A 41-year-old woman was referred to our institution for evaluation of recurrent cramping pain in the epigastrium since a couple of months. Contrast-enhanced computed tomography revealed a high grade stenosis with a hook like configuration of the celiac trunk with poststenotic dilatation (Fig. A-B, arrowheads at the celiac trunk, single arrowhead: origin, double arrowhead: stenosis and triple arrowhead: poststenotic dilatation; asterisk in aorta). Ultrasound of the abdomen confirmed the stenosis which was more pronounced during expiration. Doppler ultrasound showed peak velocities up to 278 cm/s during inspiration and 334 cm/s during expiration (Fig. C) at the level of the stenosis and poststenotic turbulent flow was clearly visible. The peak velocities in the aorta and superior mesenteric artery were all within normal range. The diagnosis of celiac artery compression syndrome was made. After multidisciplinary consultation, a conservative approach was chosen.

Comment

The celiac artery compression syndrome (CACS), also known as the median arcuate ligament syndrome (MALS) or Dunbar’s syndrome, consists of a focal stenosis of the celiac artery arising as a consequence of compression by the median arcuate ligament. This fibrous structure is bow shaped and connects the left and right diaphragmatic crura on both sides of the aortic hiatus. Occasionally, this ligament may have an intimate relationship with the celiac trunk and may cause focal indentation. Some patients show a higher than normal origin of the celiac trunk, which may also predispose to CACS. This syndrome occurs typically in young slim woman (20 to 40 years old). The celiac stenosis may present as an incidental finding on imaging or may be symptomatic. Symptoms include nausea, vomiting, weight loss, pain in the epigastrium occurring usually postprandial, or after exercise. The clinical examination sometimes reveals an epigastric bruit that varies during respiration. The gold standard for diagnosis is catheter angiography in inspiration and expiration, although this examination is currently replaced by multi-detector computed tomography (MDCT) and CT-angiography. Typically, a ‘hook like’ appearance of the celiac artery is seen. In addition, color Doppler sonography is the imaging modality of choice for functional evaluation of the severity of the stenosis which is more pronounced during expiration. This is explained by a cranial movement of the celiac trunk during expiration rendering the stenosis more significant. A peak systolic velocity greater than 200 cm/s is believed to be diagnostic. Differential diagnosis includes stenosis of the celiac trunk due to atheromatosis which is more frequently seen in the elderly patient. Furthermore, calcifications are absent in CACS. Treatment is still controversial. Decompression of the celiac trunk can be achieved by cleavage of the median arcuate ligament. Alternative methods such as percutaneous transluminal angioplasty and stenting of the celiac trunk can also be performed but with variable results.