Congenital Aneurysm of the Pulmonary Sinus of Valsalva

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SUMMARY
A case of congenital aneurysm of the pulmonary sinus (sinus of Valsalva) in a 7-year-old child is described. The child met a traumatic death and manifested no symptoms or definite signs of his valvular defect. The heart was of normal weight, and no other abnormalities of the heart or other organs were present. This is the first report of sinus aneurysm of the pulmonary valve. The pathogenetic implications and possible clinical consequences of this defect are discussed.

Additional Indexing Words:
Pulmonary artery Pulmonary valve

The lesion that is termed an “aneurysm of the sinus of Valsalva” has not been reported to involve the pulmonary valve. The present report of a dissection at the base of the sinus of the anterior pulmonary valve cusp is of further interest because of the young age of the patient and the histologic aspects of the lesion.

This defect was an incidental finding in a 7-year-old white boy who died as the result of cerebral injuries incurred when he was struck by a car. In the hospital he had a blood pressure of 130/90 mm Hg and a variable pulse rate. A grade II/VI systolic ejection murmur extending to the neck was auscultated by several observers, and the second heart sound was normally split. The electrocardiogram revealed only acute changes consistent with observed electrolytic abnormalities.

Autopsy Findings
The patient was of normal appearance and development for his age. Congenital defects were noted only in the heart. The heart, which was in the normal site, weighed 95 g with the right ventricular wall measuring 2 mm in thickness and the left ventricular wall measuring 9 mm without abnormality. The foramen ovale, interatrial septum, and interventricular septum were closed and of normal configuration. Each of the valves and related structures were normal with the exception of the sinus of the anterior pulmonary valve cusp (figs. 1 and 2). This cusp itself was also without abnormality, except that at the central portion of the base of the sinus there was a 4 by 2 mm oval defect which led into a 6 by 6 by 4-mm sac which presented immediately beneath the valve cusp (fig. 2). This saccular diverticulum rested on the myocardium of the anterior pulmonary outflow tract with its outer, delicate fibrous wall bulging slightly over the attachments to the myocardium at each lateral extent. The lining of the sac was smooth, but there were irregularities of the inner wall consisting of narrow fibrous bands passing along the wall from the mouth to the base of the sac, some of which then extended into the underlying myocardium. Most of these bands remained closely fixed to the inner aspects of the wall. There was minimal increase in the opacity of the immediately surrounding endocardium which was little different in

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The pulmonary and aortic valves viewed from above utilizing the BNA nomenclature.\textsuperscript{8} $P =$ posterior or noncoronary cusp of the aortic valve; $RA$ and $LA =$ the right and left coronary cusps of the aortic valve, respectively; $RC$ and $LC =$ the right and left coronary arteries, respectively; $RP$ and $LP =$ the right and left cusps of the pulmonary valve, respectively; $A =$ the anterior cusp of the pulmonary valve.

Thus the aortic cusp including the origin of the right coronary artery is the right cusp, and the right pulmonary cusp is the cusp of the pulmonary artery which is apposed to it. The left cusp of the aortic valve is similarly related to the left pulmonary cusp and the remaining pulmonary cusp is termed the anterior pulmonary cusp. The defect in the present case was in the anterior sinus.

The opened outflow tract of the right ventricle showing the anterior pulmonary cusp on the left. Enlarged about $\times 2$. Note the lesion presenting immediately beneath the central portion of the anterior cusp. The lower borders are delineated by the two arrows.

appearance from the endocardium beneath the adjacent cusp (fig. 2). The underlying musculature was normal.

Sections of the involved pulmonary sinus and aneurysmal cavity revealed no inflammatory reaction. Above the midportion of the involved sinus the pulmonary arterial media terminated sharply in a transverse plane at a nodular collection of almost organoid, dense, mature collagen (fig. 3). This latter structure is interpreted as representing a portion of the annulus fibrosus. Sections taken slightly to either side of the center of the sinus defect showed relatively more annulus fibrosus tissue (fig. 4) until at the most lateral reaches of the lesion, the valve cusp, annulus and pulmonary artery had almost normal relationships (figs. 5 and 6). The inner lining of the aneurysmal cavity consisted of a single layer of flattened endothelial cells. Immediately beneath the lining and parallel to it were a few thin, loosely arranged positive Verhoeff fibers. Between the inner lining and the myocardium were thin bundles of collagen that were orientated parallel to the wall of the cavity. Because of its configuration this fibrous tissue is interpreted as reactive fibrosis. The valve cusp had a normal structure, except that at the base where the fibrosa would normally blend with the well-formed collagen of the annulus fibrosus, the fibrosa of the cusp blended with the reactive and less compact reactive fibrous tissue of the wall of the aneurysm.

Microscopic examination of the other two pulmonary valve cusps and sinuses as well as all three commissural regions revealed normal anatomy. The relationship of the aortic and pulmonary arterial media to the annulus fibrosus is subject to some variation, but in the central portion of the cusp the media tapers down to extremely delicate elastic fibers ending in a wedge-shaped edge cephalad to the annulus with the narrow end of the wedge at the intimal aspect.\textsuperscript{2} This termination is sharp and differs in a constant fashion from that in the region of the cusp commissures where the media ends in a more transverse

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Figure 3
Microscopic section of the lesion taken at the central portion of the mouth of the aneurysm. Verhoeff-van Gieson stain, \( \times 11 \). At the termination of the pulmonary artery (p) which takes the black elastic tissue stain, the tissue of the annulus fibrosus (f) is seen as a collection of rounded dense collagen fibers. Below this the pulmonary valve cusp (v) does not meet the annulus to form the base of the sinus of Valsalva. Instead, this area becomes the mouth of an aneurysm (an) which extends beneath the endocardium of the right ventricle (en) as was grossly apparent in figure 2. A round, dark postmortem blood clot is present in the sac just above the “an” lettering.

Figure 4
Microscopic section, Verhoeff-van Gieson stain, \( \times 39 \), of the meeting of the pulmonary arterial media with the annulus fibrosus at the central portion of the mouth of the aneurysm. The organoid collections of collagen immediately beneath the ending of the dark elastic fibers contrast with the linear collections of collagen bundles which continue from this level to line each dissecting pocket of the lesion.

arterial media ends in the manner normally observed at the commissural region, and the small remnant of annulus tissue present is poorly formed, soon giving way to the reactive fibrous tissue which lines the entire sac.

Comment
This lesion is interpreted as an aneurysm of the pulmonary sinus of Valsalva, and this report appears to be the first of a case in which the pulmonary valve was involved.\(^3\) In the reports of aortic sinus aneurysms, the absence of inflammation\(^3,4\) has been taken as evidence for a congenital origin. Since the present lesion is regular in gross morphology and completely devoid of inflammatory cells and also because it occurred in a child without history of inflammatory cardiac disease, the
lesion is believed to be due to a congenital defect.

A number of theories of pathogenesis for similar congenital lesions in the aorta have been proposed. As originally described by Gegenbaur, the separation of the bulbus arteriosus by the bulbo-spiral septum divides the two lateral endocardial cushions and one half of each becomes the anlage of contiguous cusps of the aortic and pulmonary valves. The anterior and posterior cushions are not affected by this division. The aortic and pulmonary valve cusps maintain roughly the same relative positions as in the developmental period and therefore, the anterior pulmonary cusp and its counterpart in the aortic valve, the posterior or noncoronary cusp, are molded from endocardial cushion tissue which is not related to the bulbo-spiral septum. In the present case the involved sinus is that of the anterior pulmonary cusp, and consequently, as has been pointed out for aneurysms of the analogous aortic noncoronary cusp, a defect in the development of the distal bulbar septum will not account for an aneurysm in this sinus. In an attempt to formulate a theory of pathogenesis applicable to all aortic sinuses, Venning cites C. V. Harrison’s suggestion that the fundamental defect could be in the development of the elastic tissue at the base of the aorta. Edwards and Burchell have agreed and have illustrated the lack of continuity between the aortic media and the ring of the aortic valve.
The lesion in the present case differs from that reported by Edwards and Burchell in that the media of the pulmonary artery meets a bundle of fibrous tissue at the superior extent of the aneurysm (figs. 3 and 4). The nodular configuration of this bundle and the regular, organoid pattern of its collagen are interpreted as indicating that this structure is part of the annulus fibrosus. If this interpretation is correct, then the defect would be in the annulus fibrosus or at the junction of the annulus and the fibrosa of the valve cusp. Gibbs and Harris suggest this possibility and note that a criticism of the hypothesis that aneurysms result from defective development of the endocardial cushions is the fact that congenital aneurysms of the pulmonary sinuses have not been described. Most reports of aortic aneurysms that include histologic findings often comment only on the evidence of inflammation, rheumatic or syphilitic disease, or are able to present the histologic aspects of only a surgically resected aneurysm sac. In proposed congenital cases, attention often has not been focused on the relationship of the termination of the media to the ostium of the aneurysm. When it has, the findings of Edwards and Burchell have been confirmed by some investigators while others have encountered different anatomy.

Detailed histologic studies of aneurysms in the analogous aortic noncoronary sinus would be of particular interest since, in common with its counterpart, the anterior pulmonary sinus, the adjacent arterial media presumably develops from mesenchyme present at the site rather than from that which descends with the bullospiral septum. Therefore, in these sinuses the union of the media with the annulus might be less likely to suffer from deficiencies, and the site of the defect, as in the present case, may be found in the annulus itself. This was not the situation, of course, in the noncoronary sinus case of Edwards and Burchell and an analogy between these two sinuses is weakened by the fact that the anterior half of the noncoronary sinus is situated immediately above the membranous interventricular septum. This circumstance is of apparent importance since many of the noncoronary sinus aneurysms occur at this point.

Other interesting features in this case are the youth of the patient (7 years old) and the fact that the lesion did not present clinically. The great preponderance of congenital aortic sinus aneurysms have manifested themselves beyond the second decade, and at one time it was thought that rupture never occurred in young children. Subsequently a small number of well-documented and other less compelling cases occurring in childhood have been recorded. In this age group, sinus aneurysms must be distinguished from aortico-left ventricular tunnels. The latter are characterized by having their point of origin above the ostia of the coronary arteries and having continuation of the aortic media into the tunnel. The present lesion displays only sparse elastic tissue in its wall and its saccular configuration is much more in keeping with the congenital sinus aneurysms of the aorta. None of the reports of comparable aortic sinus cases in the pediatric age group have included information on the relationship of the media to the site of dissection. Detailed anatomic study of such cases would afford the opportunity, as did the present case, to observe the lesion relatively unaltered by infection or secondary changes. The fact that this lesion did not attain clinical significance is probably attributable to the low pressure in the pulmonary circuit. Indeed, as has been suggested, the low pulmonary artery pressure may be the reason why sinus aneurysms have not been previously encountered in the pulmonary valve.

The natural history of aneurysms of the aortic sinus of Valsalva is dissection from a local congenital defect and production of a characteristic fibrous sac which perforates into the right atrium or ventricle and ruptures. Dissection into the interventricular septum has rarely produced rupture into the left ventricle in the absence of infection. No case identical to the present one has been described in the aorta, but two cases with aneurysms of

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all three sinuses have presented gross appearances in the lesser involved cusps which are similar to that of this case. Presumably these aneurysms are lesser degrees of the accompanying lesion which produced the symptoms, and they may be considered, along with the present case, to represent an earlier stage in the development of the lesion. The present lesion could have enlarged and might have produced regurgitation without rupture as did a similar case in the aortic valve. It also may have continued to dissect and penetrate into the right ventricular outflow tract or into the pericardial sac.

The case reported adds to the spectrum of defects in the sinuses of Valsalva and presents new material for the consideration of the pathogenesis of these lesions. It is apparently the first reported case of such a defect in the pulmonary valve.

References


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