A Supernumerary Valvula in the Pulmonary Semilunar Valve

By

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Summary: A supernumerary valvula of the pulmonary semilunar valve was found in a 51-year old, male Japanese. The valve was composed of three equal-sized valvulae and one smaller one with numerous fenestrations present. The supernumerary valvula was located between the anterior and right valvulae. We found this condition in one of 1407 cadavers examined.

Introduction

A supernumerary or quadricuspid pulmonary valve has been considered to be a rare congenital anomaly (Simonds, 1923). However, de Vries (1918), Becker (1972) found that a fourth valvula or cusp in this normally tricuspid valve occurred frequently. In his review of the literature Kissin (1936) reported that the anomaly was usually associated with other clinical symptoms. The purpose of this paper is to report a case of supernumerary valvula found in the pulmonary semilunar valve in a Japanese.

Clinical History

A supernumerary valvula was found in the pulmonary valve of a fifty-one year old Japanese male in the dissecting laboratory at Nihon University School of Dentistry. The patient died in a hospital in September, 1975 following acute, cardiac insufficiency. From the hospital medical records he had several other diseases including cerebral infarction, chronic hepatitis, high blood pressure, lumbar back pain, psychasthenia and organic dementia before he died. However, there were no physical signs of pulmonary insufficiency. There was no electrocardiographic information on the status of the heart.

Gross Anatomical Findings

An extra valvula of the pulmonary semilunar valve was discovered in one of 1407 cadavers dissected between 1952 and 1987. It was located between the anterior and right valvulae and was smaller than the other three valvulae (Figs. 1, 2). Its free edge measured 1.0 cm in length and the nodule was 0.8 cm from the lowest part of the valvula. The remaining three valvulae were almost equal in size (2.1-2.2 cm in length and 1.3 cm in depth). A normal appearing nodule was located at the center of each valvula. The sinus of the
anterior and left valvulae was larger than that of the right.

The supernumerary and right valvulae exhibited fenestrations just below their edge and a fibrous band bridged each open area (Figs. 1B, 2). Two fenestrations were present in each valvula and an additional one was in their commissure region. Those in the supernumerary valvula were located closer to the commissure on each side than to the nodule. The largest one was in the right valvula near the commissure region with the supernumerary valvula. Because of the size and number of fenestrations in that region the two valvulae were essentially anchored in the wall of the pulmonary trunk by way of fibrous bands.

The heart weighed approximately 310 grams. Both coronary arteries were present and were not markedly sclerotic or calcified. The tricuspid, mitral (bicuspid) and aortic semilunar valves had the usual number of valvulae or cusps and presented smooth surfaces. The myocardium appeared normal to the naked eye. The size of the major vessels was within normal limits as they entered or left the heart and no abnormal morphology was apparent. Several pathological changes were found in organs surrounding the heart. The lungs were slightly congested but there were no pleural adhesions. Two small hemorrhagic areas were found along the lesser curvature of the stomach. The spleen was hypertrophic but exhibited no other gross pathological changes. A small infarct was evident in the liver. In the cerebral hemispheres there were several small, soften, cyst-like areas. This was especially evident in the occipital lobe. No pathological change was observed in the pancreas, small and large intestines and urogenital organs.

Fig. 1. (A) Quadricuspid pulmonary valve viewed from above
1: anterior (left anterior) valvula
2: supernumerary valvula
3: right (right anterior) valvula
4: left (posterior) valvula

(B) The pulmonary trunk has been opened with a cut between the anterior and left valvulae. The small supernumerary valvula (2) is indicated by a white arrowhead.
Discussion

Reports on the incidence of a supernumerary valvula (quadricuspid) in the pulmonary valve have varied between 1/250 (Becker, 1972) and 1/2,500 (Houck, 1929) necropsies. We found one such valve in 1407 cadavers dissected between 1952 and 1987. A pulmonary valve with four valvulae should not be considered a rare congenital anomaly according to Becker (1972). However, semilunar valves with four valvulae appear to be much less common than those with either one or two (Hurwitz & Roberts, 1973). Davia et al. (1977) found a 5:1 ratio of quadricuspid pulmonary valves to quadricuspid aortic valves. Approximately 65% of reported cases of quadricuspid pulmonary valves were found in males which was the sex in the present case (Kissin, 1936; Hurwitz & Roberts, 1973).

No other cardiac anomalies were observed in the present case but Enoch (1968) indicated that 20% (1/5) of the hearts in his cases had another abnormality. Davia et al. (1977) reported that 29% of their cases (10/35) exhibited clinical and pathological evidence of coexisting congenital cardiac defects. However, in the remaining 71% of their cases the quadricuspid pulmonary valve was an incident finding at autopsy. In their study, Hurwitz & Roberts (1973) found that a pulmonary semilunar valve with four valvulae was rarely associated with other congenital anomalies.

Symptoms or signs of pulmonary insufficiency may be present in patients with a quadricuspid pulmonary valve but this is not usually the case. Kissin (1936) claimed to have clear cut clinical evidence of pulmonary insufficiency in 12.5% (3/34) of his patients. Enoch (1968) found morbid anatomical evidence of pulmonary regurgitation in 40% (2/5) of his patients. In contrast, none of Becker's (1972) 20 cases presented evidence of pulmonary insufficiency during life. He believed that this condition has no or negligible clinical significance and gave that as the reason it is easily overlooked at necropsy. There was no convincing evidence that pulmonary insufficiency existed in the present case.

The quadricuspid valve in the present case was made up of three large valvulae and one small one (Fig. 2). This is the usual arrangement according to Kissin (1936), Hurwitz & Roberts (1973) and Davia et al. (1977). The extra valvulae in our specimen was located between the anterior and right valvulae (Fig. 1) but its site can vary according to Kissin (1936). Two fenestrations were located in both the supernumerary and in the right valvulae and an additional one was located in the intervening commissure region (Figs. 1B, 2). The supernumerary cusp is often fenestrated according to Kissin (1936). Hurwitz & Roberts (1973) commonly observed multiple small fenestrations in the eight specimens they found. Such valvular fenestrations appear to represent tissue deficits that likely have a congenital origin.

References