RARE PRESENTATION OF DUCTUS ARTERIOSUS ANEURYSM COARCTATION OF AORTA AND ORTNER'S SYNDROME

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Abstract:
Ductus arteriosus aneurysm (DAA) is a rare congenital anomaly diagnosed within the first two months of infancy usually and less frequently in the third trimester antenatally. It accounts for about 5% of all thoracic aneurysms. When symptomatic, they often mimic other nonvascular conditions, presenting with hoarseness, chest pain, recurrent nerve palsy, or recurrent pulmonary infections. Here we present a rare manifestation of ductal diverticulum aneurysm compressing the descending thoracic aorta externally which lead to coarctation of aorta with significant pressure gradient between pre and post coarctation segment and hoarseness of voice.

Keyword: ductus arteriosus aneurysm, Coarctation, Ornters syndrome.

INTRODUCTION:
Ductus arteriosus (DA) is a normal anatomic structure that provides communication between the systemic and pulmonary circulations during fetal life and closes soon after birth. The pathogenesis of ductal aneurysm in adults still remains uncertain. Incomplete obliteration of the DA at the aortic arch end has been considered as a primary abnormality for aneurysm formation. Closure of the DA normally proceeds from the pulmonary arterial side toward the aortic side. An indentation of the aortic wall at the site of insertion of the obliterated ductus arteriosus is seen in approximately 9–26% of adults on angiography studies and is referred to as a ductus diverticulum or bump.[1] However, if closure of the aortic end is delayed, DA becomes an aortic diverticulum and is exposed to aortic pressure and can thus develop into an aneurysm. In addition, the structural change of the ductal wall should be considered as a cooperative factor for aneurysm formation. In previous studies the wall of the ductal aneurysm contained poor elastic fibers in comparison to the aorta or the pulmonary artery, and the media was degenerated by a mucoid substance. Herein we report a case of ductal aneurysm causing extrinsic compression of the post subclavian aorta and compression of recurrent laryngeal
nerve resulting in ortner’s syndrome.

**CASE REPORT**
A 26 year old female presented with complaints of bilateral leg claudication pain on walking for 100 meters and hoarseness of voice of 2 months duration. There was no major significant history in the past and no history cardiac diseases in the childhood. On examination, her BP was 150/100 mm hg in both upper limbs and 120/98 mm hg in both lower limbs. Her lower limb femoral pulse was weak and there was significant radio – femoral delay present. There was no evidence of lower limb muscle wasting or other signs of arterial insufficiency. Cardiovascular examination was unremarkable. Transthoracic echocardiogram revealed an out pouching globular mass from the isthmus of descending aorta with free flow of blood from aorta to the outpouch. Turbulent flow was seen at the isthmus site and continuous Doppler revealed systolic gradient of 81.9 mm Hg with diastolic spilling of 15 mm Hg (fig 1, 2, 3) and bicuspid aortic valve. To study the extent and the origin of the mass, a cardiac catheterization study was performed through femoral artery. Pigtail catheter size 5 f entered into the globular outpouch from isthmus part of aorta and could not be negotiated to enter the arch of aorta. Pressure in the descending aorta was measured to be 100/80/70. Aortogram done at this level revealed the globular outpouching from the isthmus suggestive of aortic ductal diverticulum (Fig 4 & 5). The Diverticular pressure recorded was 90/80/70. The arch and ascending aorta were entered with difficulty after exchanging the pigtail catheter with a multipurpose catheter. Then Multipurpose catheter was exchanged with pigtail catheter and aortic root angiogram was done which showed the diverticulum to be originating from the isthmus of aorta. The pressures in the ascending aorta and Left ventricle were 160/120/90 and 150/15 respectively.
**DISCUSSION:**
Aneurysm of the ductus arteriosus may occur either spontaneously or may follow surgical treatment of a patent ductus arteriosus.[2] Spontaneous aneurysm of the ductus arteriosus is an uncommon occurrence, with only 34 reported cases in the Japanese literature till 2009.[3] Spontaneous aneurysm of the ductus arteriosus presenting in adults usually shows an obliterated pulmonary end of the ductus, unlike aneurysms in the pediatric age-group which occur in an open ductus arteriosus.[4] The presence of concomitant hypertension can be a contributory factor. Connective tissue disorders such as Marfan syndrome and Ehlers-Danlos syndrome are known to predispose to ductus arteriosus aneurysms as well.[5,6] Hoarseness of voice, cough, anorexia, and chest pain are common presenting symptoms in adults and may be secondary to involvement of the adjacent organs and nerves. Hoarseness of voice occurs due to compression of the recurrent laryngeal nerve as it courses through the aortopulmonary window and our patient had the symptom. Fetterolf and Norris[8] were the first to show that the recurrent laryngeal nerve must be compressed in the aortopulmonary window between the left pulmonary artery, the aortic arch, and the ligamentum arteriosum to produce clinical symptoms of left recurrent laryngeal nerve palsy. Spontaneous aneurysm of the ductus arteriosus is an uncommon but important cause of left recurrent laryngeal nerve palsy in adults. In our case, the hoarseness of voice had a delayed onset, despite the long-standing presence of the ductus aneurysm. And also our patient had features of lower limb claudication due to coarctation of aorta because of external compression. Rupture of the aneurysm is reported to be the commonest complication in adults.[2] Erosion into adjacent mediastinal structures (pericardium, bronchi, and esophagus), endocarditis, and thrombosis have also been reported.[2,4]

**CONCLUSION**
Spontaneous aneurysm of the ductus arteriosus is an extremely rare condition. They can present with features of non-vascular conditions like recurrent laryngeal nerve palsy, chest pain or recurrent pulmonary infections. Aneurysm of the ductus arteriosus causing extrinsic compression of the post subclavian aorta and presentation as Coarctation of aorta is exceedingly rare. To the best of our knowledge this is the first report of a case of Spontaneous aneurysm of the ductus arteriosus presenting as Coarctation of aorta and ortner’s syndrome. This case is presented for the rarity of its presentation.

**REFERENCES:**


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