Median Arcuate Ligament Syndrome

A 32-year-old woman presented with upper abdominal pain and recent weight loss. She underwent CT angiography and conventional angiography demonstrating focal extrinsic compression and associated downward deflection of the celiac trunk (A, arrow).

Median arcuate ligament syndrome (also known as celiac artery compression syndrome, celiac axis syndrome, celiac trunk compression syndrome, or Dunbar syndrome) is a rare disorder characterized by chronic, recurrent abdominal pain related to compression of the celiac artery by the median arcuate ligament (a muscular fibrous band of the diaphragm).\(^1\) It usually presents in patients 20 to 40 years of age with symptoms of abdominal pain, weight loss, and abdominal bruit, and affects women far more frequently than men (35:1). It is unclear whether symptoms occur from vascular compromise or from compression of the celiac plexus.\(^1,2\) CT angiography and conventional angiography are considered the gold standard imaging modalities.

Imaging findings are characterized by a hooked appearance of the celiac axis due to superior extrinsic compression, frequently with associated poststenotic dilation (B, arrow). Functional stenosis may lead to collateral formation between the superior mesenteric and celiac arteries, most notably though hypertrophy of the pancreaticoduodenal arcades (C, arrow). When indicated by severity of symptoms, therapy is surgical decompression or bypass.\(^2\)

The characteristic appearance of the stenosis, absence of associated disease of the superior mesenteric artery, as well as the younger age of the patient, distinguishes this syndrome from the primary differential of atherosclerotic disease. It is noted that extrinsic compression of the celiac artery may also be seen in up to 50% of asymptomatic patients if imaging is acquired during the expiratory phase.\(^1\) Therefore, imaging for accurate diagnosis should ideally be performed in the end-inspiratory phase.

References