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Review Article

Idiopathic dilatation of pulmonary artery: A review

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ABSTRACT

The diagnosis of idiopathic dilatation of pulmonary artery is challenging because its clinical recognition is difficult and various other causes of dilated pulmonary artery need to be excluded. The clinical findings mimic various common cardiac disorders and both invasive and non-invasive investigations should be done to arrive at the diagnosis. It is a known clinical entity but etiology and pathophysiology are largely unknown. The current echocardiographic and catheterization based diagnostic criteria, may not be satisfied equally in a particular patient and need to be revisited in view of newer imaging modalities. There is paucity of information about the natural history of the disease with attendant lack of clarity in treatment guidelines. Certain cases may progress to huge dilatation and consequent serious implications. It is a rare disease and is the diagnosis of exclusion.

1. Introduction

Idiopathic dilatation of the pulmonary artery (IDPA) was first reported by Wessler and Jaches.[1] The clinical relevance and challenges in the diagnosis of idiopathic dilatation of pulmonary artery lie in exclusion of various other causes of dilated pulmonary artery. Clinical diagnosis is challenging because the symptoms are highly variable and clinical findings may mimic common cardiac disorders like mild pulmonary stenosis, atrial septal defect and pulmonary artery hypertension. The purpose of this article is to emphasize on varied clinical presentations, the need for an inclusive and comprehensive evaluation of patients, given the fact that clinical and investigative findings may be not coherent and to discuss the treatment options in light of natural history of the disease.

2. Incidence

Idiopathic dilatation of pulmonary artery is an uncommon anomaly occurring in 0.6% (in isolation) of patients with congenital heart disease.[2] In a series of autopsy cases of 109,571 cases Deterling and Clagett reported an incidence of 0.0073%.[3] The exact incidence is underreported because of very benign nature of disease in most of the cases. In the past the diagnosis was based on cardiac catheterization/angiocardiogram or autopsy, but with the advancement in echocardiography, cardiac computed tomography or cardiac magnetic resonance (CMR), more number of cases are being reported.

3. Etiopathology

Not much is known about the etiology of idiopathic dilatation of pulmonary artery. Most of the cases have been reported in adults, but there is description of this disease in children which supports the theory of congenital origin.[4] Preponderance toward any sex is not described. The main pulmonary artery and the origin of the right and left main pulmonary arteries are mainly affected. Assman[5] postulated theory of an unequal division of truncus arteriosus communis as the possible mechanism which was supported by Kourilsky et al.,[6] Laubry et al.[7] and Gold[8] while Greene et al.,[9] did not agree with this theory. Laubry and Gold suggested the association of hypoplastic aorta with dilated pulmonary artery – *grosse pulmonaire – petite aorte*. According to Kaplan[10] this anomaly was due to maldevelopment of entire pulmonary tree. Carlotti et al.[11] demonstrated that there was no parallelism between the dilatation of pulmonary artery in and the pressure or cardiac output in contrast to earlier theories relating dilatation of pulmonary artery, cardiac output and pulmonary blood flow in cases of IDPA. Congenital weakness in the wall of the pulmonary artery is the most accepted theory

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keeping in with the concept of medial deficiency and aneurysm formation.[12,13] Deb et al. also suggested cystic medial degeneration of arterial wall as an etiological factor in idiopathic dilatation of pulmonary artery.[14] An association between cystic medial degeneration and increased hemodynamic forces leading to aneurysm formation is postulated, even in the absence of a bona fide connective tissue disorder.[13] Pulmonary artery aneurysms have been associated with structural cardiac anomalies, structural vascular anomalies, vasculities and infection. Idiopathic term should be applied when all these have been excluded conclusively.

4. Pathophysiology

The disease process usually involves the central pulmonary artery, however it may extend to any of the main branches. A few studies with histological evaluation reported evidence of cystic medial deficiency in such cases. With the constant blood flow across the heart and great vessels, pressure varies directly with resistance to flow (resistance = pressure/flow). As the dilatation becomes more, the resistance to blood flow in dilated pulmonary artery is decreased and as well proportional drop in pressure, which accounts for the pressure gradient as the driving force between right ventricle and the pulmonary artery during cardiac systole without right ventricular hypertension. At the same time, progressive and hugely dilated pulmonary artery results in reverse systolic flow due to increased capacitance. This reverse systolic flow persists in early diastole.

5. Clinical picture

Absence or mildness of symptoms was the most significant finding in all previous studies. Patients may present with exertional dyspnea, palpitation, fatigue and chest pain. Symptoms usually appear with onset of complications like compression of nearby structures, dissection, thrombosis or rupture. Zhao et al. reported 21 cases of IDPA. Majority of patients were asymptomatic (66.67%) and all patients had normal respiratory and cardiovascular function. It was misdiagnosed (clinically) as pulmonary stenosis (42.86%), secundum atrial septal defect (38.50%) and pulmonary artery hypertension (28.57%).[15] Around 10% patients had a duration of illness lasting for more than 20 years. IDPA has been ascribed to as one of the rare cause of angina pectoris which can be missed at times.[16] The idiopathic dilatation of the pulmonary artery may be extensive (aneurysm) and patients have more symptoms, especially breathlessness. Taussig[17] described a case of severe form of idiopathic dilatation of the pulmonary artery where postural changes in hugely dilated pulmonary artery constricted the trachea and its main branches causing attacks of cyanosis with severe dyspnea. Dilated pulmonary arteries (from any cause) can lead to compression of left main coronary artery[18] and even sudden cardiac death has been described in cases of IDPA.[19]

Clinical examination is not conclusive because IDPA can mimic findings of common cardiac disorders. The second heart sound may be normal or accentuated but splitting is usually fixed. In cases of hugely dilated pulmonary artery/aneurysm the second heart sound may be soft. The pulsations of dilated arteries may be felt in second space on the lateral sternal border. A pulmonic ejection systolic murmur is usually present. The systolic murmur in cases of idiopathic dilatation of pulmonary artery does not show typical transmittance to subclavicular or interscapular area as compared to pulmonary stenosis with dilatation pulmonary artery. The systolic murmur was thought to be originating from dilatation of pulmonary ostium and greater stress in a dilated pulmonary artery.[8,20] Kaplan et al.[10] emphasized that the only constant feature of murmur is its inconstancy. Sometimes a diastolic murmur can also be present.[7] The systolic ejection click may be present as in other conditions with dilatation of great vessels. The typical phonocardiographic findings were described by Karnegis et al.,[21] Pulmonary insufficiency of varying severity is observed in majority of cases although consistently not present in all of the patients.

6. Investigations

6.1. ECG

It is usually normal but deviation of the cardiac axis toward the right or bundle branch blocks can be present.

6.2. Chest X-ray

In idiopathic dilatation of pulmonary artery, lung parenchyma and its vascularity are normal with normal cardiothoracic ratio and dilated pulmonary artery. The characteristic picture of pulmonary stenosis e.g. enlarged main pulmonary artery with diminished vascularity is absent in this condition (Fig. 1).[22] The hyperemic lungs fields with hilar dance, common in large left to right shunts, are also absent. It is interesting that the pattern of dilatation is the same as in uncomplicated pulmonary stenosis (localization is restricted to the trunk or occasionally to one of the main branches of the pulmonary artery, but no dilatation of the peripheral branches is present).

6.3. Echocardiography

Although idiopathic dilatation of pulmonary artery is mainly the diagnosis of exclusion, authors have given criteria for diagnosing idiopathic dilatation of pulmonary artery. In 1949, Greene[9] proposed the criteria for diagnosis of idiopathic dilatation of pulmonary artery (Table 1, Fig. 2). Desmukh[23] proposed fifth criteria of normal pressure in the right ventricle and pulmonary artery.

The other echocardiographic based diagnostic criteria were described by Boutin et al.[24] in 1994. It was based on study of 30 patients in which 17 had characteristic radiological findings and 13 others had clinical signs of idiopathic dilatation of the pulmonary artery, compared with a group of 20 normal control subjects. Comparison with the control group showed significant differences ($p < 0.05$) in 4 parameters:

1) The diameter of the pulmonary artery at the bifurcation/m^2 body surface area (2.8 ± 0.4 cm versus 2.4 ± 0.4 cm)
2) The diameter of the aorta 2 cm beyond the aortic valve/m^2 of body surface area (1.7 ± 0.3 cm versus 2.1 ± 0.7 cm)
3) The ratio of pulmonary artery/aortic diameters at the valve rings (1.4 ± 0.2 versus 1.1 ± 0.02)
4) The ratio of the aorta 2 cm beyond the valve/aortic ring (1.02 ± 0.07 versus 1.09 ± 0.09).

Van Buchem et al.[25] reported that the normal diameter of pulmonary artery ranged from 22 to 33 mm. These three sets diagnostic criteria may not be satisfied together in a particular patient owing to different parameters used by different authors. Asayama et al.[26] described fine systolic fluttering of pulmonary valve in IDPA. Echocardiography is also useful in assessing parameters which help in making treatment decisions such as right ventricular function, pulmonary regurgitation and hemodynamic parameters in excluding other causes of dilated pulmonary artery.

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