QUADRICUSPID PULMONARY VALVE - A RARE CASE REPORT

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Abstract:
Quadricuspid pulmonary valve is a rare congenital cardiac malformation which is notable for its asymptomatic behaviour. With only a few hundred cases reported in literature, its clinical significance is not exactly known, although many authors have placed its implications on the Ross procedure. We report a case of quadricuspid pulmonary valve which was detected on routine cardiac screening.

Keyword: quadricuspid pulmonary valve, Ross procedure

An asymptomatic 13 year old boy was referred from school health screening for further evaluation of an incidentally detected cardiac murmur. There was no history of pre existing cardiac disease. On examination, the vitals were found to be normal. The results of clinical and cardiac evaluation are summarized in the following table.

Cardiac CT angio and Cardiac MRI could not be done as the patient was not affordable to do them.

Table 1: Clinical and Cardiac evaluation.

<table>
<thead>
<tr>
<th>Evaluation</th>
<th>Findings</th>
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<tbody>
<tr>
<td>Cardiac auscultation</td>
<td>Ejection systolic murmur grade 2/6 in the pulmonary area</td>
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<tr>
<td>Electrocardiogram</td>
<td>Incomplete right bundle branch block</td>
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<tr>
<td>Chest radiograph</td>
<td>Normal</td>
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<tr>
<td>Echocardiogram</td>
<td>1. Quadricuspid pulmonary valve with mild valvular pulmonary stenosis (pressure gradient of 23 mm Hg)</td>
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<td>2. Trivial pulmonary regurgitation</td>
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<td></td>
<td>3. Dilatation of pulmonary trunk</td>
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<td></td>
<td>4. Intact interatrial septum</td>
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<td>5. Normal biventricular function</td>
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Discussion:
Quadricuspid Pulmonary Valve (QPV) is a rare congenital anomaly accounting for approximately 0.005-0.05% of all congenital heart diseases in major autopsy series. It is usually detected incidentally during a routine cardiac testing and is not associated with any additional anomalies, although there are some reports of coexisting aortic valvular abnormalities. Presently, the number of published cases of QPV is less
than 400. The overall incidence is quoted to be at 1 in 400 to 1 in 20000 autopsies (1). The absence of significant symptoms and its strategic position in the thoracic cage makes it difficult to identify the abnormality during life and most case series report only an autopsy incidence, thus underestimating the actual incidence of QPV in general population. There is frequent requirement of specialized techniques like cardiac catheterization and computerized tomography for the diagnosis of QPV.

There are several case reports of QPV with additional cardiac abnormalities. The earliest published case series of QPV was by Enoch et al who reported four cases of isolated QPV and one case of QPV with rheumatic mitral stenosis (1). Two of these patients had pulmonary regurgitation. Kissin et al reported a case of QPV with pulmonary regurgitation as early as 1936 and later noted that the most frequent pattern of the abnormal valve was that of interposition of a rudimentary accessory cusp in between three normal cusps. The other less common pattern was a combination of two larger and two smaller cusps (2). The large case series by Davia et al reported 47 cases of quadricuspid semilunar valves, of which 35 were QPV and the rest were quadricuspid aortic valves (QAV), indicating a clearly higher incidence of QPV compared to QAV. In this series, only ten out of 35 patients were symptomatic due to the presence of additional congenital abnormalities (3).

Reyes et al reported a case of congenital QPV with double valvular lesions and post stenotic dilatation of left pulmonary artery (4). Chehab et al reported a case of QPV complicated by aneurysm of the pulmonary trunk (5). Hwang et al reported two cases of quadricuspid semilunar valves, one of which was a symptomatic QAV and the other was an asymptomatic QPV, which was incidentally detected on autopsy (6). Ricci et al reported a case of QPV with transposition of great arteries (7). The present case of QPV was an isolated anomaly of the pulmonary valve where the patient was asymptomatic and the abnormality was picked up on a routine echocardiogram.

Although several authors have reported that the usual long term consequence of QPV is regurgitation, several others have argued that the condition has no clinical implication. The most important clinical implication of QPV is during the Ross procedure and the importance of the diagnosis of a QPV has been stressed by several authors. Present evidence suggests that the QPV has unfavourable hemodynamics which makes it unacceptable for a Ross procedure. The ability of QPV to maintain normal hemodynamics is restricted to the pulmonary circulation and this can lead to serious consequences in the systemic circulation. It has thus been stressed that the presence of a QPV should be diagnosed before planning a Ross procedure to avoid untoward hemodynamic consequences (8).

Conclusion
Quadricuspid pulmonary valve is a rare, most often isolated congenital cardiac abnormality which has minimal, if any, clinical consequences. Its importance arises when the Ross operation is being contemplated, making it mandatory for early detection.

References:
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