Congenital Incomplete Fusion of Superior Mesenteric Artery Mimicking Dissection

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Conflict of interest: None declared

Patient: Male, 62
Final Diagnosis: Superior mesenteric artery anatomic variant
Symptoms: Abdominal pain • diarrhea • transient ischemic attacks
Medication: —
Clinical Procedure: CT of abdomen and pelvis
Specialty: Surgery

Objective: Congenital defects/diseases
Background: Both spontaneous SMA dissection and anatomical variants of GIT vasculature are well known entities. We present a case initially diagnosed as an SMA dissection on CT, but upon detailed review of the imaging findings was considered to be incompletely fused ventral segmental arteries – a rare anatomic variant not well described before. This finding is clinically significant, as it can mimic a vascular dissection and such a wrong diagnosis will lead to unnecessary investigation and intervention.

Case Report: A 62-year-old male patient presented with abdominal pain of uncertain etiology. The initial CT revealed an abnormal appearance of the superior mesenteric artery (SMA) which was diagnosed as SMA dissection. However, the appearance of this ‘dissection’ was unusual and there was a mismatch between the clinical presentation and radiological findings. The scan was reviewed and a 3D reconstruction of the abdominal aortal and visceral arteries was performed. The abnormal appearance of the SMA was deemed to be from a congenital anatomical variant. A review of the embryological origin of gut vasculature provides a likely explanation for this appearance.

Conclusions: Ours is an unusual case of a developmental variant that has not been well described hitherto. Attention to the ancillary radiological signs and understanding the embryological origin of the abdominal vasculature is important to distinguish such variants from pathology.

MeSH Keywords: Anatomic Variation • Dissection • Mesenteric Artery, Superior

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Background

Spontaneous superior mesenteric artery (SMA) dissection is a known entity, the appearance of which has been well documented and classified by various authors. We present a case initially diagnosed as an SMA dissection but upon detailed review of the imaging findings was considered to be incompletely fused ventral segmental arteries.

Incomplete and anomalous regression of the primitive arteries has been implicated as a cause for the earlier described variants of gastrointestinal tract (GIT) vasculature. However, the finding of incompletely fused ventral segmental arteries has not been described in the literature before. This finding is clinically significant, as it can mimic a vascular dissection and such a wrong diagnosis will lead to unnecessary investigation and intervention.

Case Report

A 62-year-old Chinese man with a background of hypertension, transient ischemic attack and vertebrobasilar insufficiency presented to the emergency department complaining of central abdominal pain and 3 episodes of blood-stained stools over 24 hours. The pain was slow in onset, generalized and constant, with no radiation. There was no history of fever, vomiting, constipation, or jaundice.

Examination elicited mild tenderness at the right iliac fossa and around the umbilicus. Proctoscopy demonstrated internal hemorrhoids, which did not show any active bleeding. The full blood count (FBC) revealed an elevated total leukocyte count (TLC) of 19.1×10^9/L with 82% neutrophils. The routine urine microscopy and chest radiograph were unremarkable. A provisional diagnosis of abdominal sepsis was made and a CT scan was performed (Figure 1).

The CT scan was initially reported as SMA dissection although its appearance did not fit into known classification systems described by Luan et al. or Yun et al. [1,2]. Also, there was an apparent mismatch between the radiological diagnosis and the clinical signs and symptoms. Therefore, a review of the original CT images was warranted, including 3D reformatting (Figure 2). After a review of embryology of gastrointestinal tract (GIT) vasculature, this unusual appearance was deemed to be from an abnormality of fusion rather than a dissection.

The patient was managed conservatively in the ward with bowel rest, intravenous fluids, and antibiotics, and subsequently improved. A follow-up scan after 3 weeks showed a stable appearance of the SMA.

Figure 1. Contrast-enhanced CT images of the abdomen in the arterial phase in oblique-coral (A–D) and axial planes (E, F) show abnormal appearance of the SMA (closed arrow) with a linear hypodense ‘septum’ dividing its lumen into 2 parts, resulting in a “double-barrelled” appearance. This ‘septum’ starts 2.5 cm distal to the ostium and measures 4 cm in length. The inferior pancreaticoduodenal artery arises from the right aspect of the SMA (B, open arrow). No intra-luminal thrombosis, surrounding fat-stranding, signs of bowel ischemia, or free fluid is evident.
Discussion

The ‘septum’ of the ‘dissection’ in our patient appeared abruptly in the straight descending portion of the SMA (distal to the curve and 2.5 cm from the ostium). This ‘septum’ was as thick as the vessel wall, instead of being of imperceptible thickness. Both lumens were nearly of the same size, rather than a larger false lumen. In a true dissection, the false and true lumens are orientated superior and inferior, respectively, but appeared side-by-side in our case. Neither lumen in our case was blind-ending, with the apparent “false” lumen continuing smoothly into the inferior pancreaticoduodenal artery. This appearance can be explained by a hypothesis based on the knowledge of embryological origin of GIT vasculature and previous reports of anatomical variants in other major visceral arteries.

The fetal vascular system consists of a pair of dorsal aortic anlagen (Figure 3). Each anlage gives off branches that can be divided into 3 groups [3]:
1. Ventral segmental branches;
2. Lateral segmental branches;
3. Dorsolateral intersegmental branches.

The 2 dorsal aortas fuse together to form a single dorsal aorta by the middle of week 5. The pairs of ventral segmental arteries (from each aortic anlage) also fuse in the midline, forming the celiac trunk as well as the superior and inferior mesenteric arteries [3]. If this fusion is incomplete, it will result in an appearance as seen in our patient.

The developmental variants of arteries supplying the GIT are not infrequent [4]. The frequency of anatomical variations in the celiac and superior mesenteric arteries ranges from 3.6% to 10.1% [5,6]. The available literature frequently describes variations such as absent celiac axis, single celiac axis, celiacomesenteric origin (common origin of coeliac and SMA), celiac-bimesenteric origin (common origin of the celiac and both mesenteric arteries), and replaced right or left hepatic arteries [4–6]. Incomplete and anomalous regression of the primitive arteries has been implicated as a cause for these [7].

However, the finding of incompletely fused ventral segmental arteries has not been described in the literature before. This finding is clinically significant, as it can mimic a vascular dissection and such a wrong diagnosis will lead to unnecessary investigation and intervention, ultimately exposing the patient to treatment-related morbidity/mortality.

Figure 2. 3D volume-rendered reconstruction of the abdominal aorta shows a dilated SMA trunk (solid arrow) with a septum (dotted arrow) dividing its lumen into 2 parts. The inferior pancreaticoduodenal artery (black arrowheads) is seen coming off the right lateral aspect of the distal SMA trunk.

Figure 3. A simplified depiction of embryogenesis of fetal vasculature at beginning (A) and end (B) of the 5th week. 1. Dorsolateral intersegmental branches of the aorta. 2. Dorsal aorta. 3. Lateral segmental branches of the aorta supplying the mesonephros, later forming the renal arteries. 4. Ventral segmental branches of the aorta supplying the gastrosplenic tract. 5. Ventral branches of the dorsal intersegmental branches of the aorta forming the intercostal arteries. 6. The gastrointestinal tract.
Conclusions

Ours is an unusual case of a developmental variant that has not been previously well described. Understanding the embryological origin of the abdominal vasculature is important in distinguishing such variants from pathology. Attention should be paid to the ancillary radiological signs and the patient's clinical status to confidently regard this as a benign entity.

Statement

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