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Asymmetrical Hypertrophy of the Heart: Two Sudden Deaths in Adolescents

Since Teare's report of the first eight cases describing the pathologic features of asymmetrical hypertrophy of the heart [*1*], there have been few subsequent documentations of this condition in the forensic literature [*2*]. This seems rather surprising, since most of these deaths are in the sudden and unexpected category and should be candidates for proper medicolegal investigation. Misdiagnosis, certification without autopsy, and lack of documenting isolated cases are probably equally responsible for this phenomenon.

While mentioned as occurring in sporadic form, Teare and others have also recorded familial incidences [*3,4*]. Marshall has reviewed his nine-year experience, during which he investigated sixteen cases [*5*]. Additional reports have stressed the clinical hemodynamic findings in the patients under study [*6,7*]. It is at least a variant, but may be the fundamental anatomic defect in the condition known as idiopathic hypertrophic subaortic stenosis (IHSS), the pathogenesis of which has not been completely elucidated [*8*]. Tumor, infection, metabolic storage disease, and congenital malformation of muscle have long been considered as possible etiologic agents [*9–11*]. Gross findings of heart specimens may have some similarities to hypertensive cardiac conditions, but the important clinical history of an elevated blood pressure has usually not been elicited. Therefore, when death occurs in nonhypertensive individuals, especially when they are young, the pathologist should be more alerted to this unusual entity. We thought it appropriate to add two additional instances observed within a brief time period (one month) from this office.

Case 421

A 15-year-old white male had completed about fifteen minutes of limbering-up exercises in spring football training at the local high school athletic field when he suddenly collapsed. He was taken to a nearby hospital and expired in the emergency room. He had played varsity sports in junior high school and the preseason interscholastic league physical examination, which had not included a chest X-ray, revealed no abnormalities. According to classmates, his only complaint the day of demise was that of a headache.

At autopsy, the heart weighed 440 g (normal is 230 g) and there was a prominent bulge of the left ventricle. In the subaortic area, the left ventricle measured 40 mm and narrowed to 10 mm at the apex. The hypertrophied portion involved the septum, as well as the anterior left ventricle (Fig. 1). The myocardium at this point appeared lighter tan brown with prominent trabeculae coursing through the muscle bundles. A few small, somewhat

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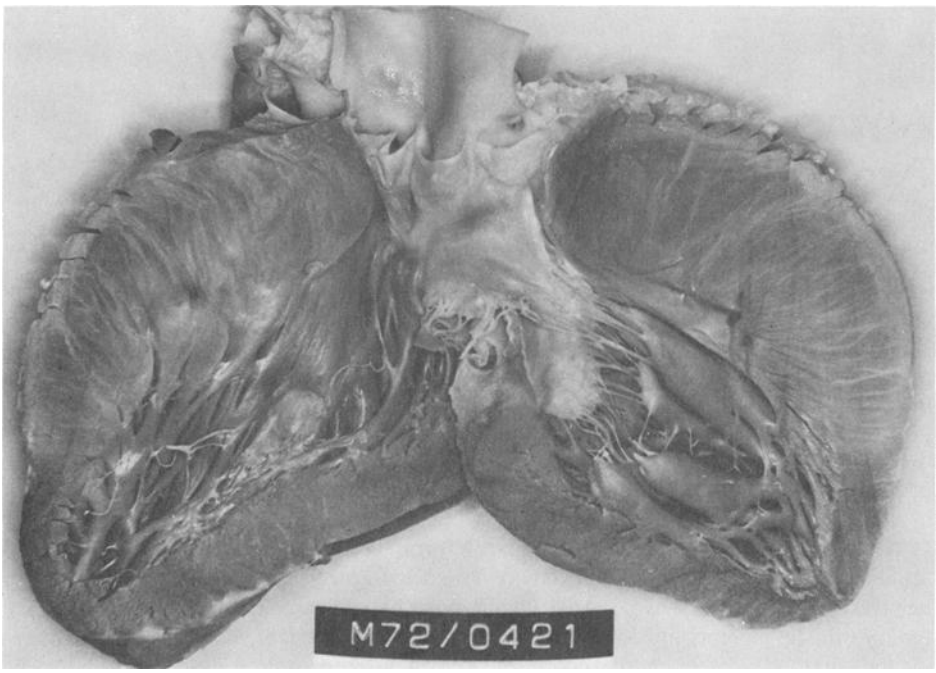

FIG. 1—*The opened left ventricle demonstrating the marked hypertrophy of the interventricular septum and the anterior wall of the left ventricle. Small lucent gray foci of ischemic necrosis are noticeable.*

lucent gray fibrotic foci were apparent. There were no other cardiac defects, congenital abnormalities, or natural disease processes present.

Microscopic examination of the hypertrophic myocardium revealed, in addition to the striking jumbled pattern of myocardial fibers, occasional small areas of healing ischemic necrosis (Fig. 2). These varied from loose vascular connective tissue with mononuclear cells to a more dense fibrous tissue. Occasional small coronary artery branches displayed focal eccentric intimal fibrosis compromising the lumen. Nerve trunks were prominent in some sections. The hematoxylin-basic-fuchsin-picric acid (HBFP) stain revealed acute ischemic change in small groups of myocardial fibers in a few scattered areas.

The pertinent history revealed that the parents of this teenager are separated and the whereabouts of the father is unknown. The mother, a 39-year-old white woman, has been followed for two years with organic heart disease, thought to be rheumatic in nature. One 20-year-old brother and one 24-year-old sister are reported to be in good health. Following the autopsy findings in this case, examination of one 18-year-old sister revealed a slight heart murmur. Right and left heart catheterization was performed and a slight peak systolic gradiant was recorded across the outflow tract of the left ventricle during Isuprel® infusion. These findings are compatible with hypertrophic myocardiopathy with a slight obstructive component. Other family members have not submitted to similar studies.

Case 465

An 11-year-old white female was walking with her father near their residence when she "tripped" onto the sidewalk, falling on her face. She was dead on arrival at a nearby hospital. The parents stated that she had recently complained of "fatigue," but had no other major medical problems. There was no history of heart disease in the family, which included one healthy 14-year-old sister. Shortly after birth, one pediatrician noted a "fast heartbeat," but no diagnostic studies were performed and no complaints or findings referable to the heart were subsequently noted.

At autopsy, the heart weighed 275 g (normal is 122 g) and was somewhat globular in shape. The majority was taken up by the left ventricle, which measured 16 mm in thickness in the subaortic aspect, but narrowed to 10 mm at the apex. The septum adjacent to the left ventricle was thickened to 20 mm, which narrowed to 10 mm at the apex of the ventricular septal margin (Fig. 3). The right ventricle varied from 3 to 5 mm in thickness. The left auricle was 3 mm in thickness and was slightly dilated; the foramen ovale was closed. The papillary muscles were proportionately enlarged, but neither softened nor fibrotic. The chordae tendineae appeared to be slightly shortened but were not fused. The valve structures, ostia, and coronary arterial system were normal with the right coronary artery appearing slightly dominant. There was no endocardial fibroelastosis, abnormal venous return, or septal defect. There were no other cardiac defects, congenital abnormalities, or natural disease processes documented at autopsy. The brain was slightly swollen with minimal frontal subarachnoid hemorrhage, consistent with the terminal fall.

Microscopic study of the myocardium from the affected area revealed the classic, bizarre pattern of varying-sized fibers, mostly devoid of the usual interlacing pattern (Fig. 4). Atrophy of individual cells consisted of a "vacuolated" appearance, with many of the nuclei hyperchromatic and some mitoses noted (Fig. 5). A slight increase in fibrous tissue surrounded the myocardial bundles in these areas (Fig. 6). Other areas of the myocardium showed less obvious fiber jumbling and the vacuolated appearance was not observed. None of the sections revealed giant cells, stainable organisms, or inclusion bodies.

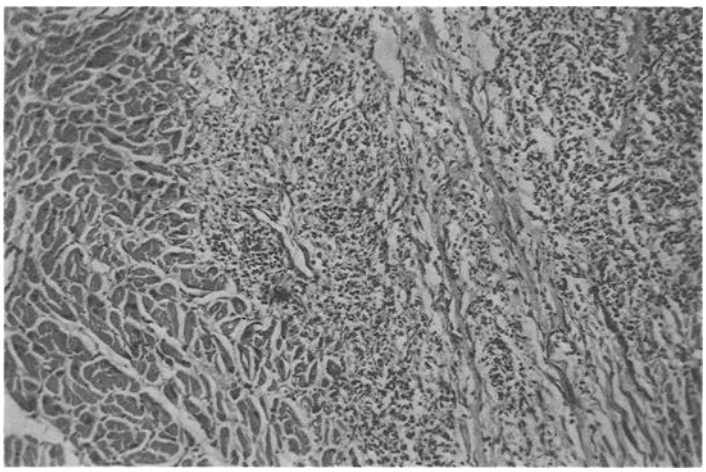

FIG. 2—*Focal areas of ischemic necrosis and repair consisting of vascular connective tissue and mononuclear cells (original magnification ×130).*

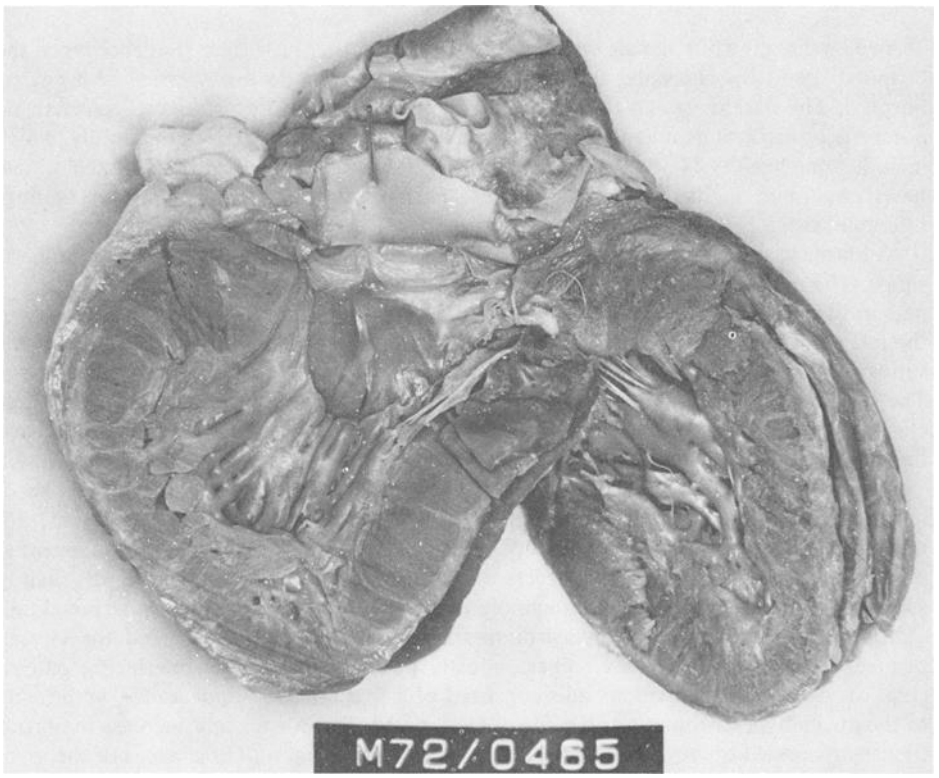

FIG. 3—*The opened left ventricle showing similar hypertrophy of the interventricular septum and the anterior wall of the left ventricle.*

The intramural arteries appeared marginally thickened. No inflammatory cells were observed. Special stains, including HBFP, were not revealing. Enzymatic stains were not performed. Attempts to persuade the remaining members of the family to undergo clinical cardiovascular studies have been unsuccessful.

Discussion

The other previously reported series of deaths from asymmetrical hypertrophy indicate a male predominance and an age range from 10 to 72 years with many occurring in the teenage period (7 of 16 cases in Marshall's series). Four of Teare's eight cases had previous cardiac symptoms, whereas ten of Marshall's reported deaths were symptom-free. The complaints ranged from occasional shortness of breath and palpitation to "blackouts" and classic epileptic fits for five years. Exercise is said not to be a precipitating factor, although one of Marshall's and two of Teare's cases were riding bicycles and another of the latter group was "running to catch a bus." However, ten of Marshall's cases were "at rest": in bed, sitting down, or standing quietly. It is of interest that two other cases in this group were sudden deaths precipitated by a blow on the chest, supporting the concept of traumatically induced arrhythmias. The study of patients by successive cardiac

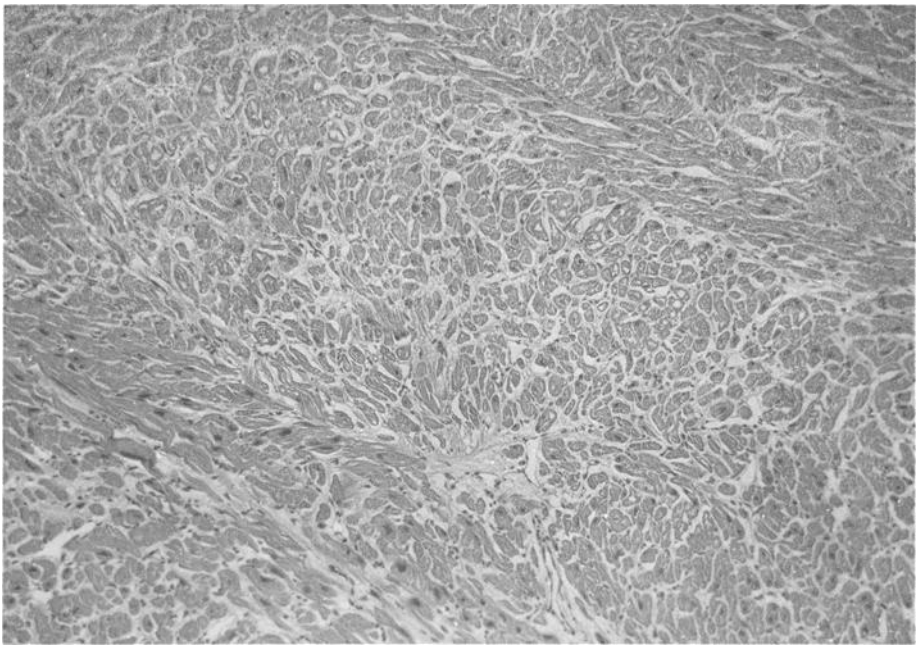

FIG. 4—*The typical jumbled pattern of the myocardial fibers with intramyocardial fibrosis and some nuclear vacuolization (original magnification ×130).*

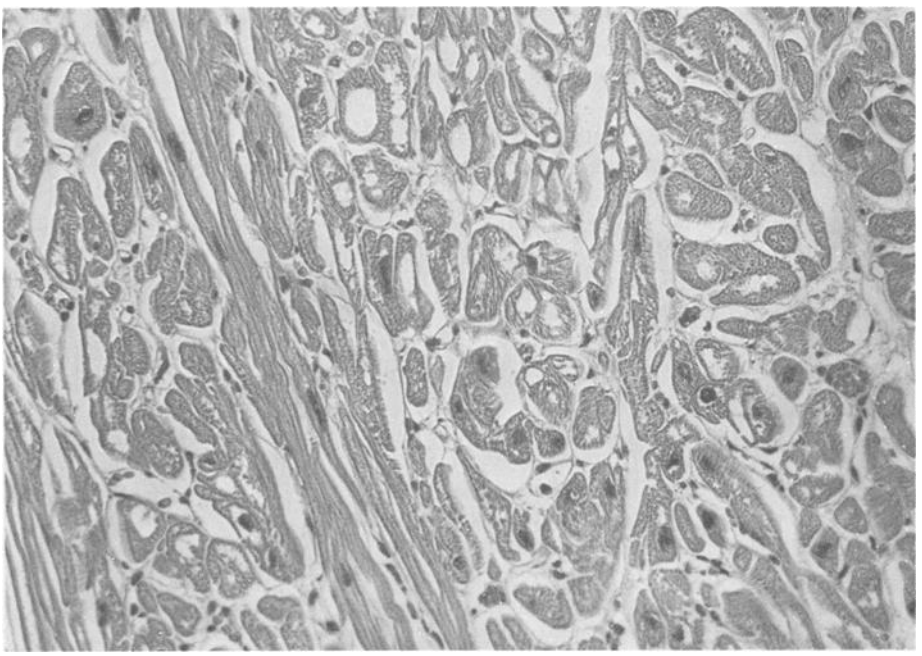

FIG. 5—*Focus of myocardium revealing the vacuolated appearance of the cells. Hyperchromatism and mitoses are observed with some cells showing atrophic changes (original magnification ×350).*

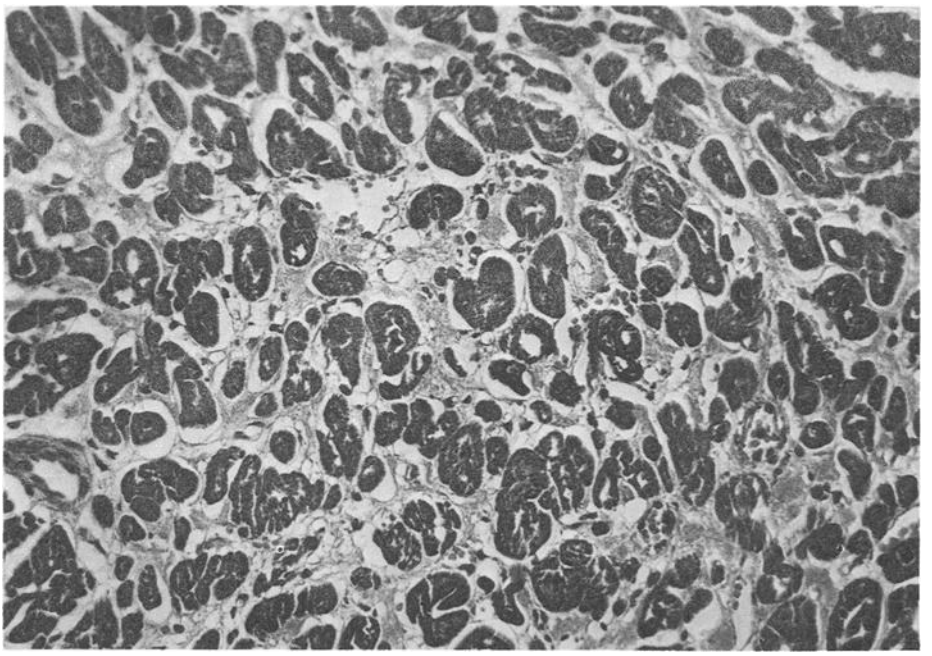

FIG. 6—*Increase of fibrous tissue surrounding the individual myocardial bundles. Hyperchromatism and mitotic figures are noted, but vacuolization is less prominent (original magnification ×350).*

catheterizations progressing from a nonobstructive hypertrophy to a typical obstructive cardiomyopathy has been documented, and a recent subject followed for five years showed such changes accompanied by a Type-B Wolff-Parkinson-White syndrome noted by electrocardiography [*6*].

The distinction has been made between the variants of location of the hypertrophy. Some instances show a combination of septal and anterior wall involvement, whereas in other cases it is confined to one or the other position, and occasional cases involve the right ventricular septum. The two cases in our series showed the combined involvement, as did half (eight) of Marshall's cases. The histologic picture appears rather constant, with variations occurring predominantly in terms of severity. However, one of Marshall's cases demonstrated the typical bizarre fiber jumbling away from the hypertrophied segment, that is, in the posterior left ventricle and in the wall of the right ventricle. Similar involvement of the atria has not been described, although mention is made of left atrial dilatation.

An aspect of public health and preventive medicine resulted from the publicity attendant to the young male athlete (Case 421). As this was the second cardiac death in the high school sport population within a short period of time, a review of physical examinations prior to athletic competition was undertaken. It was shown that chest X-rays were seldom included as part of this program and shortly thereafter, following attendant publicity, mobile units provided by the local Respiratory Health Association began giving free chest X-rays to junior and senior high school athletes. Statistics are not available on the number of students found medically unfit to participate in sporting events, but no deaths related to athletics in the high school population were reported to our office during the 1972–1973 academic year.

Summary

Two sudden deaths in adolescents from asymmetrical hypertrophy of the heart are reported. This condition may be incorrectly diagnosed and underreported for several reasons, although gross and microscopic findings are constant and striking, varying only in severity. Familial considerations and public health aspects, especially in the high school athletic population, are stressed.

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