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Background

- Complex arch anomalies including Double Aortic Arch (DAA) has been associated with PHACE syndrome.
- Atresia of the proximal segment of the left arch in DAA is extremely rare and can pose a diagnostic and management conundrum due to formation of a loose vascular ring.

Case Presentation

- A 5-year-old male with PHACE syndrome and an incidental finding on head imaging of a right aortic arch (RAA) with an aberrant left subclavian artery at an outside hospital.
- Clinically, he was asymptomatic, growing well with no respiratory symptoms or dysphagia.
- Cross-sectional imaging with gated CT angiogram was performed which aided in the diagnosis of this rare DAA variant.

Diagnostic Images

Figure (1A) and Figure (1B): 3 D reconstruction using multiplanar format ting showing the double aortic arch with ipsilateral head and neck vessels and an atretic proximal left aortic arch. Asc Ao: Ascending Aorta, LCC: Left common carotid, RCC: Right common carotid, LSCA: Left subclavian artery, RSCA: Right subclavian artery.

Figure 2: The presumed atretic proximal left aortic arch ligament between the right and left aiches is shown as a blue bar. The left sided ductal ligament connects the dorsal aorta with the distal pulmonary trunk as evidenced by the presence of ductal dimple under the left common carotid artery (yellow bar).

Discussion

- Diagnostic clues include:
  - Proximity of the RCC and LCC arteries.
  - Symmetric four vessel sign at the thoracic inlet.
  - Posterior course of a patent segment of the atretic left arch.
  - Presence of the ductal dimple under the LCC artery.
- Surgical intervention has been performed in limited reported cases when patients had symptoms of airway compression or associated left to right shunt related to PDA.

Conclusion

- Given the "loose vascular ring" and asymptomatic nature, patient specific discussion should be considered with the family to assess optimal management strategy.
- Complementing cross sectional imaging with baseline echocardiography even in asymptomatic cases can be useful in the diagnosis of this rare DAA variant.

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