A 79-year-old man was admitted due to hoarseness. He had received treatment for hypertension but had no history of sepsis, infective endocarditis, or chest trauma. He had no clinical features suggestive of Marfan syndrome or Ehlers-Danlos syndrome. There was no heart murmur on admission. Laryngoscopy demonstrated left recurrent laryngeal nerve palsy. Chest roentgenogram indicated an abnormal shadow on the left upper hilar region, and contrast-enhanced computed tomography (CT) showed a saccular aneurysm approximately 5 cm in largest diameter in the space between the aorta and the pulmonary trunk. 3D-CT (Figure A) demonstrated an abnormal vessel (arrow) originating from the distal aortic arch, descending between the aortic arch and connecting to the undersurface of the aneurysm. Selective angiography (Movie S1; Figure B) demonstrated an abnormal vessel with a diameter of approximately 4 mm originating from the distal aortic arch, elongated and tortuous, draining into the main pulmonary artery near the origin of the left branch pulmonary artery. The interior of the aneurysm was also stained on selective angiography (Figure B, arrow). The coronary artery was intact, and right heart catheterization indicated normal pulmonary artery pressure (17/4 mmHg) with no significant left–right shunt on oxymetry. The aortopulmonary fistula was occluded with 0.035-inch Embolization Coils (Cook Medical). Although Gianturco coils may not be suitable for transcatheter closure of moderate–large patent ductus arteriosus (PDA), we chose this coil because it was more readily available than the specific PDA coils or amplatz duct occluders and is considered to be suitable for occlusion of non-conical-shaped vascular structures, as in the present case. Follow-up CT demonstrated the absence of blood flow.

Figure. (A) Contrast-enhanced 3-D computed tomography showing an abnormal vessel (arrow) originating from the distal aortic arch, descending between the aortic arch and connecting to the undersurface of the aneurysm. (B) Selective angiography showing an abnormal vessel originating from the distal aortic arch, taking a tortuous course and draining into the main pulmonary artery (MPA). Arrow, staining of the aneurysm.
Aortopulmonary Fistula Associated With Aneurysm

into the aneurysm.

We encountered a case of thoracic aneurysm originating from an abnormal vessel that arose from the distal aortic arch and draining into the main pulmonary artery. To the best of our knowledge, such an aortopulmonary fistula has not been reported previously. Although the location of the fistula is consistent with PDA, the markedly elongated nature of the fistula raises the possibility that it is developmentally different from PDA. The ductus arteriosus undergoes a characteristic structural change immediately after birth.\textsuperscript{2,3} The medial smooth muscle fibers in the ductus arteriosus contract due to the abrupt increase in oxygen tension, which results in wall thickening, lumen obliteration, and shortening of the ductus arteriosus. Within the next 2–3 weeks, infolding of the endothelium along with subintimal disruption and proliferation result in fibrosis and a permanent seal. The resulting fibrous band persists as the ligamentum arteriosus. Thus, the wall of the PDA consists of degenerative tissues with poor elastic fibers, and the fragility of the ductal wall following structural change in an incomplete closing process is considered to be a potential pathogenesis of ductal aneurysm formation.\textsuperscript{4} Aneurysms of PDA are most commonly present in infancy associated with an underlying disorder such as trisomy 21, trisomy 13, Smith-Lemli-Opitz syndrome, type IV Ehlers-Danlos syndrome, or Marfan syndrome.\textsuperscript{5} To the best of our knowledge, there have been only two adult cases of aneurysms of PDA without an underlying disorder.\textsuperscript{6,7} In contrast to the present case, however, those PDA were associated with a significant left–right shunt, and the aneurysms arose from the proximal portion of the PDA, which had a short communication with the distal aortic arch and pulmonary artery. Thus, the aortopulmonary fistula in the present case appears to be structurally and functionally different from PDA. Its markedly elongated and tortuous course suggests that normal vascular components remained in the vessel wall after birth, and the vessel underwent persistent growth. An aneurysm may arise from the vessel wall associated with atherosclerotic change due to aging.

References


Supplemental Files

Supplemental File 1

Movie S1. Selective angiography demonstrating an abnormal vessel originating from the distal aortic arch, taking a tortuous course, carrying blood into the aneurysm and draining into the main pulmonary artery near the origin of the left branch pulmonary artery.

Please find supplemental file(s);