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CASE REPORT

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Cardiac Hemangioma: A Rare (or Possible) Cause of Sudden Death in Children

Recently we had occasion to study an apparently healthy 13-year-old girl who died suddenly and unexpectedly. Necropsy revealed that her death was most likely caused by an arrhythmia induced by hemangiomatous involvement of the ventricular conduction system. Cardiac neoplasms have commonly been described in literature of the English language [*1*], but hemangiomas, especially those of small size, have rarely been reported [*1-9*]. This lack of reports may be attributed to a failure of detection. It would appear from published cases that some lesions, whether or not hemangiomatous, may be so diminutive as to pass unnoticed even at necropsy examination [*1,10-13*]. For this reason, as well as the rarity of our encounter, we report the following case.

Case Report

A 13-year-old white female was found unresponsive in bed. Despite vigorous resuscitation efforts, she died. Except for having complained of a mild earache several days earlier, she had been in good health. In particular, there was no history of a cardiac disorder or of trauma.

At necropsy the well-developed, well-nourished body was 164 cm long with a weight of 45 kg. Pathologic findings were confined to the 350-g, slightly enlarged heart. A 4.0 by 3.5 by 3.5-cm spongiform lesion from which blood could easily be expressed extended from the subbasal region of the anterior ventricular septum to the apex (Fig. 1). With light microscopy the lesion was seen to consist of a cavernous hemangioma characterized by large vascular channels with large muscular venous or simple endothelial walls (Fig. 2). The ventricular septal Purkinje fibers were invaded by hemangioma (Fig. 3) and some were surrounded by dense fibrous tissue at foci wherein the neoplasm was undergoing sclerosis. The atrioventricular node and the bundle of His were free of neoplasm. The coronary arteries followed a normal distribution and were widely patent. Hemangiomas were not found in any other organ.

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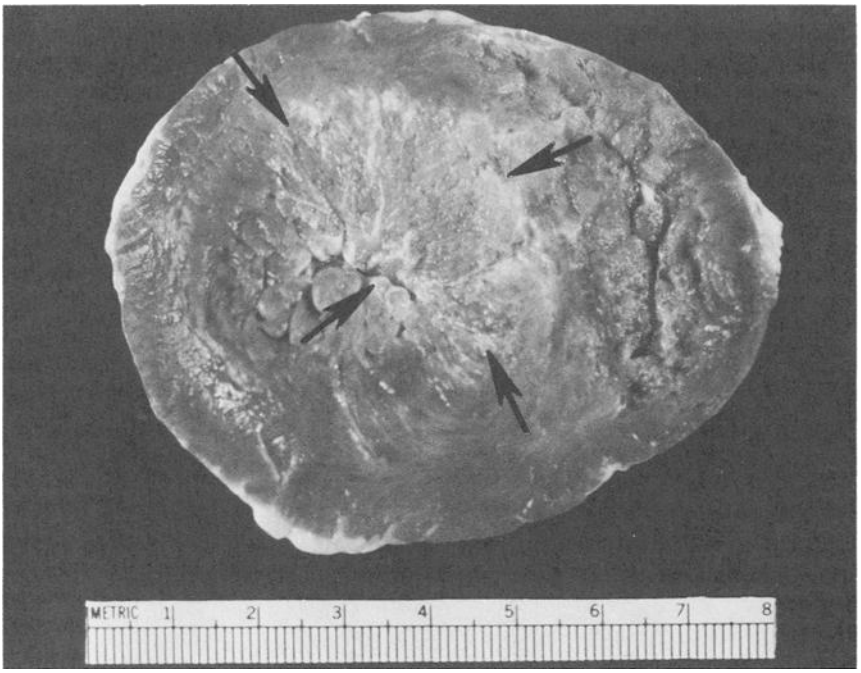

FIG. 1—*The ventricular septum is markedly widened by the presence of the hemangioma, which is outlined by the arrows.*

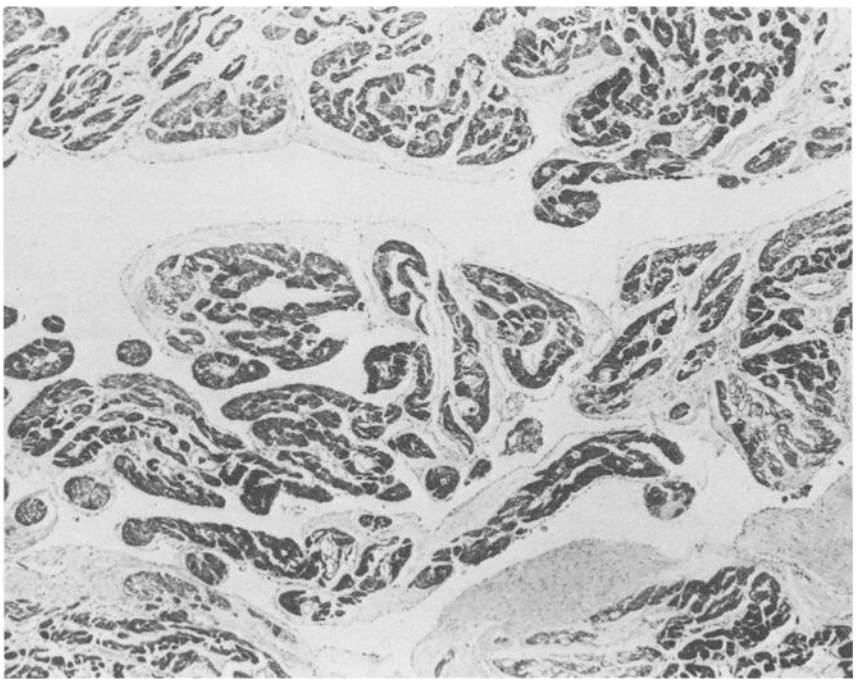

FIG. 2—*Cavernous vascular channels separate myocardium into numerous islands. Various degrees of endocardial fibrosis are present. (Gomori trichrome, ×25.)*

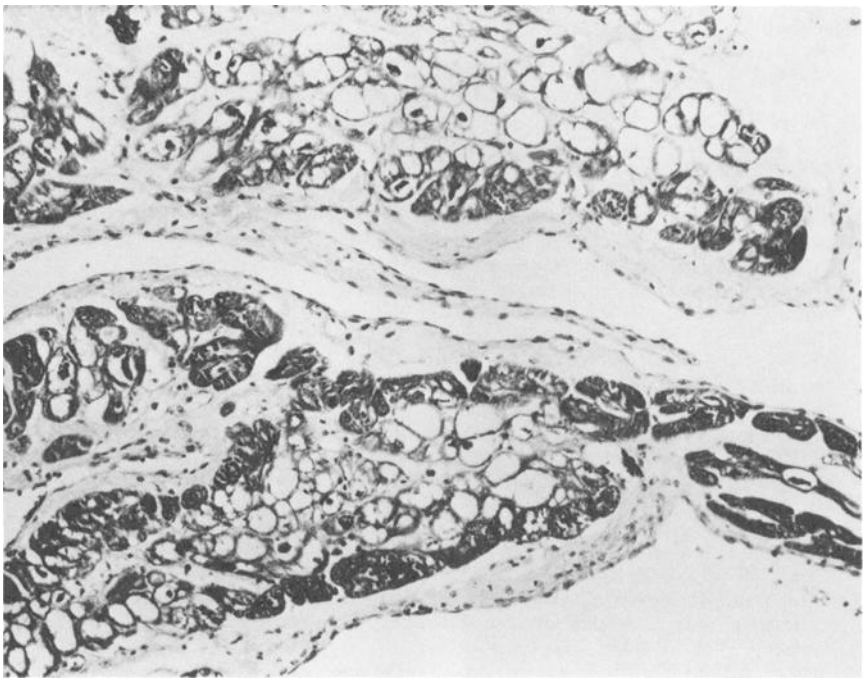

FIG. 3—*Subendocardial Purkinje fibers of the ventricular septum are intimately involved by cavernous vascular channels. Blood cells are artifactually absent. (Hemotoxylin and eosin, ×100.)*

Discussion

We have been able to find only about 40 reported cases of cardiac hemangioma in the world literature [*2*]. The patients with cardiac hemangiomas greater than 2.0 cm in diameter ranged from 8 to 57 years of age; they had neoplasms either within the myocardium or tumors that were pedunculated from the epicardial or endocardial surface [*1-7*].

A history of conduction disturbances was prominent in two adult patients reported to have cardiac hemangioma [*2,7*]. Other than for occasional reports [*2,7-13*], cardiac neoplasms or hamartomas have not been associated with arrhythmias or with sudden death.

In the English literature only nine of the described cardiac hemangiomas were larger than several millimetres [*1-9*]; one was associated with sudden death [*8*]. In each of the patients hemangiomas were found only in the heart, the other organs being spared. In a cooperative international study in which 254 pediatric cases of sudden death of cardiovascular causes were reported, there was not a single instance of sudden death associated with cardiac neoplasm; congenital cardiac malformations accounted for the vast majority of fatalities, and inflammatory and cardiomyopathic conditions were observed in another 20% [*14*].

Recently, Wedemeyer and Breitfeld [*13*] reported necropsy findings of an infant who had been treated with pulmonary artery banding in palliation of an atrial and ventricular septal defect; the patient had suffered paroxysmal atrial tachycardia which, at necropsy, was shown to be associated with a hamartoma within the cardiac conduction system [*13*].

In our patient the ventricular septal Purkinje fibers were invaded by hemangioma. The tumor mass was large—4 by 3.5 by 3.5 cm—and thus was readily recognizable. It may be, however, that more often than is realized sudden death of obscure cause might result from cardiac neoplasm or hamartoma of much less conspicuous size.

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