Papilloma of Cusp of the Aortic Valve

Report of a Patient with Sudden Death

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SUMMARY
This is a case report of sudden death in a 50 year old man with a history of recent onset of atypical anginal pain. Death appears to have been caused by a small papilloma attached by a pedicle to the edge of the right cusp of the aortic valve. This papilloma occluded the right coronary ostium in a heart with a predominant right coronary circulation.

Additional Indexing Words:
Angina Right coronary ostium Intermittent occlusion

This patient died 23 years ago. The case was not reported at the time because it was thought to be too rare, that it would be impossible to diagnose by methods then available, and that it would require color illustrations which were then not used by most medical journals.

At this time many of these problems have changed and the case is now presented briefly because with modern methods of cineangiography and perhaps echocardiography such a diagnosis might be made in vivo and corrected surgically.

History
The patient was a 50 year old male with no significant family history or past history of cardiovascular disease. Eight days prior to admission to the University Hospital the patient developed sudden onset of severe constricting precordial pain with radiation into both arms. This persisted intermittently and was aggravated by exercise but also occurred at rest. After three days he consulted a physician who did not take an electrocardiogram but prescribed nitroglycerine which gave some, but not always complete, relief. He was seen by one of us (CAP) five days later. The electrocardiogram showed an isoelectric T in aVL and negative T waves in the chest leads 4 and 5 (these may have been CF leads). The patient was hospitalized the same day with the diagnosis of lateral wall myocardial infarction.

Physical Examination. Physical examination in the hospital was not remarkable in any way except that the BP was 150/90 and 170/110 on two occasions.

Laboratory Findings. RBC 4.5 million/mm³, Hgb 14.5 g/100 ml, WBC 9,500/mm³ with a normal differential. The sedimentation rate was 1 mm/hour and the serum cholesterol was 175 mg/100 ml.

Course. The patient was placed on dicumarol therapy. We were puzzled by the atypical nature of the anginal attacks which came on without any particular relation to activity, ingestion of food or emotional upsets during hospitalization and often occurred when the patient was sitting or lying quietly. An electrocardiogram was taken five days after admission and although the T in aVL was slightly negative the T waves across the precordium (V leads) were all positive. On the eighth hospital day the patient was found dead in bed at 4:15 p.m.; death had evidently occurred only a short time before.

Autopsy
The pertinent findings as recorded by the pathologist were as follows:

Heart: Weighs 400 grams and measures 13.0 × 9.0 × 6.5 cm. The relation of the great vessels is natural. The epicardium is everywhere smooth and glistening. The organ feels firm. The left auricle is not remarkable. The mitral valve has a circumference of 9.5 cm. Its leaflets are pliable and competent. The chordae tendineae are long, thin and straight. The papillary muscles are normal. The
left ventricle is hypertrophied. Its wall averages 1.8 cm in thickness. The myocardium appears natural. The aortic valve measures 7.5 cm in circumference. Its cusps are thin and pliable. The noduli Arantii are prominent. On the appositional edge of the right cusp there is a small polypoid structure which measures 4 mm in diameter and is attached by a narrow pedicle. It is pinkish in color and appears gelatinous. Fine strands of fibrous tissue radiate from its pedicle through its substance. The polyp is freely movable and, at the time the heart was opened, was seen to lie directly over and pushed into the ostium of the right coronary artery, effectively plugging it up (fig. 1). The ostium itself is depressed below the aortic intima in a funnel-like manner. The polyp is easily removed from the position by the natural motion of the valve, but tends to fall back again when the valve is opened. The ostium of the left coronary is unremarkable.

On tracing out the coronary circulation, the vessels are noted to be patent throughout and remarkably free of atheromatous thickening. The right coronary artery is seen to predominate in supplying the heart, the lateral descending vessels of the left ventricle arising from the exceptionally long right circumflex artery. The left coronary supplies only the anterior surface of the left ventricle and part of the left lateral margin. The myocardium is everywhere firm and of normal color.

Figure 1
(Upper) Right cusp of aortic valve with polyp occluding the right coronary ostium found when the heart was opened at autopsy. (Lower) Same view but polyp has been pulled out of ostium of right coronary and is draped over the edge of the valve cusp with a piece of paper behind it so that the size can be delineated.
Microscopic Examination

Heart: Section D is of the polyp of the aortic valve. It is composed of a papillated mass whose stroma is acidophilic and homogeneous and which is lined by a single layer of large endothelial cells. Trichrome Stain: Reveals the stroma to contain bands of delicate wavy collagen fibers. Mucicar-mine and Twort Stains: Show evidence of mucus secretion.

Discussion

We have made no attempt to search the literature in detail, but in the intervening years we have not encountered any references to a similar case.
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