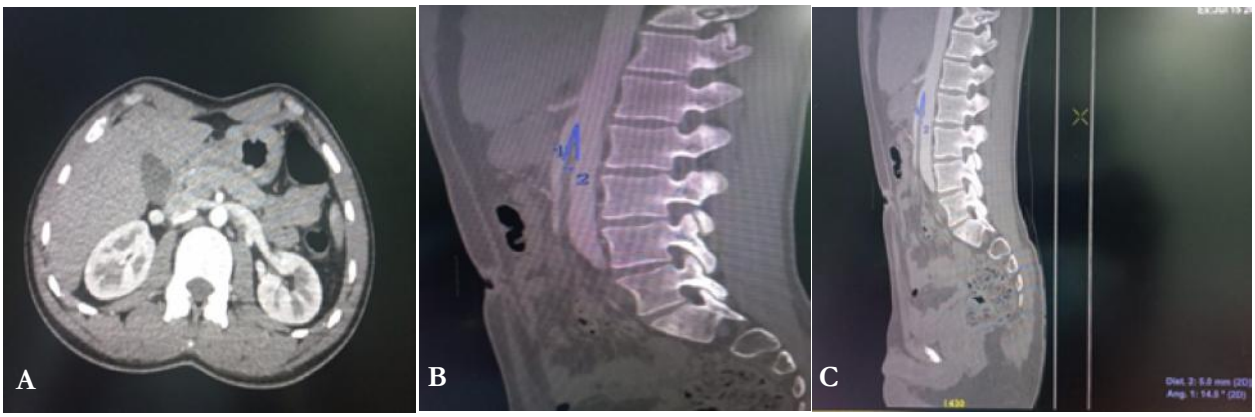
Keywords: Nutcracker syndrome, Abdominal pain, Radiology

INTRODUCTION

Nutcracker syndrome (NS), also called left renal vein (LRV) entrapment syndrome, is a rare vascular compression disorder referring to extrinsic compression of the LRV by the superior mesenteric artery (SMA) anteriorly and the aorta posteriorly.[1] It can lead to lead to renal vascular congestion manifesting as hematuria, proteinuria, orthostatic hypotension, pain or renal dysfunction.[2] The severity of the NS is variable and affected individuals may be completely asymptomatic, in which case it is referred to as nutcracker phenomenon, or in the most severe cases, experience severe pelvic congestion.[3] NS may be associated with a SMA compression disorder where the SMA compresses the third part of the duodenum.[4] Diagnosis can be challenging and variable, frequently involving a combination of Ultrasound Doppler, cross-sectional, and invasive imaging.[5] Management depends on severity of clinical features. Treatment ranges from expectant management to surgical intervention.

Case Presentation 1

HSH, a 20-year-old male adult, presented to the emergency room with acute exacerbation of chronic



A - Transverse contrast-enhanced Abdominal CT image showing the let renal vein compressed between the aorta and the superior mesenteric artery
B - Sagittal section showing the SMA overlying the bowel, with aorta behind
Inset 1- Superior mesenteric artery, Inset 2- Abdominal aorta
C - Sagittal section showing the anomalies with angle and distance measurements (inset at right lower corner)

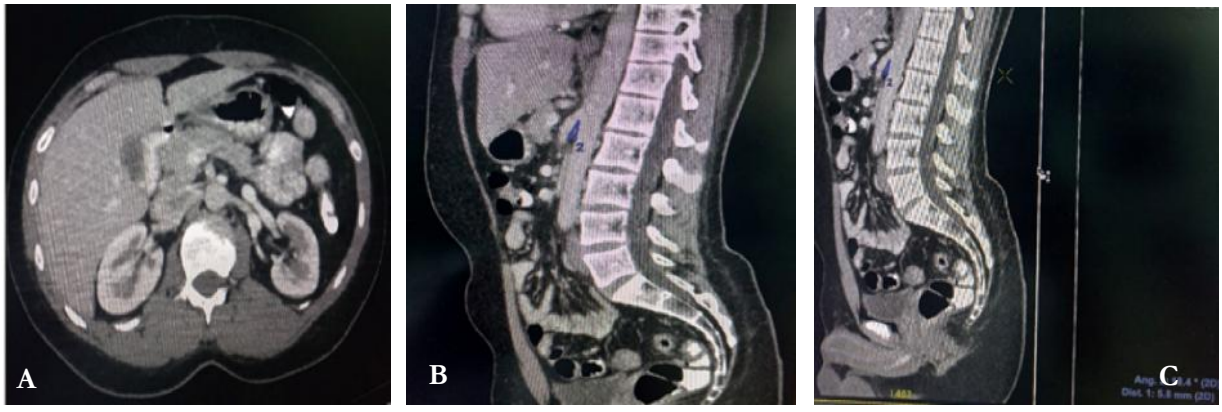
upper abdominal pain. The pain was described as dull and dragging, with associated nausea and occasional vomiting. He has been to other hospitals in the years past for similar presentation. There was no history of blunt trauma to the abdomen and no history of abdominal surgery. Clinical examination revealed a young man, well preserved, not pale, hydrated with stable vital signs. Abdomen is flat, with diffuse and vague tenderness in the epigastric and peri-umbilical regions, no positive, expansile cough impulse. Abdominal sonography done was essentially normal. He later had an abdominal computerized tomography (CT) scan done that revealed an aortomesenteric distance of 5-6mm, while aortomesenteric angle was 14^0, with left renal vein compressed between the SMA and the aorta. All laboratory investigations were essentially normal.

Case Presentation 2

AMA, 29 year old, male patient who presented to the surgical out-patient clinic with recurrent upper abdominal pain, nausea with occasional vomiting. Patient also has a history of Crohn's disease and he has been receiving treatment for this. Clinical examination shows a young man, well preserved with stable vital signs. Abdominal examination showed a flat abdomen, with mild tenderness on deep palpation at the epigastrium. All laboratory parameters were within normal limits. Abdominal sonography was equivocal while he has abdominal CT scan done which revealed the presence of Nutcracker syndrome (Illustration 2). He was counselled and referred to the gastrointestinal surgery unit for further follow-up.

phenomenon.[1] However, the earliest pathological illustration of this condition was provided by the anatomist Grant who described the position of the LRV between the aorta and the SMA as similar to a nut lying between the jaws of a nutcracker.[6] This condition is also called "anterior nutcracker syndrome". Less commonly, a circumaortic (up to 17%) or completely retroaortic (3%) left renal vein may be compressed between the aorta and vertebral body, which is termed "posterior nutcracker syndrome".[7] Epidemiologically, the prevalence of the disease is not known however, it is estimated to be relatively more common in females and usually presents in the 3rd or 4th decade of life.[8] Clinical presentations actually depends on the extent of vascular compression hence acute (if there is significant vascular compression) and acute-on-chronic (with insidious history of vague abdominal pain and other constitutional symptoms).[3] Although female preponderance has been earlier reported, our index patient was a male in his second decade of life with a clinical presentation of insidious onset of abdominal pain.

The underlying pathology of this entity has been linked to the loss of the protective, supporting fat cushion around the SMA thereby reducing the gap between the aorta and the vessel, making this vessel prone to external compressive forces from the surrounding structures.[9] One explanation has suspected a low body mass index (BMI), with some authors noting that symptoms resolve with increasing BMI particularly in children.[10] Other authors suggest renal ptosis as a s



A - Transverse contrast-enhanced Abdominal CT image showing the left renal vein compressed between the aorta and the superior mesenteric artery
B - Sagittal section showing the SMA overlying the bowel, with aorta behind
C - Sagittal section showing the anomalies with angle and distance measurements (inset at right lower corner)

DISCUSSION

The term Nutcracker syndrome, was first described by De Schepper in 1972 as an incidental finding of vascular compression with clinical symptoms, differentiating it from the asymptomatic Nutcracker cause, a condition in which the kidney descends within the retroperitoneum upon position change from supine to upright, and has been implicated in LRV stretching over the aorta and subsequent venous congestion.[11]

The diagnosis of NS is hinged on radiological assessment. Sonographic findings in support of NS include reduction in both aorto-mesenteric angle (<25^0) and distance (<8-10mm).[6] However, this modality is operator dependent and such can be a major limitation. The CT scan has a better resolution and tissue plane delineation showing a reduction in aorto-mesenteric angle (6^0-22^0) and distance (2-8mm).[8] Our index patients had an aortomesenteric distance of 5-6mm each, and aortomesenteric angle of 14^0 and 18.4^o respectively, which are all in conformity with what is expected for NS.

The treatment of NS depends largely on the mode of presentation. In patients with incidental nutcracker phenomenon where there are no clinical presentations, expectant management is favored. In cases of mild NS with non-specific constitutional symptoms and no back-flow changes on radiological imaging, conservative treatment is commonly explored first. Cases of persistent severe hematuria may resolve spontaneously in adolescents as a consequence of increased height.[12] For patients with associated SMA syndrome who present with features of bowel obstruction, they are managed according to established protocol for managing acute bowel obstruction which entails, placing such patients on NPO, intravenous fluids, nasogastric decompression and instituting nutritional rehabilitation either nasoenteral or total parenteral nutrition.[13] Generally, conservative therapy is often appropriate in management of NS, especially in children and adolescents. Endovascular, laparoscopic, and open surgical treatments have been utilized successfully in severe and persistent cases.[5]

Our index patients were admitted and managed conservatively with generous opioids analgesia, NPO and intravenous fluids. Both patients eventually recovered from their symptoms and were commenced on oral feeding as advised by the nutritionist. Both patients were eventually discharged home from the hospital after 5 days on admission to be followed up at the out-patient clinic.

CONCLUSION

Rare pathologies like NS could pose a diagnostic dilemma to the attending physician and radiologist. The radiologist therefore should arm him/herself with the requisite knowledge of this rare entity in order to make a diagnosis. The clinician as well should have a high index of suspicion when patients present with vague abdominal pain. The benefit of multidisciplinary team working together to establishing this diagnosis and providing prompt treatment remains invaluable.

Authors' contribution: Dr. Osobu was involved in the evaluation of this patient. Both authors conceived the idea of writing this article and both were also involved in literature search, typesetting and final review of this manuscript.

Conflict of Interest Statement

The authors affirm that they have no conflict of interests to declare.

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