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RESEARCH ARTICLE

MISLEADING POST PRANDIAL PAIN: MEDIAN ARCUATE LIGAMENT SYNDROME REVIEW OF LITERATURE AND CASE REPORT

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Abstract

Introduction: Median arcuate ligament syndrome (MALS) is a rare disease with different names. Patients are usually young females, presenting with abdominal pain and weight loss. The symptoms are caused by compression of the celiac artery by the median arcuate ligament.

Case report: A 35-year-old male patientwas admitted as a case of COVID, with a chronic history of postprandial abdominal pain. The patient was treated before for a helicobacterpylori infection after presentation with the same complaint. When he resolved his COVID condition, an abdominal computed tomography incidentally showed celiac artery stenosis with a hooking appearance. He underwent a laparoscopic release. Interestingly, he was readmitted after 25 days and diagnosed to have Addison's disease.

Discussion: Due to the syndrome's rarity, it is usually diagnosed after the exclusion of more common differentials. Although that the European Society for Vascular Surgery (ESVS) is considering MALS as the most common cause of single-vessel arterial stenosis. The diagnosis is made through the presentation of radiological evidence of celiac trunk compression. The release is the mainstay of treatment for the best outcome and is trending nowadays to be by the laparoscopic procedure.

Conclusion: There is not enough MALS awareness, and there are needs to be established. This can happenthrough more proper case reviews, trials of new approaches, and management.

Introduction:-

Median arcuate ligament syndrome is a rare condition. Usually, the patients are young females. The clinical presentation mostly will show abdominal pain, which can be unprovoked, post-exertion, or postprandial epigastricpain. There are associated signs and symptomssuch as weight loss, abdominal bruit, or nausea. And at the end, the reason would finally be due to a compression of the median arcuate ligament to the celiac artery. [1]

To date, there is no agreement about specific criteria to suspect MALS from the initial presentation, which makes a difficulty to address a specific modality for establishing the diagnosis. [2]

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Using computed tomography angiography can show the hooking manifestation, which is characteristic of MALS. [3]

In 1917, Lipshutz et al, described a compression of the celiac trunk by the median arcuate ligament in autopsies which was the 1st time to be mentioned in the literature review. Publishing about a release of compression of the celiac artery was described for the first time in the 1960s by Harjola, J P Carey then Dunbar et al. [4,5,6,7]

Release of the median arcuate ligament, whatever the way the procedure opens, laparoscopic, robotic, or even endoscopic can provide symptomatic relief, but there are insufficient data on the long-term outcome. [2]

Case Report:

We are presenting a case of a 35-year-old Saudi male patient with, a known case of asthma, who was admitted to East Jeddah Hospital as a case of COVID pneumonia with acute kidney injury due to dehydration.

The patient had a history of chronic epigastric pain, for almost 18 months. The pain is dull, with no history of shifting or radiation. The pain was usually post-prandial and aggravated by walking, with no specific relieving factors, and he had multiple visits seeking medical help for pain control. There is a history of weight loss of almost 12 kilograms only during the last 5 months. Sometimes the condition is associated with nausea and vomiting with food content.

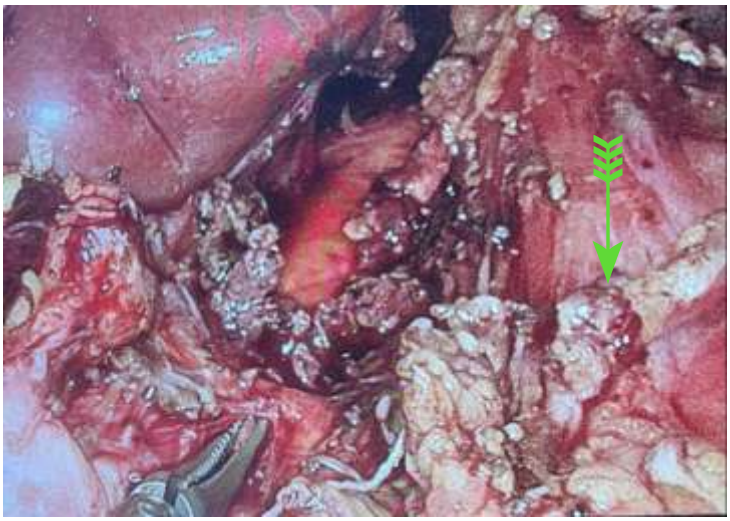
The patient was following with gastroenterology, and he had a previous Esophagogastroduodenoscopy, and the findings were mild chronic active gastritis with histopathology results proven to be positive for helicobacter pylori infection, he toke the full course of treatment and proven later to be cured and that the infection was eradicated, but unfortunately, the symptoms did not improve.

After resolving the patient from the COVID condition, the patient was still complaining about the same abdominal pain, although the abdomen was soft and lax since admission. Abdominal computed tomography was done and incidentally was found that he is having median accurate ligament syndrome, which was proven by abdominal computed tomography angiography.

The image showed that there is a focal narrowing of the proximal celiac axis with the characteristic hooked appearance. There was post-stenotic prominence but there was no dilation or collaterals formation.

Abdominal computed tomography angiography showing a deflection of the celiac trunk by

A laparoscopic release was done, The dissection was started from the lesser curvature area exposing, isolating, and following the left gastric artery until reaching the celiac trunk and identifying all of its branches. Careful Dissection of the right crus exposing the aorta, the median arcuate ligament was identified at this point and was divided very carefully using a combination of hook cautery and harmonic shears. The anterior surface of the aorta was exposed for about 3 cm. The celiac trunk was completely skeletonized down to the trifurcation area. All fibrotic tissue and nerve plexus overlying the celiac axis were resected.



Green arrow: The median arcuate ligament anterior to the celiac artery.

The patient was shifted to a regular ward, tolerated the surgery, and was discharged on the 2nd-day post-operative. The patient was seen later in the clinic doing well with much improvement, he was eating well with no complaints.

Interestingly, on the 25th day after the operation, the patient revisit the emergency department complaining of postprandial abdominal pain, associated with vomiting of food content and constipation for 2 days.

On examination, he was alert, conscious, and oriented. His abdomen was soft and lax but he was having generalized tenderness. He was hypotensive (Mean arterial pressure: 63), tachycardic (Pulse: 98), hypoglycemic (Random blood sugar: 41), and afebrile.

Dextrose was given to the patient, and his blood sugar was improved. But regardless of resuscitation, his blood pressure was becoming worse, which led the end to start him into traps and he was admitted to the intensive care unit.

He did not have leukocytosis, but his hemoglobin was mildly low (11.7 g/dL). And his potassium was mildly high (5.32 mmol/L) and sodium was low (122 mmol/L).

Abdominal computed tomography was done And showed nothing except for improvement of the previously mentioned stenosis and there were no signs of bowel ischemia.

Dehydroepiandrosterone sulfate was ordered and Intravenous hydrocortisone was given to the patient and he was markedly improved. Dehydroepiandrosterone sulfate results were revealed laterally and it was low (27.4 ug/dL) to confirm that the patient is having Addison disease.

The patient has weaned off inotropes, shifted later on after a few days to the regular ward, continued to follow up by the medical endocrinology team, and was discharged later on after optimization on oral hydrocortisone.

Discussion:-

Median arcuate ligament syndrome, Celiac artery compression syndrome (CACS), or Dunbar syndromeis a group of signs and symptoms, that may present all or some of them, usually in young females with median age is 40 years. It

is a rare condition with different names. It is caused by compression of the median arcuate ligament to the celiac artery. Leading to abdominal pain mostly, which can be unprovoked, post-exertion, or postprandial epigastricpain. There are associated signs and symptoms such as weight loss, abdominal bruit, or nausea, ordered according to their percentages respectively. [1]

This syndrome had been reported may lead to more complications such as celiac artery aneurysms or different visceral arterial aneurysms distal to the stenosis. [8]

Curiously, studies are showing that presence of unprovoked abdominal pain before the surgery is predicting of poor outcomes from the release of the compression. [9]

There was also an observation of an association between psychiatric illnesses including anxiety and mood disorders. [10]

Usually, at the level of the aortic hiatus and cranial to the celiac artery afibrous tissue, the median arcuate ligament, is connecting the diaphragmatic crura. In MALS the median arcuate ligament was found to be caudal to the celiac artery. Leading to the compression of the artery. [11]

In the literature review, it was found that the compression may lead to histological changes in the arterial wall with the chronicity of the condition. It was recognized that the clinical presentation may not be only due to the compression of the artery that causes abdominal angina but maybe also due to the compression of the celiac ganglion. The severity of the presentation may depend also on the respiration variation. [12,13]

To date, there is no agreement about specific criteria to suspect MALS from the initial presentation, which makes a difficulty to address a specific modality for establishing the diagnosis. As a result, usually, it will be detected later after the exclusion of the other more common differential diagnosis, although the European Society for Vascular Surgery (ESVS) is considering MALS as the most common cause of single-vessel arterial stenosis. [2,14]

However, there is still insufficient data to correlate the percentage of stenosis, not the angulation, and the development of symptoms. [15]

By Imaging, especially duplex ultrasound because it is less invasive, cost-effective, and can easily measure the blood flow velocities, the celiac trunk of the symptomatic patient with MALS will be deflected > 50 degrees and the peak flow velocities should be measured during respiration which makes this modality appropriate initial imaging. [16]

Besides computed tomography angiography, Magnetic resonance angiography, and digital subtraction angiography, all of them can detect the hooking appearance, the aneurysms that developed due to Celiac artery stenosis, and they can detect the collaterals that may predict better prognosis after the compression release. [3,17]

Regarding the collaterals, there is a correlation between the stenosis percent of the development of the collateral. [18]

Another pre-operative predictor is performing celiac plexus block which will result in temporary symptomatic ease in some patients. Those who will experience this improvement, most of them will benefit from the release of the compression. [19]

The Laparoscopic release had higher post-operative symptom relief, lower recurrence rates, faster time to feeding, and shorter duration of hospitalization than open release. Besides that, it had also a shorter operative time than robotic release. Which made this trend towards laparoscopic release in recent years. [20,21,22]

Generally, the patients who managed conservatively had worsening outcomesthan those who managed by actual releasing of the compression. [23]

Endovascular intervention solely has shown in the litterer review that its inadequate treatment, but it may be needed for persisting stenosis after releasing the compression. [24]

On the other hand, the endovascular intervention was found to be promising, if it was combined with the endoscopicretroperitoneal release. This makes a question about the results if the endovascular intervention complained with the cases that were managed with the surgical release and complicated with recurrence. [17,25]

Conclusion:-

There is not enough MALS awareness, and there are needs to be established. This can happenthrough more proper case reviews with long-term follow-up. Meanwhile, we can consider using new approaches, including activating multidisciplinary meetings and management, including internists, radiologists, psychiatrists, and vascular and laparoscopic surgeons. There is also a need to try new combined treatmentssuch as using endovascular intervention before the surgical release.

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