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Letter to the Editor Open Access

Pulmonary sinus of Valsalva aneurysm: A rare and important entity

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We read the case report by Gajjar et al.[1] with a great interest. We, firstly, congratulate the authors and thank them for sharing their rare case and experiences with us. Nonetheless, we would like to make some contributions to this issue.

Pulmonary artery aneurysms (PAAs) or pulmonary sinus of Valsalva aneurysms (PSVAs) may be associated with connective tissue diseases, such as Ehlers-Danlos syndromes type VI, Marfan syndrome, Rubinstein-Taybi syndrome, Loeys-Dietz syndrome, and arterial tortuosity syndrome, Noonan syndrome, pulmonary valve stenosis or absent pulmonary valve syndrome, or the three most frequent congenital heart defects with a left-to-right shunt such as ventricular septal defects, atrial septal defects, and patent ductus arteriosus.[2-4] The aortic valve pathologies such as hypoplastic and bicuspid aortic valve disease may present concomitantly, as well.[4] In addition, PAAs or PSVAs can be associated with coronary artery anomalies and congenital keratoglobus.[2] On the other hand, acquired factors include chronic obstructive pulmonary disease, chronic pulmonary embolism, valvular heart diseases resulting in pulmonary arterial hypertension (PAH), vasculitis such as Behçet's disease, Hughes-Stovin syndrome or giant-cell arteritis, infections such as mycosis, tuberculosis, syphilis, septic pulmonary embolism, rheumatic disease such as sarcoidosis, or congenital pulmonary arteriovenous fistula or aneurysm, extravascular or intravascular post-trauma, and degenerative diseases including atherosclerosis and cystic medial degeneration.[2,4,5] Beyond this, several PAA cases without any underlying cause have been also reported in the literature. They are usually seen in the younger age group and affects both sexes equally.[4] The most common symptoms in patients with a PAA are shortness of breath, fatigue, palpitations, chest pain, syncopal episodes, cough, hemoptysis, and hoarseness. Pulmonary artery dissection or rupture, which is a fatal complication, develops in approximately one-third of cases with a PAA. Other severe complications include right cardiac failure due to pulmonary valve regurgitation, coronary ischemia secondary to left main coronary artery pressure, airway obstruction due to tracheal bronchial pressure, hoarseness due to recurring laryngeal nerve pressure, and formation of a thrombus inside the aneurysm, leading to pulmonary thromboembolism.[2] Hemoptysis is associated with a possible symptom and may be a warning sign for the presence of a ruptured aneurysm.

Dissection of a PAA is a rare, but life-threatening complication which occurs in approximately 20% of all PAA patients without PAH.[4] A structural collagen tissue disorder or infection, thinning/weakening of the pulmonary artery wall due to chronic inflammation, wall tension caused by degeneration, and chronic pressure/volume overload have been blamed for the basis of the pathophysiological mechanism of PAAs.[2] Medical treatment is recommended to improve clinical signs and symptoms of PAAs. If PAH accompanies a PAA, calcium channel blockers, anticoagulant treatment, diuretics, vasoactive substances such as endothelin receptor antagonists, phosphodiesterase type 5 inhibitors, and prostacyclin derivatives can be used.[2,4] However, it should be kept in mind that the majority of patients with normalized pressure still have a risk of increase in the PAA diameter.[4] Shunt

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flow (left-to-right) or valvular pathologies (aortic or pulmonary valve) cause persistent hemodynamic stress, leading to PAA formation and dilatation and must, therefore, be a contraindication for conservative medical treatment. Nonetheless, there is no clear consensus on the surgical treatment for a PAA. Some authors have advocated surgical repair and recommended surgical intervention, when the pulmonary artery is dissected or the pulmonary artery diameter increases progressively.[2] However, surgical treatment is required in cases of right ventricular dysfunction, severe valvular insufficiency, thrombus formation in the aneurysm sack, confirmed PAH, compression of adjacent structures, and symptomatic patients with chest pain, shortness of breath, coughing, and hemoptysis, an absolute PAA diameter of ≥5.5 cm, an increase in the diameter of the aneurysm of ≥0.5 cm within six month, and signs of rupture or dissection.[2,4] Possible alternatives for surgical treatment of a PAA include interposition with a Dacron or homograft, repair with a pericardial patch, aneurysmorrhaphy, and arterioplasty.[2] In general, patients with PAH should be seriously considered for the surgical treatment. It is considered that an aggressive surgical approach has the risk of dissection and rupture in these patients. Nevertheless, surgical therapy seems to be the only treatment with the possibility of effective long-term survival.[4] In patients with vasculitis, immunosuppressive therapy seems to be reasonable. Interventional therapy is a relatively new treatment method for PAAs, and coil embolization seems to be a good treatment option for iatrogenic causes and small branches of the pulmonary artery. Moreover, there is even a report of complete occlusion of a dissected PAA by a covered stent.[4]

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