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IMAGES in PAEDIATRIC CARDIOLOGY

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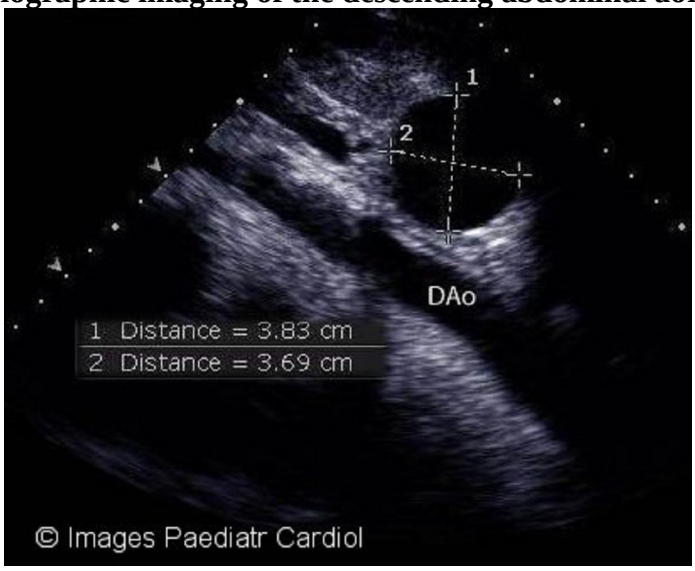
Introduction

Loey's Dietz syndrome is a disorder of connective tissue caused by a mutation in the genes that encode transforming growth factor (TGF) beta receptor 1 and 2.[1] It is an autosomal dominant disorder similar to Marfan's syndrome but with a more aggressive clinical course.[2] Patients with Loey's-Dietz syndrome have progressive dilatation of the aortic root that can lead to aortic dissection and rupture. The location of non-aortic arterial aneurysms may be wide spread but often occur in the head and neck vessels.[1]

Patient

We present a case of a 13 year old boy with Loey's Dietz syndrome and a history of a David procedure (valve sparing aortic root replacement) when he was 9 months old with aortic valve and aortic root replacement at 2 years of age with subsequent post-operative complete heart block requiring pacemaker insertion. He ultimately underwent a Bentall procedure at 12 years of age. Upon routine cardiology clinic follow-up an incidental finding of a cystic structure in the liver was noted during echocardiography (Figure 1).

Figure 1: A 3.83 cm by 3.69cm echo dark structure is seen within the liver on subcostal echocardiographic imaging of the descending abdominal aorta (Dao).



He was asymptomatic from a cardiac and gastrointestinal standpoint. The remainder of his echocardiogram showed normal mechanical aortic valve function and normal left ventricular function. His pacemaker function was normal.

An abdominal CT was obtained to further characterize the lesion. Compared to a CT obtained 4years previously a new, well demarcated, heterogenously enhancing mass or aneurysm was identified between the lesser curvature of the stomach and the liver measuring 3.9 cm x 5.3 cm x 4.3 cm (Figure 2 and Figure 3). A small branch off the left gastric artery appeared to enter the left anterior aspect of the mass (Figure 4).

Figure 2: Abdominal CT axial image demonstrating the large heterogenous mass or pseudoaneurysm (arrow) between the stomach and the liver. It measures 3.9 cm (anterior-posterior) x 5.3 cm (transverse) x 4.3 cm (cranio-caudal).

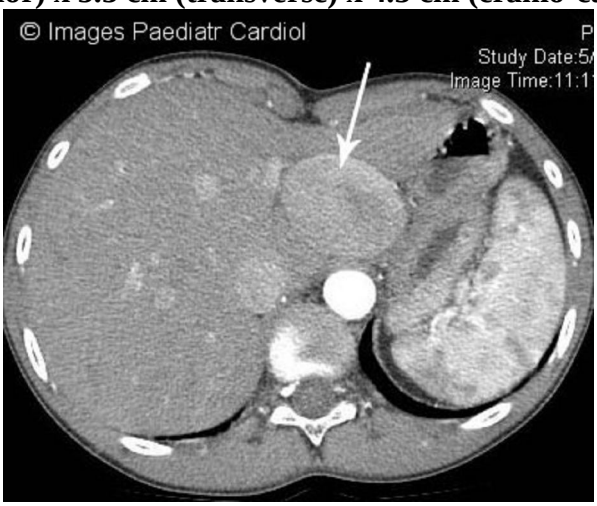


Figure 3: CT scout image demonstrating the mass vs pseudoaneurysm (arrow). Contrast seen on this image is from a recent prior outside CT scan.

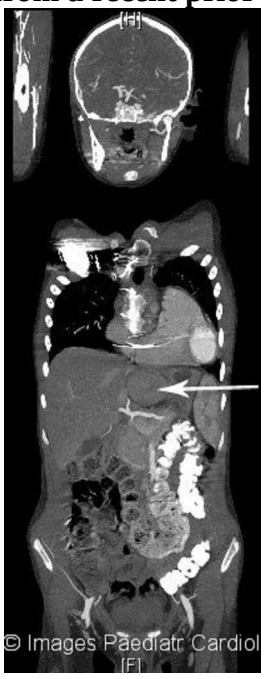
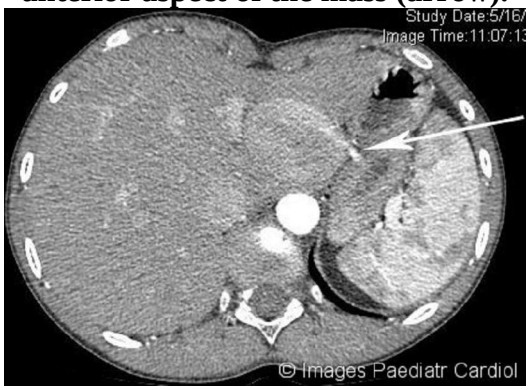


Figure 4: CT axial images showing a small branch off the left gastric artery entering the left anterior aspect of the mass (arrow).



An abdominal ultrasound with Doppler performed after the CT scan was consistent with a patent aneurysm or pseudoaneurysm impressing upon the left hepatic lobe (Figure 5 and Figure 6). Color Doppler flow into the aneurysm was demonstrated from the left gastric artery. No thrombus was identified.

Figure 5: Abdominal ultrasound showing the pseudoaneurysm to measure 4.4 cm x 3.98 cm in this plane.

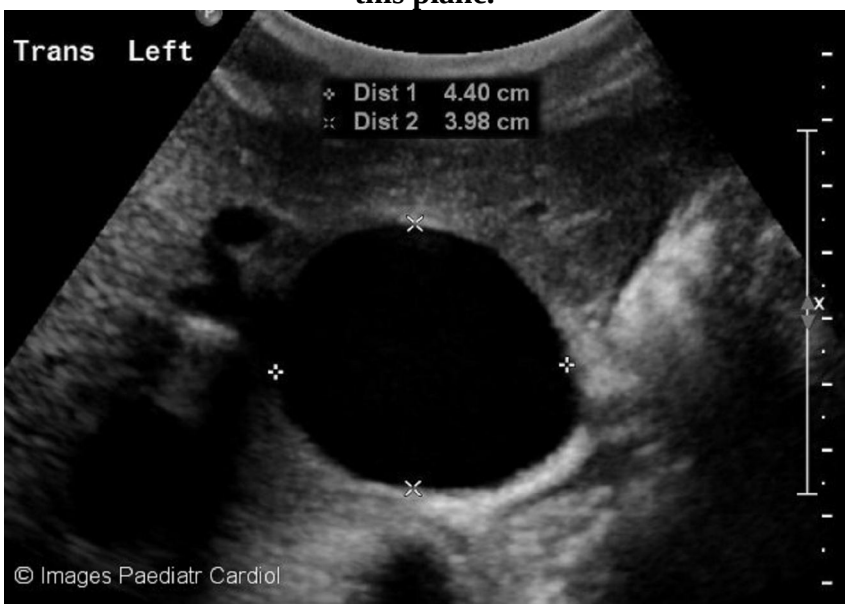
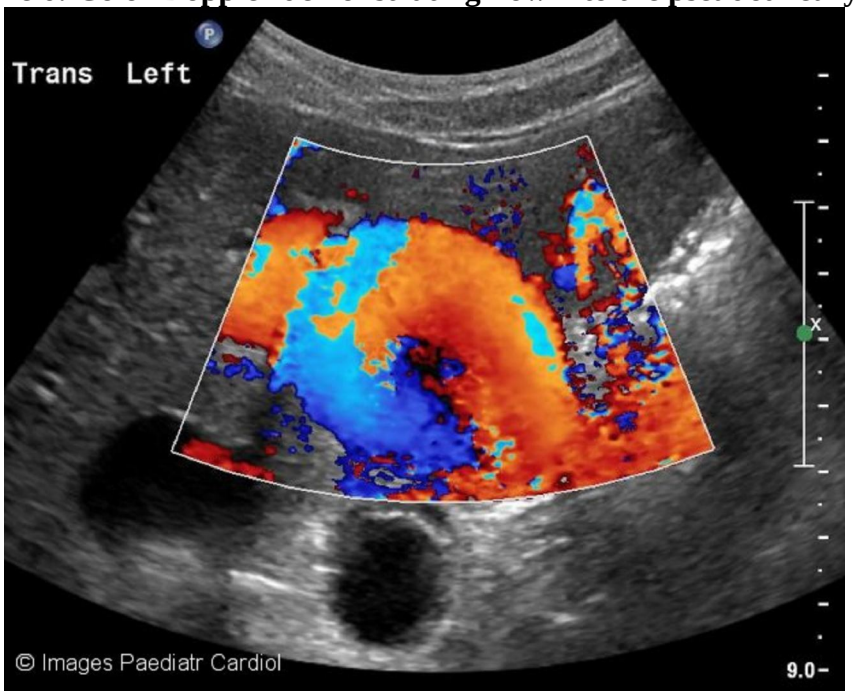


Figure 6: Color Doppler demonstrating flow into the pseudoaneurysm.



Successful embolization of what was found to be a left gastric artery pseudoaneurysm (Figure 7) by interventional radiology was performed using 5 detachable GDC coils (Boston Scientific Corporation, Natick, MA) in the left gastric artery at the neck of the pseudoaneurysm (Figure 8).

Figure 7: Selective angiogram demonstrating flow into the left gastric artery and feeder vessel (black arrowheads) filling the large pseudoaneurysm (white arrows).

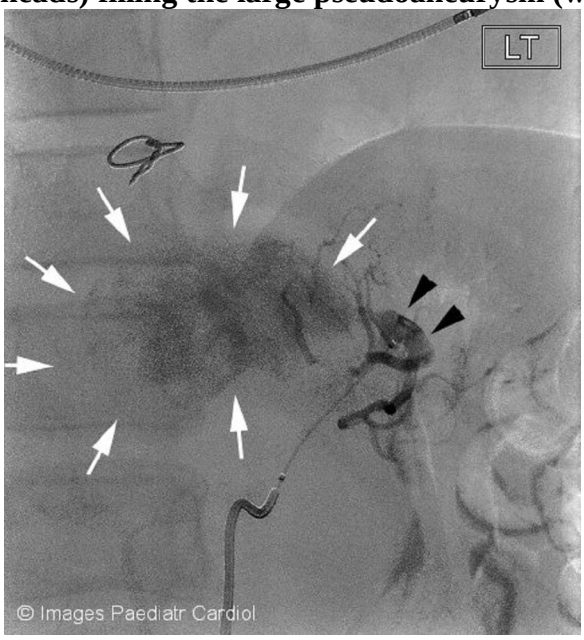
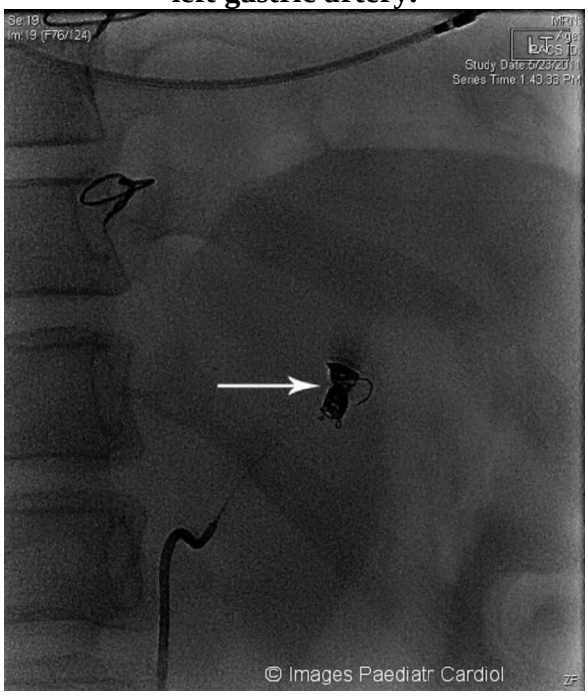


Figure 8: Selective angiogram after placement of GDC coils (arrow) with no flow seen to the left gastric artery.



He was also noted to have a markedly ectatic superior mesenteric artery (SMA) with several irregular beaded second and third order vessels from the SMA thought to possibly represent manifestations of his disease. Two months following the coil embolization, an abdominal CT scan with contrast demonstrated the excluded large pseudoaneurysm with no enhancement noted within it. Nine months later, an abdominal ultrasound showed interval resolution of the embolized and thrombosed pseudoaneurysm without any residual abnormalities demonstrated in the area. There were no new aneurysms identified. He remains stable from a cardiac perspective as well.

Discussion

Loey's Dietz syndrome is a disorder characterized by an aggressive arteriopathy resulting in arterial tortuosity and aneurysms. These patients can suffer significant morbidity and mortality from aortic or arterial dissection or rupture. Dissection of the thoracic aorta is the most common cause of death in patients with this syndrome.[3] However, more than 50% of patients have more distal aneurysms. Our patient illustrates the importance for clinicians to recognize aneurysms or dissections early so appropriate interventional or surgical management can be undertaken before a catastrophic event occurs.[4] Due to the progressive and severe nature of this disease, it has been suggested that annual MRA from head to pelvis as well as every 3-6 month echocardiograms for surveillance may be appropriate for these patients.[5,6]

References

1. Van Hemelrijk C, Renard M, Loeys B. The Loeys-Dietz syndrome: An update for the clinician. Curr Opin Cardiol. 2010;25(6):546-551.

2. Kalra VB, Gilbert JW, Malhotra A. Loeys-Dietz syndrome: Cardiovascular, neuroradiological and musculoskeletal imaging findings. Pediatr Radiol. 2011;41(12):1495-1504.

3. Loeys BL, Chen J, Neptune ER, et al. A syndrome of altered cardiovascular, craniofacial, neurocognitive and skeletal development caused by mutations in TGFBR1 or TGFBR2. Nat Genet. 2005;37(3):275-281.

4. Loeys BL, Schwarze U, Holm T, et al. Aneurysm syndromes caused by mutations in the TGF-beta receptor. N Engl J Med. 2006;355(8):788-798.

5. Williams JA, Loeys BL, Nwakanma LU, et al. Early surgical experience with Loeys-Dietz: A new syndrome of aggressive thoracic aortic aneurysm disease. Ann Thorac Surg. 2007;83(2):S757-63.

6. Yetman AT, Beroukhim RS, Ivy DD, Manchester D. Importance of the clinical recognition of Loeys-Dietz syndrome in the neonatal period. Pediatrics. 2007;119(5):e1199-202.

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