Persistent 5th Aortic Arch in Patient with Known Intrauterine Thalidomide Exposure

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Clinical History

A 52 year-old male with a history of multiple congenital defects from intrauterine thalidomide exposure presented to his primary care physician with tenderness and a mass-like sensation in his right supraclavicular and right neck area. Although the patient had some mild tenderness on palpation, there was no severe pain, restriction of motion, fever, or nuchal rigidity. The supraclavicular region was “boggy” on palpation, but no mass was identified.

Findings

Sagittal multiplanar reformat (MPR) and 3-D volume rendered reconstruction (VRT) from a chest CT with contrast showed bifurcation of the aortic arch into two arches (4th and persistent 5th) at the level of the brachiocephalic artery origin. The two arches fused after a few centimeters, and the remainder of the aorta was normal. Evaluation of the osseous structures on the chest CT demonstrated fusion anomalies of the vertebrae and ribs (Figure 2). The neck CT demonstrated additional common bony defects associated with thalidomide exposure, including a cleft palate (Figure 3). No abnormality was present in the right supraclavicular to explain the patient’s symptoms.

Discussion

Persistence of the 5th aortic arch is a rare congenital vascular anomaly which may be seen in isolation or associated with other congenital cardiac or vascular anomalies. Many patients present at birth due to associated defects such as ventricular septal defect, pulmonic stenosis, pulmonary atresia, interruption of the aortic arch, and complex congenital heart disease. This anomaly exists in two distinct forms, a systemic-to-systemic connection and a systemic-to-pulmonary connection. In the systemic-to-systemic type, the anomalous arterial branch begins at the origin of the brachiocephalic trunk and reconnects with the descending aorta, as in this case. In the systemic-to-pulmonary type, the 5th arch connects with the embryologic remnant of the 6th aortic arch, which is usually the left pulmonary artery. There is no reported association of persistent 5th arch anomalies with thalidomide in humans, but animal studies have shown an association between various arch anomalies and intrauterine thalidomide exposure.

REFERENCES

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