Elephant Trunk Sign of Rajul: A Rare Radiological Finding in Agenesis of Celiac Axis

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Abstract
Agenesis of celiac axis is a rare congenital anomaly associated with variable patterns of origin of its various branches. Among this, separate origin of common hepatic and splenic arteries arising nearly at the same level directly from aorta is rarer. This article describes the congenital narrowing of common hepatic & splenic arteries near their origins origin, which has not been described in medical literature.

Keywords: Trunk; Agenesis; Celiac Axis

Introduction
Agenesis of celiac axis is a rare anomaly of abdominal aorta with an estimated incidence of 0.1–2.5% in cadaveric studies [1,2]. Less than forty cases have been reported in the medical literature with majority reported on either angiographic or anatomic studies [2,3]. The frequent Multi-detector Computed Tomography (MDCT) evaluation of abdomen has made incidental detection of these anomalies commoner. Information about these congenital and physiological variations may be useful during laparoscopic and robotic procedures and in predicting the course of diseases related to the affected vessels [4]. We present a rare appearance of common hepatic & splenic arteries in a rare case of agenesis of celiac axis.

Case Report
A 30-years old male consulted our department for Contrast-Enhanced Computed Tomography (CECT) of abdomen for detection of cause of low-grade pyrexia of unknown origin. CECT was performed on 128-slice scanner with arterial and venous phases following optimal distention of stomach and small bowel by a neutral contrast agent i.e. water. All laboratory tests were unremarkable except for slight raised Erythrocytic Sedimentation Rate (ESR).

The CECT abdomen did not reveal any radiological finding that could be attributed to low-grade fever. However, careful examination revealed agenesis of celiac axis consequent to which both common hepatic and splenic arteries were seen arising directly from abdominal aorta, just cranial to superior mesenteric artery at L1 vertebral body level (Figures 1–3). Further evaluation

Figure 1: Post-contrast MIP CT image in trans-axial plane shows separate origins of common hepatic (thick white arrow) and splenic (thin white arrow) arteries directly from aorta at the same level.

Figure 2: Post-contrast MIP CT image in sagittal plane shows origin of common hepatic artery just cranial to superior mesenteric artery at the same vertebral level (white arrow) with obvious narrowing at its origin before coursing superiorly in the form of elephant trunk.

Figure 3: 3D-VRT-CT image shows origin of common hepatic & splenic arteries just cranial to superior mesenteric artery as (white arrow) with elephant trunk sign of Rajul.
revealed flattening and narrowing of the proximal part of the common hepatic & splenic arteries which were seen coursing superiorly in elephant-trunk manner to supply its territory. As the patient did not have any clinical signs associated with narrowing of above-described arteries, the final diagnosis of agenetic celiac axis with congenital narrowing and elephant-trunk course of common hepatic & splenic arteries was made.

The left gastric artery was noted arising from the common hepatic artery. No evidence of any similar abnormality is noted in any other vessel.

Discussion

With increasing utilization of CECT abdominal examinations for a variety of clinical conditions, more and more variations in branches of abdominal aorta are being detected. However, among these agenesis of celiac axis is quite uncommon being reported up to 2% in many studies [1−3,5]. In such cases common hepatic & splenic arteries may have anomalous origin from the abdominal aorta [5]. The overall incidence of aortic origin of these arteries has also been reported to be up to 2% [5−8].

However, in previously reported cases of anomalous origin of above-described arteries abdominal aorta, asymptomatic congenital narrowing with elephant trunk course as seen in our case has not been reported, that has been described in our article as positive elephant trunk sign of Rajul. This finding is important as any further narrowing of already-compromised lumen of common hepatic & splenic arteries with advancing age and atherosclerosis may lead to ischemic changes in their territorial distribution leading to related morbidity. This finding will also be relevant, should any form of hepatobiliary or pancreatic surgery including laparoscopic or robotic procedures are contemplated in future.

The important differential diagnosis in our case is atherosclerotic narrowing of vessel lumen which can be easily excluded in our case by young age and absence of mural thickening/fatty-plaques/mural calcification.

Conclusion

Agenesis of celiac axis is a rare congenital anomaly that can be detected on the routine MDCT abdominal examinations. In these cases, common hepatic & splenic arteries may be seen, arising from abdominal aorta or superior mesenteric artery. In cases of abdominal aortic origin of common hepatic artery, if elephant-trunk course of these arteries is noted then an extra-note should be made of the presence or absence of elephant trunk sign of Rajul i.e. congenital narrowing of lumen as this may become severe with advancing age and may become symptomatic.

References


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