Clinical Case Report

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Investigation of an anatomically variant isolated bicuspid pulmonary valve

A case report

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Abstract

Introduction: We provide a discussion of the anatomical characteristics of the bicuspid pulmonary valve (BPV) in this paper. We performed an autopsy of an isolated BPV found in the heart of a deceased individual. The deceased was a man in his 60s and had no previous history of cardiovascular disease. The heart weighed 260 g and had mild right ventricular hypertrophy. The pulmonary valve had a fish-mouth-like shape that was convex to the pulmonary trunk and both cusps were thickened and hardened. The anterior and left semilunar cusps of the pulmonary valve were fused. Post-stenotic dilatation was noted.

Conclusions : In comparing the present case with previous reports, we found that, in human BPVs, cusps are fused in at least 2 patterns.

Abbreviations: AV = aortic valve, BPV = bicuspid pulmonary valve, PV = pulmonary valve.

Keywords: autopsy, cardiac anomaly, isolated bicuspid pulmonary valve

1. Introduction

The presence of a bicuspid pulmonary valve (BPV) without other cardiac malformations (i.e., isolated BPV) is rare and generally of little clinical significance.[1,2] Therefore, reviews of isolated BPV cases are sparse. For example, in 1999, Gerlis published a detailed report about BPV,[1] and Ford et al[3] reviewed 15 previously reported cases of isolated BPV, including their own case, in 1956. Both reports concluded that isolated BPV is generally a benign entity. One mechanistic study by Fernández et al[4] included a detailed examination of the anatomy and formation of congenital BPVs in Syrian hamsters.

In this study, we described the autopsy results of a man in his 60s who had an isolated BPV. The sample had an anatomically unique characteristic compared with the observations of Fernández et al[4]. There seems to be a difference between the formation of a BPV in humans and Syrian hamsters. We further discussed the anatomic findings of this case of isolated BPV.

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2. Methods

2.1. Case History

An alcoholic man in his 60s was found dead in his friend's garden. No resuscitation was attempted. An autopsy was performed 2 days later. His medical history included a complaint of epigastric pain approximately 10 years prior and right chest pain and respiratory discomfort approximately 1 year prior. He had no previous history of cardiovascular disease, but had undergone craniotomy for acute subdural hematoma after he suffered a fall 3 years prior. Informed consent for the publication of this report was obtained from the patient's younger brother, who was the only living relative.

3. Results

The length and weight of the body were 160 cm and 51 kg, respectively. The palpebral and bulbar conjunctivae were jaundiced, and pitting edema was observed on the dorsal surfaces of both feet. The heart weighed 260 g. The aortic valve (AV) and pulmonic valve (PV) roots measured 7.2 and 10.2 cm in circumference, respectively, and the pulmonary trunk exhibited dilatation. Although the AV was normal, the PV was bicuspid with a fish-mouth-like shape that was convex to the pulmonary trunk (Fig. 1). Both cusps were thickened and hardened. The anterior and left semilunar cusps of the pulmonary valve were fused, and a hypoplastic commissura was observed at the center of the fused valvular cusp (Fig. 2). A raphe-like ridge was observed in the left sinus (Fig. 3). No vegetation or calcification was observed on the pulmonary valve. The left and right ventricular wall and interventricular septum thickness were 1.0, 0.6, and 1.0 cm, respectively. The columnae carneae and papillary muscles of the right ventricle were enlarged. The foramen ovale was closed, and no ventricular septal defect or subvalvular obstruction of the right outflow tract was observed.

The coronary artery exhibited severe calcification, with no other anomalies. The left and right anterior descending arteries

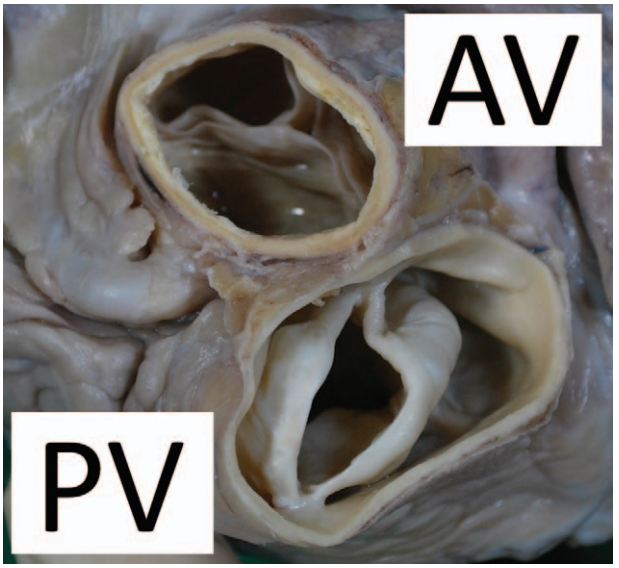

Figure 1. Aortic (AV) and pulmonary (PV) valves. The AV is normal, whereas the PV is bicuspid and forms a fish-mouth-like shape that is convex to the pulmonary trunk. Both cusps are thickened and hardened. Alignment of the figure: right is left, bottom is anterior.

were found to be approximately 50% and 40% stenosed, respectively, and slight stenosis was observed in the left circumflex branch. No thrombus was observed in the coronary artery. The right and left lungs weighed 580 and 840 g, respectively, and exhibited congestion and edema. The liver was hard, nodular, atrophic, and weighed 1200 g. The patient had accumulated 2100 mL of ascites. The pancreas weighed 50 g, and was hardened and fibrous. No other intracranial lesions were found other than a craniotomy scar on the right parietal bone and old cerebral contusions at the base of the left frontal and temporal lobes.

A histological examination revealed fibrotic thickening of the pulmonary valve. Inflammatory cell infiltration, hemorrhage, calcification, and tumorous lesions were not seen in the pulmonary valve. However, a large scar was observed at the posterior part of the interventricular septum. In addition, scattered and small fibrotic foci, broad edema, and contraction band necrosis were observed in the right ventricle. In the right ventricle, hemorrhage, neutrophilic infiltration of the interstitium, and neutrophilic

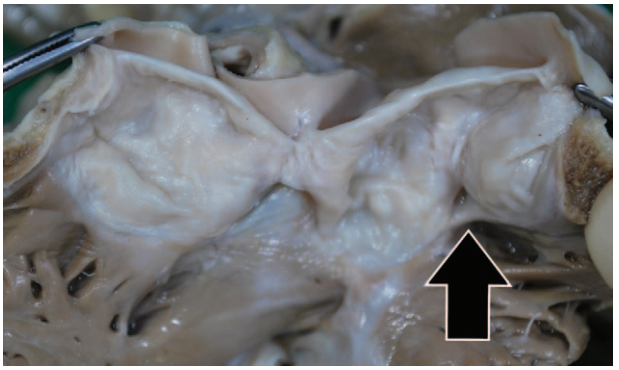

Figure 2. The pulmonary valve as seen from the right ventricle after incision. The anterior and left semilunar cusps of the pulmonary valve are fused and a hypoplastic commissura is observed at the center of the fused valvular cusp (arrow).

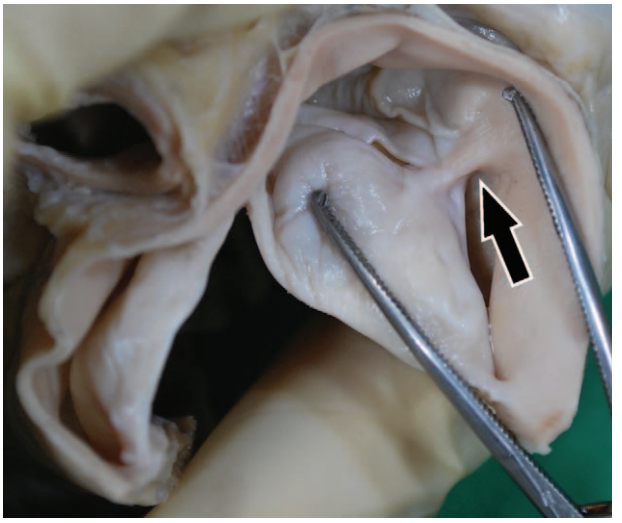

Figure 3. The left sinus of the pulmonary valve. A raphe-like ridge was observed (arrow).

accumulation in the blood vessels were detected locally. Moderate sinoatrial and atrioventricular node fibrosis was also observed. The liver exhibited massive fibrotic tissue replacement and pseudolobule formation.

According to a toxicology report, ethanol was not detected in the patient's blood. Similarly, a Triage test (Alere, Inc., Sawyer, MA) yielded no positive results. We determined the cause of death to be liver failure due to liver cirrhosis.

4. Discussion

BPV is usually observed in patients with other congenital heart diseases, particularly tetralogy of Fallot. In contrast, isolated BPV is rare, with a prevalence of approximately 1 in 3000 autopsy cases.[1] As the clinical course of a BPV appears to depend on the associated anomalies, a BPV itself is thought to have almost no clinical significance.[1,2]

Fernández et al[4] investigated BPVs in 206 Syrian hamsters belonging to a single laboratory-inbred family and discussed the mechanism of the development of this anatomical abnormality. In doing so, they found that 45 hamsters had BPVs, of which all exhibited fused right and left cusps. Consequently, ventral and dorsal sinuses were formed. From these observations, Fernández et al[4] concluded that a BPV occurs due to fusion of the right and left cusps. Gerlis[1] reported a case of isolated BPV in accordance with this theory (i.e., 2 cusps were on the anterior and posterior [ventral and dorsal in animal] side and the pulmonary valve was opened in the right to left directions). On the other hand, in our case, 2 cusps were on the right and left side, and the pulmonary valve was opened in an anterior to posterior (ventral and dorsal in animal) direction since the anterior and left cusps were fused. The isolated BPV in Gerlis's report was similar to our fish mouth-like cusps in shape. Thus, the BPV reported herein is an approximately 90° rotated version of the BPV reported by Gerlis. Furthermore, Mahato[5] reported a case of isolated BPV in which the direction was the same as that in our case (i.e., cusps were right and left and the opening was ventral to dorsal), but without thickened and hardened cusps. It is not clear which anatomical features of human BPV are the most common since there are few reports; however, the mechanism by which BPV forms in humans is different than that of Syrian hamsters in some cases.

In the present case, both pulmonary cusps were thickened, which is a frequently observed feature in cases of valvular pulmonary stenosis.[6,7] Furthermore, the fish-mouth-like shape of the valve was also thought to reduce the size of the pulmonary valve orifice. These findings suggest that pulmonary stenosis occurred in this case, which was supported by observations of right ventricular hypertrophy and pulmonary trunk dilatation in the deceased's heart. However, right ventricular hypertrophy in not severe and decompensated cirrhosis was readily observed. Thus, we determined the cause of death to be liver failure due to liver cirrhosis. Patients with advanced chronic liver disease sometimes exhibit hyperdynamic syndrome and increased cardiac output.[8] Pulmonary valve stenosis may lower right heart cardiac output and cardiac reserve, which may adversely affect the patient's general condition; however, it is unlikely that isolated BPV in this patient was the cause of death.

We conclude that there are a variety of mechanisms by which BPV forms in humans, which are different than those of Syrian hamsters, as reported by Fernández et al.[4]

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