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Aneurysms of the azygos vein

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ABSTRACT

Objective: Azygos vein aneurysms (AVAs) are uncommon and infrequently diagnosed. When confronted with a patient presenting with an AVA, physicians can rely on only a few case reports after an extensive literature search. To date, no guideline, no rule, and no review on the optimal treatment strategy for these patients exist.

Methods: A PubMed and MEDLINE database search for papers and case reports describing AVA was performed. Cases from our own institutions were also reviewed.

Results: The literature search identified 57 published case reports that were reviewed for inclusion. Of those published cases, etiologic factors can be classified into idiopathic, acquired, and traumatic causes. Most AVAs are limited to the azygos arch, a congenital anatomic weak point. Clinical symptoms generally remain nonspecific. Computed or magnetic resonance tomography scans are effective diagnostic tools, although the optimal therapeutic plan remains unclear. Complications include rupture, thromboembolism, mediastinal mass effects, and pulmonary artery hypertension.

Conclusions: Conservative treatment along with oral anticoagulation may be reasonable for some AVAs, but to date, there is no clear guideline or evidence-based threshold for surgical or interventional therapy. In review of the existing data and from our clinical and scientific knowledge, interventional or surgical treatment should strongly be considered in cases with clinical symptoms, pulmonary embolism or pulmonary arterial hypertension, thrombus formation within the AVA in patients with oral anticoagulation or for patients with a contraindication to oral anticoagulants, considerable increase in diameter or compression of adjacent structures, saccular AVA, or an underlying connective tissue disease. The most common procedure is surgical ligation of the AVA, although endovascular occlusion of the aneurysms is becoming more frequent. (J Vasc Surg: Venous and Lym Dis 2017;5:576-86.)

Azygos vein (AV) aneurysms (AVAs) are extremely rare. The first publication of a patient with an AVA originates from 1963.[1] To date and to the best of our knowledge, only 57 cases of AVA have been published in the literature.

The AV is the cranial continuation of the right ascending lumbar vein. Passing through the aortic hiatus of the diaphragm, it is joined by the hemiazygos vein. It extends rightward and forward, arching over the right bronchus to drain into the superior vena cava (SVC) on the right tracheobronchial angle.[2,3] In radiologic studies, the normal diameter of the AV is defined as no more than 1 cm.[4] Whereas the diameter of the azygos arch is variable, it may reach 1.5 cm in healthy subjects or in pregnancy.[3] In postmortem examinations, the mean diameter of the AV measures 0.4 cm at its origin and 0.8 cm at its termination.[5]

In general, an aneurysm is defined as a focal dilation of a blood vessel involving all three layers of the vessel wall.[6] In the extremities, a venous aneurysm is defined as a persistent venous dilation with a diameter twice the size of a normal vein.[7] Owing to filling-, pressure-, and breathing-dependent variability in size of AVAs,[8-10] it is—in our opinion—reasonable to increase the threshold for the definition of an AVA to 2.5 times the normal diameter of the AV. Therefore, we define an AVA in the adult patient as a focal, persistent dilation of the AV exceeding 2.5 cm and exceeding 3.75 cm in the AV arch. To the best of our knowledge, there is no other congruent definition of an AVA in the literature that is used as a diagnostic threshold.

Corresponding to morphologic descriptions of aortic aneurysms, AVAs can be classified by their macroscopic shape and size into fusiform-type aneurysms or saccular-type aneurysms on cross-sectional computed tomography (CT) or magnetic resonance imaging (MRI).[11] Fusiform AVAs (or spindle-shaped aneurysms) are circumferential dilations of the AV with variable diameter and length; saccular AVAs are localized dilations within a portion of the AV wall.[11]

Some authors have suggested that fusiform or saccular aneurysm development depends on the cause of AVA

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formation.[12] However, other authors reported no connection between the AVA's origin and its morphologic presentation.[11] Because of the small number of cases, it is not yet possible to resolve this controversy.

When confronted with a patient presenting with an AVA, physicians can rely on only a few case reports after an extensive literature search. To date, no guideline, no rule, and no review on the optimal treatment strategy for these patients exist. This review provides an overview of the mechanisms behind an AVA, the clinical symptoms, and the diagnostic tools and offers a treatment plan for those patients.

METHODS

An electronic search of PubMed and MEDLINE databases for all relevant literature published up to 2016 was performed. The search was limited to any reported cases or series of true AVAs. Also, cases from our own institutions were reviewed. Reports were included when the full text was available or when the abstract provided sufficient information. Reports were excluded in case of insufficient details.

RESULTS

The literature search identified 57 published case reports that were reviewed for inclusion. Because of the retrospective nature of the systematic review and lack of statistical power in the recruited studies, statistical analysis was not reasonable. Therefore, all reported cases were tabulated in Table I, which includes 45 patients; 12 case reports were not included in the table because either the full text or the abstract was not available. Also, case reports describing post-traumatic pseudoaneurysms of the AV were not included and were not counted as AVAs. In our institution, we were able to identify one patient with an AVA who we treated within the last 4 years. Moreover, within the same time period, we treated 20 congenital heart disease patients with various forms of thoracic venovenous collaterals and thoracic venous aneurysms using Amplatzer vascular plugs (St. Jude Medical, St. Paul, Minn).

With the exception of two cases being documented in a pediatric patient[13,14] and one case being documented in an adolescent,[15] all documented AVAs were diagnosed in adult patients. About two-thirds of those patients were female and one-third were male. In the documented cases, AVAs are limited or adjacent to the azygos arch while the remaining part of the AV usually appears normal.[11]

DISCUSSION

Origin

Various causes of AVAs have been described and allow differentiation between idiopathic AVA, acquired AVA due to pressure or volume overload, and traumatic AVA.

Idiopathic AVA. Idiopathic AVAs are generally considered congenital venous malformations.[12] Alternative causes for AV enlargement must be ruled out for the diagnosis of an idiopathic AVA. During vasculogenesis, the AV derives from the upper right supracardinal vein; the azygos arch arises from the upper segment of the right posterior cardinal vein.[2] As most AVAs are localized at this anatomic junction, some authors have described it as a congenital anatomic weak point[16,17] in the formation of AVA. Most AVA patients lack contributive acquired causes for AVA, and the majority of AVAs are limited to the azygos arch.[11] Therefore, it is hypothesized that the upper segment of the posterior cardinal vein may be responsible for AVA formation.[2,11] Such findings support the postulation that the origin of most AVAs may be congenital in nature.[11]

Connective tissue disorders may represent another congenital cause of AVA formation. D'Souza et al[18] have described the only case of AVA in a patient with Ehlers-Danlos syndrome type IV so far.

Acquired AVA. It has been hypothesized by some authors that pressure or volume overload due to collateral circulation through the AV system may be responsible for dilation of the AV.[2,3,15] Underlying causes of AVA formation that have been proposed include portal hypertension,[2,10,12] arteriovenous fistula,[19] cardiac decompensation,[2,3,11] pregnancy,[11] and compression of the SVC due to neoplasms[10,11] or thrombus formation.[11] Iatrogenic compression of the SVC that causes an increased flow through the AV and the formation of an AVA has also been suggested.[20]

Some authors have postulated that high central venous pressure from cardiac decompensation may be the most frequent cause of AVA formation.[17] However, from our review of the literature and larger case series,[11] most AVA patients are lacking acquired causes for AVA formation. Because of the limited number of patients with AVA, it is impossible to define the most frequent reason for AVA formation with absolute certainty, but in review of the published AVA cases, all but two of those cases were of idiopathic origin (Table I).

Traumatic AVA. Traumatic pseudoaneurysms of the AV resulting from blunt injury or catheter insertion have been described rarely[12,21] and are not addressed further in this review.

Pathophysiologic considerations

The exact pathophysiologic mechanisms underlying the formation of AVA are unknown,[12] and the venous wall after surgical resection has been described to be extremely thin.[10,19] In accordance with the pathogenesis of arterial aneurysms, this may be due to a congenital weakness or degenerative changes in the venous wall caused by abnormalities in the connective tissue.[22] Other authors have suggested onset of a venous hypertrophic process or endothelial phlebosclerosis, a degenerative process characterized by the loss of connective tissue and smooth muscle cells, as the leading pathologic mechanism.[12]