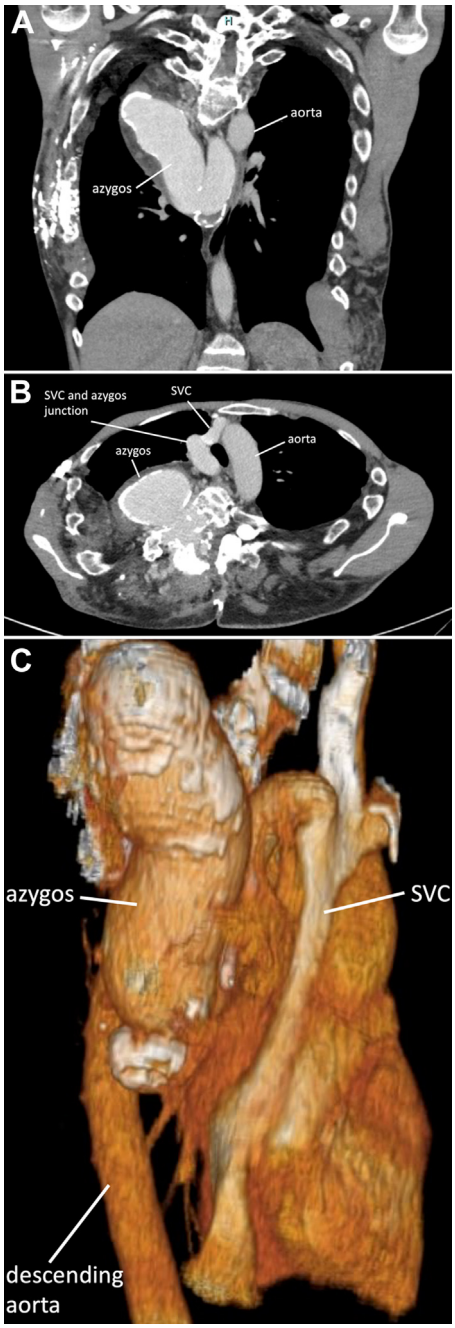
Asymptomatic azygos phlebectasia associated with CLOVES (congenital lipomatous overgrowth, vascular malformations, epidermal nevis, spinal/skeletal anomalies/scoliosis) syndrome

Claire A. Ostertag-Hill, MD,[a] Steven J. Fishman, MD,[a] Melisa Ruiz-Gutierrez, MD, PhD,[b] *and* Rush H. Chewning, MD,[c] *Boston, MA*

A 43-year-old man was referred to our multidisciplinary vascular anomalies center for evaluation and management of multiple vascular malformations and congenital anomalies. He was born with an extensive lymphatic malformation (LM) of his thorax and back with spinal canal involvement, a left scrotal LM, a chest wall capillary malformation, lower extremity limb length discrepancy, and scoliosis. During childhood, he had undergone debulking of his truncal LM, spinal decompression, spinal radiotherapy, and interferon-alpha treatment. To evaluate disease involvement, he underwent computed tomography, which again demonstrated his thoracic LM and additionally demonstrated significant anomalous, fusiform dilatation of his azygos vein, consistent with azygos phlebectasia (*A*, *B*/Cover, and *C*). The dilatation measured 6.2 cm × 4.5 cm and contained no thrombus. He denied any associated symptoms or history of chest trauma or pulmonary embolus. To better understand the history of this azygos dilatation, thoracic imaging from 5 years prior was reviewed, which showed stability in size. The superior vena cava, inferior vena cava, and hemiazygos vein were patent with normal anatomy. Considering this patient's constellation of vascular malformations and anomalies, he was diagnosed with CLOVES (congenital lipomatous overgrowth, vascular malformations, epidermal nevis, spinal/skeletal anomalies/scoliosis) syndrome, an extremely rare condition marked by congenital lipomatous overgrowth, vascular malformations, epidermal nevi, and scoliosis/skeletal/spinal anomalies in the PIK3CA-related overgrowth spectrum.[1] Given the high likelihood that his azygos phlebectasia represented a congenital vascular anomaly as part of his CLOVES syndrome, the large amount of blood flow through this vessel, and the demonstrated stability on imaging for ≥5 years, conservative management was recommended.

Phlebectasia refers to congenital venous dilatation[2] and is sometimes used interchangeably with venous aneurysm. Azygos vein aneurysms are extremely rare, with a recent review of 57 reported cases suggesting that most azygos vein aneurysms are idiopathic, more commonly saccular, and identified incidentally.[3] However, central and thoracic phlebectasia,



From the Department of Surgery,[a] Division of Hematology/Oncology,[b] and Division of Vascular and Interventional Radiology,[c] Vascular Anomalies Center, Boston Children's Hospital.

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E-mail: Rush.Chewning@childrens.harvard.edu.

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including azygos and hemiazygos phlebectasia, are common with CLOVES syndrome.[2] The presence of concurrent anomalies in lymphatic and venous channels, as seen with the CLOVES, Klippel-Trenaunay, and Proteus (progressive deformities of bone, skin, and soft tissue) syndromes, are likely related to their shared embryonic precursor.[2] Although these patients have an elevated risk of pulmonary embolism secondary to central, thoracic, and/or peripheral phlebectasia, the significance of azygos phlebectasia specifically is unclear.[2,4] Although we have elected to not yet initiate anticoagulation therapy, prophylactic anticoagulation might warrant consideration in a future perioperative setting.

The patient provided written informed consent for the report of his case details and imaging studies.

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