Dunbar syndrome as an unusual cause of exercise-induced retrosternal pain

Egzersiz ile şiddetlenen göğüs ağrısının nadir bir nedeni olarak Dunbar sendromu

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Summary– The median arcuate ligament is a fibrous band connecting the left and right diaphragmatic crura across the aortic hiatus at the level of the T12/L1 vertebral bodies. The low insertion point of this ligament causes significant stenosis of the proximal portion of the coeliac artery in a small group of patients, and contributes to ischemic symptoms known as coeliac artery compression syndrome (CACS). It is also referred to as median arcuate ligament syndrome or Dunbar syndrome. Symptoms include especially postprandial epigastric or retrosternal pain, weight loss, nausea, vomiting, diarrhea and reduced appetite. In severe cases, exercise related abdominal pain may be caused by steal phenomenon, whereby blood is shunted to the skin and relevant muscles during exercise. Computed tomographic angiography and mesenteric angiography are the gold standard diagnostic modalities to confirm diagnosis of CACS. Surgical therapy with release of the median arcuate ligament usually is the primary treatment of choice. Here, we present a 46-year-old male CACS patient with postprandial and especially exercise-induced retrosternal pain radiating to the epigastric region, which may be misperceived as a coronary symptom.

Özet– Medyan arkuat ligaman T12/L1 vertebra seviyesinde aortik hiatusu çaprazlayan sağ ve sol diyafram kruslarını birbirine bağlayan fibröz bir bağdır. Az sayıda hastada bu ligamanın düşük seviyeli yerleşimi çölyak arterin proksimalinde ciddi darlığa neden olarak çölyak arter bası sendromu olarak bilinen iskemik semptomlara yol açar. Bu durum ayrıca medyan arkuat ligaman sendromu ya da Dunbar sendromu olarak da bilinir. Semptomlar arasında özellikle yemek sonrasında gelişen epigastrik ya da retrosternal ağrı, kilo kaybı, bulantı, kusma, ishal ve iştahsızlık sayılabilir. Ciddi olgularda egzersiz sırasında kan akımının cilde ve kaslara yönlenmesine bağlı olarak gelişen çalma fenomeni sonucunda egzersiz ile ilişkili karın ağrısı gözlenebilir. Bilgisayarlı tomografik anjiyografi mezenter anjiyografi ile birlikte çölyak arter bası sendromu tanısında altın standart tanı yöntemidir. Medyan arkuat ligamanın cerrahi tedavi ile gevşetilmesi genellikle ilk tercih edilen tedavi seçeneğidir. Burada yemeklerden sonra olan ve özellikle egzersiz ile şiddetlenen epigastrik bölgeye de yayılan retrosternal ağrı şikayeti ile başvuran 46 yaşında erkek hastada koroner semptomları ile de karışabilecek çölyak arter bası sendromu sunuldu.

Mesenteric ischemia is most commonly caused by atherosclerotic disease, but extrinsic compression of the coeliac artery may also lead to similar symptoms. Coeliac artery compression syndrome (CACS), also known as Dunbar syndrome, is a rare disorder characterized by postprandial intestinal angina caused by insufficient coeliac blood supply to the gastrointestinal organs. CACS is thought to arise from compression by the median arcuate ligament that traverses the aortic hiatus of the diaphragm.[1] The characteristic clinical features of the syndrome are postprandial abdominal pain associated with nausea, weight loss and an abdominal bruit. Exercise-induced abdominal pain has been infrequently reported in association with the syndrome.[2] Diagnostic methods may include Doppler ultrasound, spiral computed tomographic angiography (CTA), selective catheter angiography or magnetic resonance angiography.[3]

Abbreviations:

CACS *Coeliac artery compression syndrome*
CTA *Computed tomographic angiography*

Here, we present a patient with CACS who had postprandial and exercise-induced retrosternal pain radiating to the epigastric region.

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CASE REPORT

A 46-year-old man was admitted to our hospital with complaints of exercise-induced lower retrosternal pain radiating to the epigastric region over a 2-year period. His symptoms were aggravated especially after meals. Physical examination showed a flat and non-tender abdomen with normal bowel sounds. He had a history of hypertension, but no known history of coronary artery disease or diabetes mellitus. His blood pressure was 145/90 mm Hg and pulse rate was 82 beats/min. Electrocardiogram revealed sinusal rhythm and laboratory findings were unremarkable. Chest X-ray was normal and transthoracic echocardiography revealed normal left ventricular systolic functions with moderate concentric hypertrophy. An exercise stress test was performed and the patient had abdominal pain after 12 METs exercise without any electrocardiographic changes. He described rapid weight loss, especially in the previous 6 months. Subsequently, pelvic and abdominal ultrasonography was performed which was also unremarkable. Esophagogastroduodenoscopy revealed normal upper gastrointestinal tract mucosa without any suspicion of malignancy. Afterwards enhanced thoracoabdominopelvic CTA was performed which revealed a focal narrowing in the proximal coeliac artery in the horizontal (Figure 1a) and reconstructed sagittal (Figure 1b) views with excessive fibers arising from the diaphragm. Three-dimensional reconstructed CTA (Figure 1c) confirmed the diagnosis of CACS and minimally invasive laparoscopic surgery was offered to the patient by gastrointestinal surgeons. Laparoscopic release of the median arcuate ligament was performed and the patient became absolutely asymptomatic one month after surgery.

DISCUSSION

CACS is defined as abdominal pain related to compression of the coeliac artery by fibers of the median arcuate ligament.[1] CACS was first described in 1963 in a case report by Harjola et al.[4] It is also referred to as median arcuate ligament syndrome or Dunbar syndrome.[5] Since then, the topic has been the focus of numerous controversies regarding its pathophysiology, definitive diagnosis and optimal treatment.

In 10–24% of patients with CACS, compression leads to a significant reduction in coeliac blood flow and causes clinical symptoms that include postprandial and vague epigastric or low retrosternal pain, weight loss, nausea, vomiting, bloating, diarrhea and reduced appetite.[1] In severe cases, exercise related abdominal or low retrosternal pain may be caused by steal phenomenon, when blood is shunted from the gastrointestinal tract to the skin and relevant muscles during exercise, as described in the present case.[2]

There are two major theories that may explain the causes of symptoms in CACS. In the first and more widely-accepted theory, it is thought to be due to a low insertion of the diaphragmatic crus or by malposition of the median arcuate ligament, sometimes in association with hypertrophy of the diaphragmatic muscle fibers.[1] On the other hand, the second theory is related

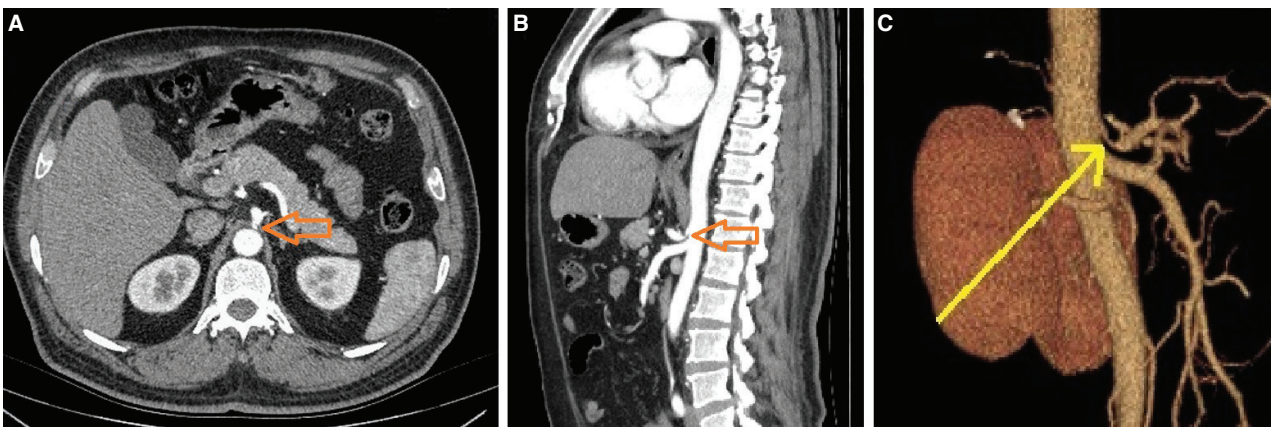

Figure 1. Enhanced thoracoabdominopelvic computed tomographic angiography (CTA) which revealed a focal narrowing in the proximal coeliac artery with excessive fibers arising from the diaphragm in horizontal **(A)** and reconstructed sagittal **(B)** views. **(C)** Three-dimensional reconstructed CTA confirmed the diagnosis of coeliac artery compression syndrome.

to neurogenic stimulation caused by compression of the coeliac ganglion and plexus. The neurogenic stimulation theory proposes that pain results either from coeliac plexus stimulation leading to splanchnic vasoconstriction and ischemia, or via direct sympathetic pain fiber irritation.[6]

Diagnosis of CACS may be made by Doppler ultrasound, spiral CTA, selective catheter angiography, and magnetic resonance angiography.[3] Doppler US has been reported as having a high sensitivity for the diagnosis of CACS and is proposed as the initial modality of choice.[7] However, the gold standard diagnostic methods are still selective angiography and CTA, which also provide a radiological approach to obtain a sagittal view of the coeliac artery by using three-dimensional reconstruction.[8]

A variety of surgical techniques are employed to manage this syndrome, including median arcuate ligament transection, coeliac ganglion destruction and revascularization of the decompressed coeliac artery. However, the limited number of cases, and short-term follow-up periods have made it difficult to evaluate the best treatment strategy. The traditional operative approach involves a midline laparotomy with division of the anomalous fibrous diaphragmatic bands overlying the coeliac artery, along with the coeliac plexus and lymphatic tissues. Laparoscopic management of CACS in carefully selected patients results not only in long-term outcomes similar to those in which the traditional approach is used, but also involves a shorter inpatient hospital stay, earlier return to normal activities and better cosmetic results.[9]

Angioplasty and stenting of visceral vessels have been described as effective methods in the treatment of atherosclerotic disease, but their use in the treatment of CACS appears questionable, particularly because extrinsic compression may prevent adequate dilatation of the vessel during stent implantation. Indeed, if the symptoms are due to involvement of the splanchnic nerve plexus, the role of endovascular treatment is out of question.[10]

CACS is related to compression of the coeliac artery by fibers of the median arcuate ligament and should be considered in the differential diagnosis of exercise-induced low retrosternal or epigastric pain. Surgical therapy with minimally invasive release of the median arcuate ligament should be the primary treatment of choice, since several investigators have failed to demonstrate success with endovascular treatment.

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Key words: Coeliac artery/abnormalities; median arcuate ligament; computed tomography.

Anahtar sözcükler: Çölyak arter/anormallik; medyan arkuat ligaman; bilgisayarlı tomografi.