Two giants: Giant cell arteritis causing a giant pulmonary artery aneurysm

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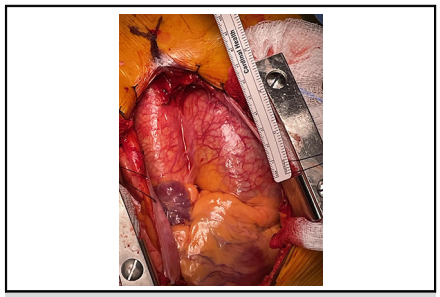

A giant aneurysm of the PA in an elderly woman with GCA.

CENTRAL MESSAGE

GCA involving the PA is rare and can lead to a giant PAA. Local inflammation in vasculitis-induced aneurysms presents unique surgical challenges.

Giant cell arteritis (GCA) is an autoimmune vasculitis most commonly involving the temporal artery, aorta, and great vessels. Isolated involvement of the pulmonary artery (PA) due to GCA is rare, with only 1 prior documented case.[1,2] Pulmonary artery aneurysms (PAAs) occur with an incidence of 1 in 14,000 and may result in lethal sequelae.[3] They are often asymptomatic and discovered incidentally. We report a case of GCA causing a giant aneurysm of the pulmonary trunk and arteries. Per institution, Institutional Review Board approval was not required. The subject provided informed written consent for the publication of the study data.

CLINICAL SUMMARY

A 76-year-old woman was referred for an incidentally noted true aneurysm of the proximal PA measuring 9 cm (Figure 1, *A* and *B*). Her only presenting symptom was exertional dyspnea. Medical comorbidities included hypertension and hyperlipidemia without a history of other cardiopulmonary or rheumatologic conditions. Her laboratory workup demonstrated antinuclear antibody titers (<1:160) and positive anti-La, but was otherwise unremarkable with normal inflammatory markers. Transthoracic echocardiography demonstrated severe pulmonic regurgitation with preserved ejection fraction. Computed tomography angiography revealed a 9-cm true aneurysm of the main PA with mild dilation of the right and left PAs (3.1 and 3.0 cm, respectively). Echocardiography demonstrated severe pulmonic valve insufficiency (PVI) with normal right ventricle function (Figure E1).

Given the aneurysmal size, dyspnea, and severe PVI, the patient was offered surgical repair. The patient subsequently underwent surgery with cardiopulmonary bypass, and the giant PAA (GPAA) was noted to be very inflamed and densely adhered to the pericardium and ascending aorta. Opening the PA unveiled annular dilation of the pulmonic valve and the right ventricular outflow tract, with a clear transition zone before aneurysmal degeneration. The pulmonic valve was replaced with a 27-mm bioprosthetic valve (Inspiris Resilia, Edwards Lifesciences, Irvine, Calif), and the GPAA was repaired with aneurysmorrhaphy in an inverted T-shape (Figure 2). A small amount of bleeding was noted from a short segment of the ascending aorta where the aneurysm was previously adhered. Because of denudation and friability of the aortic wall after freeing the inflamed aneurysm, this short segment of the ascending aorta was replaced with an interposition 28-mm Gelweave Dacron tube graft (Terumo Aortic, Sunrise, Fla).

Unexpectedly, pathologic review revealed transmural chronic inflammation with medial degeneration and disruption of the elastic layer by lymphocytes, histiocytes, and giant cells thought to be most consistent with GCA (Figure 1, *D*). Treponemal, mycobacterial, fungal, and immunoglobulin G-4 stains were negative. Rheumatological workup was not suspicious for any other signs of active vasculitis.

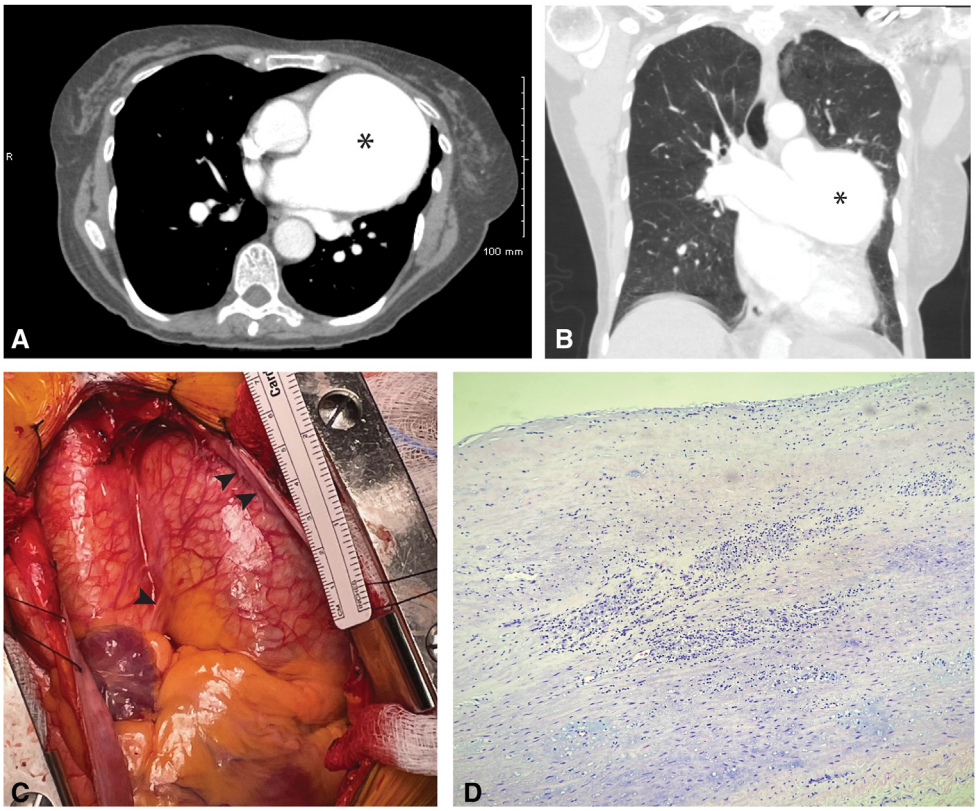

FIGURE 1. A, Axial section of cardiac computed tomography angiographic scan demonstrating main PAA (*asterisk*) measuring 9 cm. B, Coronal section of cardiac computed tomography angiographic scan showing dilatation of pulmonary trunk (*asterisk*) and the *right* and *left* PAs. C, Intraoperative photograph of inflamed and dilated PA with adhesions to the aorta and pericardium (*black arrows*). The aorta dwarfs in comparison with the aneurysmal pulmonary trunk. D, Histopathology of PAA showing transmural chronic inflammation with medial degeneration and disruption of the elastic layer by lymphocytes, histiocytes, and giant cells.

There was no history or evidence of ocular manifestations of GCA on examination by ophthalmology. Because of definitive treatment of the GPAA operatively and the lack of rheumatological sequelae, steroids were not recommended. Given her normal preoperative inflammatory markers and absence of systemic manifestations, outpatient positron emission tomography scan was recommended to assess for residual local or distant active vasculitis.

DISCUSSION

We did not expect GCA to be the cause of the GPAA because of the lack of classic GCA symptoms. Of note, 1 early instance of PAA associated with GCA involved simultaneous aortic aneurysms and PAAs requiring aortic valve replacement with ascending aortic and PA interposition grafting.[4] More recent cases demonstrated isolated distal and proximal PAAs associated with vasculitis, with both cases progressing in size, ultimately needing surgical repair.[1,5] In a review of 46 cases of PAA secondary to autoimmune disease, the underlying cause was most commonly attributed to Behçet syndrome (n = 31) and only once to GCA.[2] Our patient presented in a similar manner to the previously described cases. In patients presenting with shortness of breath, PVI, and isolated GPAA, it is important to consider GCA as a possible etiology and to be aware of the massive inflammation that may be encountered intraoperatively. Long-term data are lacking regarding repair of GPAA with aneurysmorrhaphy versus replacement. In our case, because of involvement of the entire PA trunk and vasculature, we elected to repair with aneurysmorrhaphy. Replacement should be considered for all inflammatory aneurysms of the PA. For non-GCA aneurysms, the literature supports repair of the PA when the aneurysm is greater than 5.5 cm.[3] In those with severe PVI, pulmonic valve replacement may be necessary as well. Finally, a positron emission tomography scan can be a helpful adjunctive tool in following patients with GCA GPAA.

CONCLUSIONS

Our case alerts cardiothoracic surgeons and medical practitioners to a unique presentation of GCA in which there is sole involvement of the pulmonary trunk and arteries. It is imperative to note that a GPAA and PVI can occur secondary to GCA without involvement of the aorta

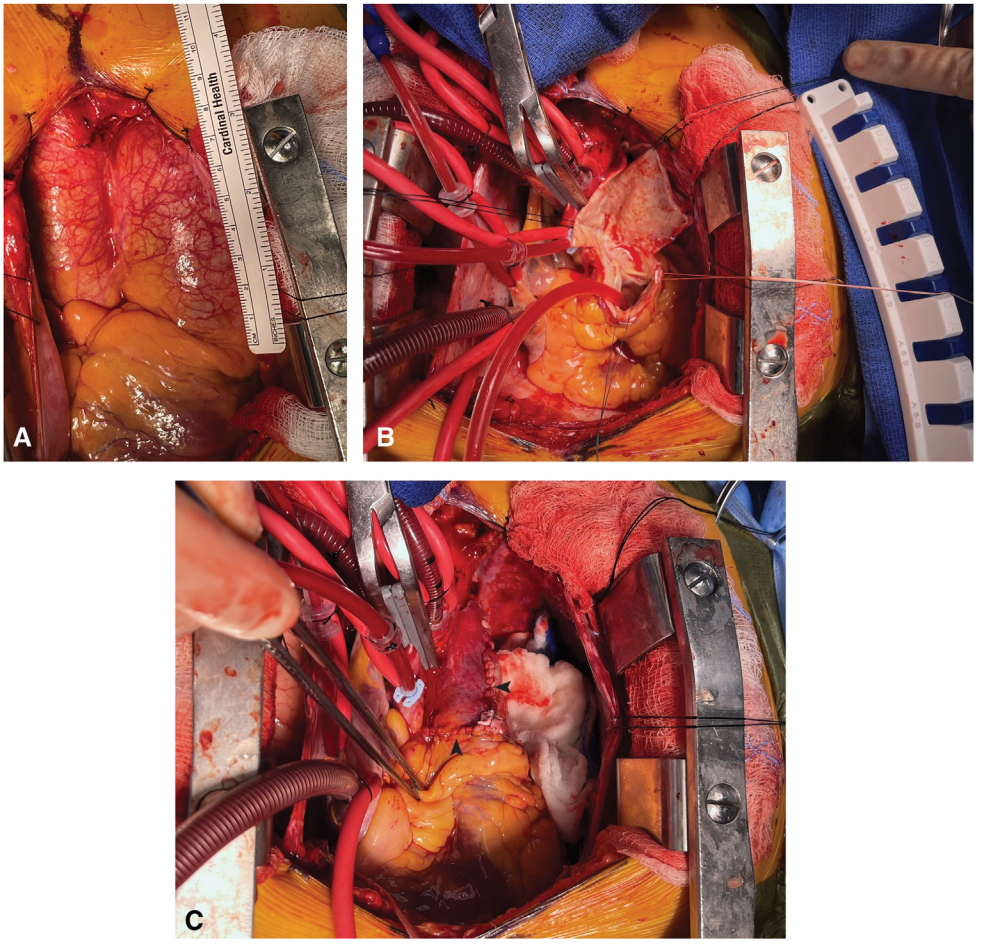

FIGURE 2. Intraoperative photographs. A, Dilated PA with inflammation of the PA and aorta. B, Pulmonic valve before replacement showing small cusps and annular dilation. C, PA aneurysmorrhaphy in an inverted T-shape (*black arrows*).

or other great vessels. Local inflammatory changes of the GPAA should be anticipated.

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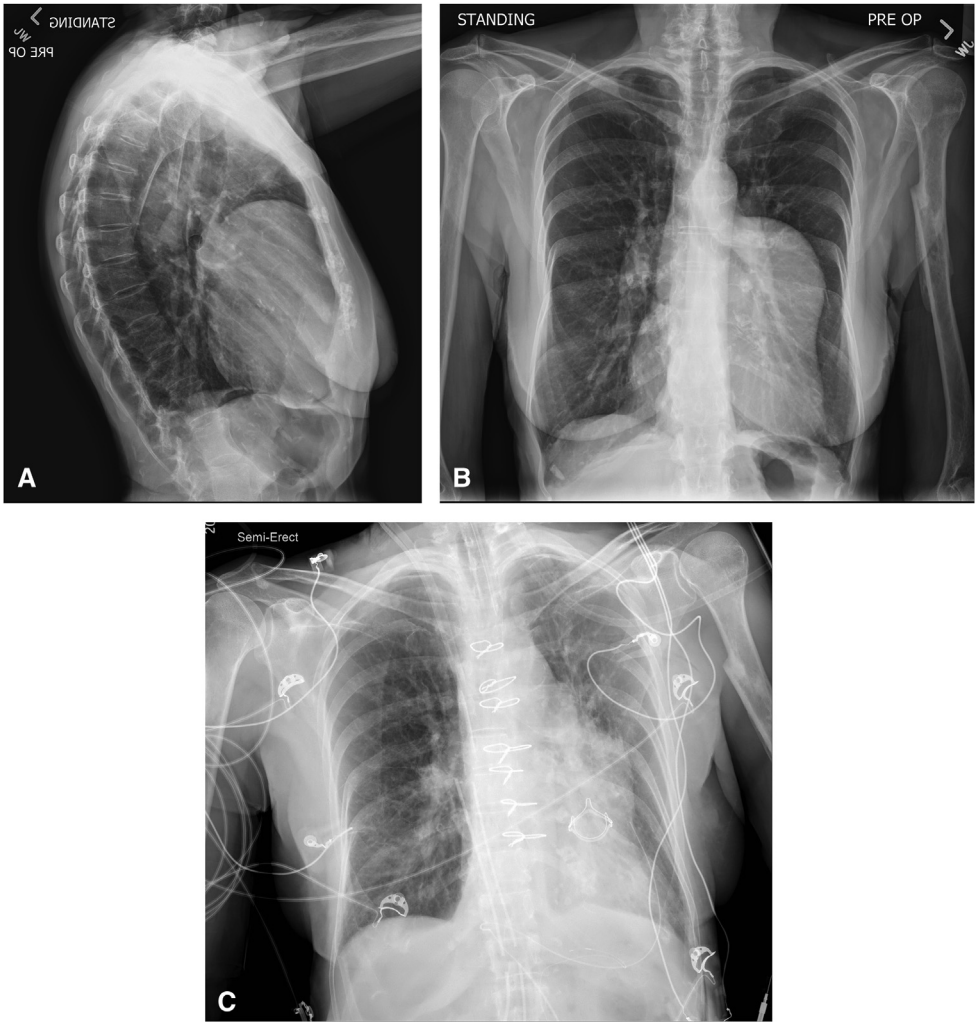

FIGURE E1. Preoperative lateral (A) and posterior-anterior (B) chest radiographs demonstrating dilation of the PA. C, Postoperative posterior-anterior chest radiograph with prosthetic valve. *PRE OP*, Preoperative.