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QUADRICUSPID AORTIC VALVE – A RARE CONGENITAL ANOMALY

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ABSTRACT

Quadricuspid aortic valve is a rare congenital anomaly. It usually appears as an isolated congenital malformation, but it may be associated with other anomalies, the most common being is the coronary arteries abnormalities. Its diagnosis is often missed even with the use of trans-thoracic echocardiogram. Many of these patients progress to aortic incompetence later in life, hence requiring surgical intervention. We report the case of a 60-year –old female with diabetes mellitus, hypertension and history of brain tumour became unresponsive and was taken to the hospital and was pronounced dead. At autopsy a quadricuspid aortic valve was noted.

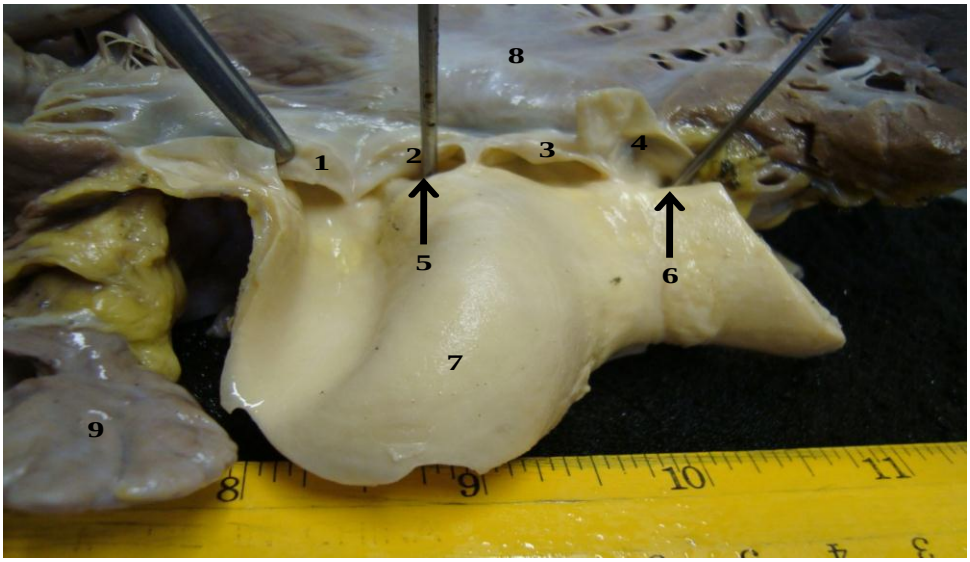
INTRODUCTION

Quadricuspid aortic valve is a rare cardiac anomaly, with an estimated incidence of 0.003 to 0.043% of all congenital heart anomalies. It appears as an isolated congenital malformation [1,2,3], but it may also be associated with other anomalies, including patent ductus arteriosus, ventricular septal defect, pulmonary valve stenosis, mitral valve malformation, left ventricular outflow tract obstruction and the most common being coronary artery anomalies [4,5, 6]. Quadricuspid aortic valve was first identified on autopsy by Ballington in 1862 [7], the first in vivo description was reported by Robicsek et al., in 1968 [8]. Since then the review of literature suggest that there have been 200 cases described, most in adults. The largest review published to date included 184 patients, aged between 2 and 84 years, with slight male predominance. We report the case of a 60-year old female with quadricuspid aortic valve noted during the autopsy.

CASE REPORT

A 60 year old female with diabetes mellitus, hypertension and history of brain tumour became unresponsive and was taken to the hospital, despite resuscitation, the patient was pronounced dead. Autopsy was done and it was noted that the brain was weighing 1190 gm with multiple lytic lesions along with osteoma was noted in the skull bone. Right lung was weighing 700 gm and the weight of the left lung was 500 gm, both the lungs showed severe congestion and oedema. The heart weighed 600 gm. The left anterior descending artery was calcified and 50 % occluded, posterior-lateral wall of heart had myocardial fibrosis, a quadricuspid aortic valve was noted [Fig. 1]. The total length of the Aortic valve was measuring 7 cm and four cusps of equal size one anterior, one posterior and two lateral in its position were noted in aortic valve along with the corresponding aortic sinuses. Right coronary artery was arising from the anterior aortic sinus and the left posterior aortic sinus was giving origin for the left coronary artery. Left ventricle thickness was 2 cm. The total length of the mitral valve was 11 cm, tricuspid valve was 10 cm and the pulmonary valve was measured 7 cm respectively. The thickness of the right ventricle wall was measuring 1cm. There were no any other abnormalities was noticed.

Figure 1. 1,2,3,4 – Aortic cusps, 5 – Left posterior aortic sinus, 6 – Anterior aortic sinus, 7 – Wall of the ascending aorta, 8 – Cavity of the left ventricle, 9 – Left auricle



DISCUSSION AND CONCLUSION

Quadricuspid aortic valve is a rare congenital anomaly, most cases have been discovered incidentally at autopsy or during aortic valve surgery, or by aortography or by endocardiography [9-13]. Hurwitz and Roberts classified the quadricuspid aortic valve into seven types on the basis of cusp size and the degree of cusp equality. According to their research, around 85% of cases are of type A, which has four equal cusps. Our case report also supports their findings. The mechanism of the congenital malformation is not fully known. Development of semilunar valve begins during the fourth week of gestation by two mesenchymal ridges which appears in the cephalic portion of the truncus arteriosus. These truncoconal ridges fuse and descend in a spiral fashion into the ventricles forming the aorticopulmonary septum. Each semilunar valve is formed from three mesenchymal swellings at the junction of the conus and truncus. These mesenchymal swellings grow to form triangular shaped valves with their endothelium covering become excavated on their distal aspect to form the cusps. This process of formation of the cusps will get completely developed by ninth week of gestation. Different mechanism has been reported that could alter the number of cusps. These includes anomalous septation of the conotruncus, excavation of one of the valve cushions, division of one of the mesenchymal ridges that normally give rise to the three aortic valves [14,15].

Development of the aortic valve occurs just after the coronary artery origins from the aortic sinus. It is possible that these two anomalies may therefore be embryologically related. Awareness about details of variation of quadricusp aortic valve helps the surgeons to properly place the prosthetic valve at the time of valve replacement and may also serve as a useful guide for both radiologists and cardiothoracic surgeons. It may help to prevent diagnostic errors, influence surgical tactics and interventional procedures and avoid complications during surgical procedures.

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