Vascular Compression Syndromes In Pediatric Population

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Abstract

Vascular Compression Syndromes, namely Median Arcuate Ligament Syndrome, Nutcracker Syndrome, May-Thurner Syndrome, and Superior Mesenteric Artery Syndrome is rare and patients go undiagnosed with various mutisystemic symptoms for long time with often debilitating consequences. Here we present several cases of pediatric patients each having multiple compression syndromes with their varied presenting symptoms, diagnostic work up, and treatments modalities. Certain abdominopelvic vascular structures may be compressed by adjacent anatomic structures or may cause compression of adjacent hollow viscera.

Such compressions may be asymptomatic; when symptomatic, however, they can lead to a variety of uncommon syndromes in the abdomen and pelvis, including median arcuate ligament syndrome, May-Thurner syndrome, nutcracker syndrome, superior mesenteric artery syndrome, ureteropelvic junction obstruction, ovarian vein syndrome, and other forms of ureteral compression. These syndromes, the pathogenesis of some of which remains controversial, can result in nonspecific symptoms of epigastric or flank pain, weight loss, nausea and vomiting, hematuria, or urinary tract infection. Direct venography or duplex ultrasonography can provide hemodynamic information in cases of vascular compression.

However, multidetector computed tomography is particularly useful in that it allows a comprehensive single-study evaluation of the anatomy and resultant morphologic changes. Anatomic findings that can predispose to these syndromes may be encountered in patients who are undergoing imaging for unrelated reasons. However, the diagnosis of these syndromes should not be made on the basis of imaging findings alone. Severely symptomatic patients require treatment, which is generally surgical, although endovascular techniques are increasingly being used to treat venous compressions.

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When symptomatic, such compressions can result in a variety of uncommon syndromes in the abdomen and pelvis, including median arcuate ligament syndrome (MALS), MayThurner syndrome, nutcracker syndrome, superior mesenteric artery (SMA) syndrome, UPJ obstruction, ovarian vein syndrome (OVS), and other forms of ureteral compression (Table). In this article, we refer to this heterogeneous group of disorders as “vascular compression syndromes,” since they all involve either the compression of vascular structures or the compression of hollow viscera by vascular structures.

Controversy surrounds the pathogenesis of some of these syndromes. Anatomic or morphologic findings that predispose to such compression may occasionally be encountered in asymptomatic patients who undergo imaging for unrelated causes. Thus, caution should be exercised to avoid overdiagnosis of these syndromes.

Symptoms resulting from such compressions can be vague, nonspecific, and obscure, resulting in delayed, incorrect, or missed diagnoses. Although many of these syndromes were described decades ago, they remain poorly understood. If unrecognized and untreated, they can be associated with significant morbidity. These syndromes may be encountered by physicians in a variety of disciplines and can present a diagnostic dilemma. In this article, we aim to familiarize radiologists with the multidetector CT appearance of these syndromes and the added benefit of MPR in diagnosis. When conservative management is not indicated or fails, surgery is the mainstay for treatment. Open surgical techniques are now being replaced by less invasive laparoscopic techniques. Outcomes following surgery may vary, and the decision to treat should be made only in those patients who are experiencing disabling or severe symptoms. In this article, we review vascular compression syndromes in the abdomen and pelvis in terms of relevant anatomy, pathogenesis, clinical presentations, imaging findings (with emphasis on findings at multidetector CT), and treatment options.

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