CASE REPORT

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Sudden Unexpected Infant Death due to Fibroma of the Heart

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ABSTRACT: A 7-month-old previously healthy female infant was found dead in her crib by her mother shortly after having been laid down to sleep following the noontime feeding. Because the child did not suffer from an acute illness and no other evidence pointed to a cause of death, it was initially assumed by the police that she had died of sudden infant death syndrome. At autopsy, however, the cause of death was determined to be cardiac arrhythmia secondary to fibroma of the heart.

KEYWORDS: forensic science, sudden death, infant, heart tumor, fibroma

Primary cardiac tumors in infants are relatively rare, and in the majority of cases, grow slowly (1). The most common cardiac tumor in children is rhabdomyoma, followed by fibroma and teratoma (2). Unlike rhabdomyomas, which show a tendency to regression after birth, fibromas tend to grow rapidly (3,4). Although neoplasms of the heart in children often exhibit no clinically relevant symptoms, surgery is indicated in cases where massive tumor growth constricts the pulmonary artery/ascending aorta or infiltrates the conduction system with consequent arrhythmias (3). We describe the case of an infant girl whose lack of clinical symptoms contrasted sharply with the finding of extensive tumor growth at autopsy.

Case Report

The 7-month-old female infant was put to bed by her mother after her noontime bottle feeding. A short time later, the mother found the child lying lifeless and immediately called an ambulance. Resuscitation by the emergency medical technician (EMT) to revive the child was unsuccessful. While inserting an intubation tube, the emergency physician noted food particles in the infant's esophagus and trachea. Because the child died suddenly and unexpectedly, it was initially assumed by the investigating police officers that sudden infant death syndrome was the cause of death.

The family's pediatrician reported that they had regularly brought the infant in for check-ups. The pediatrician had examined the child for the first time one week after birth and for the last time three weeks prior to death. The child had been in good general health and exhibited age-appropriate development. Neither the pediatrician nor the parents had noticed any symptoms of disease in the child.

Autopsy Findings

The pericardium contained 10 mL of a clear yellow fluid. Opening of the pericardium revealed a 2.5 × 1.5 cm yellow cauliflower-like growth on the epicardium overlying the septum (Fig. 1). The heart weighed 108 g (age appropriate weight: 31–37 g) and was normally developed. The principal finding was a 6 × 4 cm, sharply delimited tumor infiltrating large portions of the interventricular septum and the posterior wall of the left ventricle, almost completely occluding the chamber. The cut surface was pale yellow and filiform in appearance (Fig. 2). The tumor contained grossly visible foci of calcification. There was focal fibrosis of the endocardium in the left atrium plus endo- and epicardial fibroses overlying the tumor tissue.

Secondary findings included aspirated chyme as well as petechiae of the serosa and parenchyma of the thymus. All other organs showed age-appropriate development and were normal. Chemical-toxicological studies were negative.

Microscopic Examination

Histological studies established the diagnosis of the tumor as a fibroma. Trichrome staining showed the lesion to consist mainly of collagen fibers, fibrocytes, and elastic fibers as well as foci of calcification, typical of fibromas in areas of hyalinization (Fig. 3). The cells appeared to be monomorphic and the number of mitoses was not elevated. Except for a few displaced myocardial cells still residing between the tumor tissue structures, no cells resembling muscle fibers were found. The endocardium of the left atrium and the epicardium overlying the tumor tissue had a slightly increased number of connective tissue fibers, with hyalinization and focal lymphocytic reaction.

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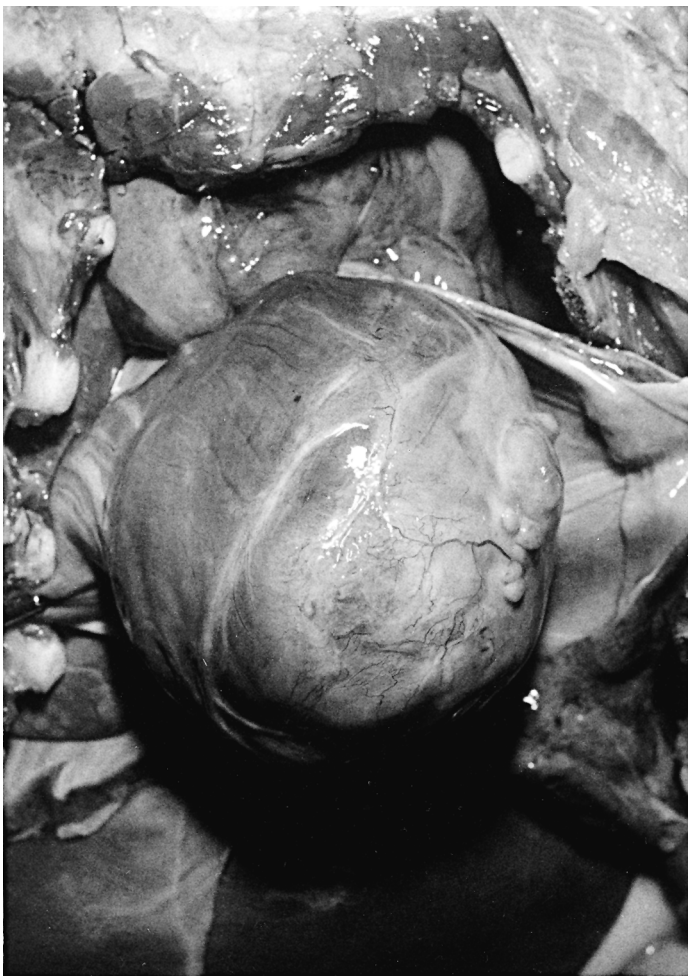

FIG. 1—*Gross photograph of the lesion consisting of a 2.5 × 1.5 cm cauliflower-like growth on the epicardium overlying the interventricular septum.*

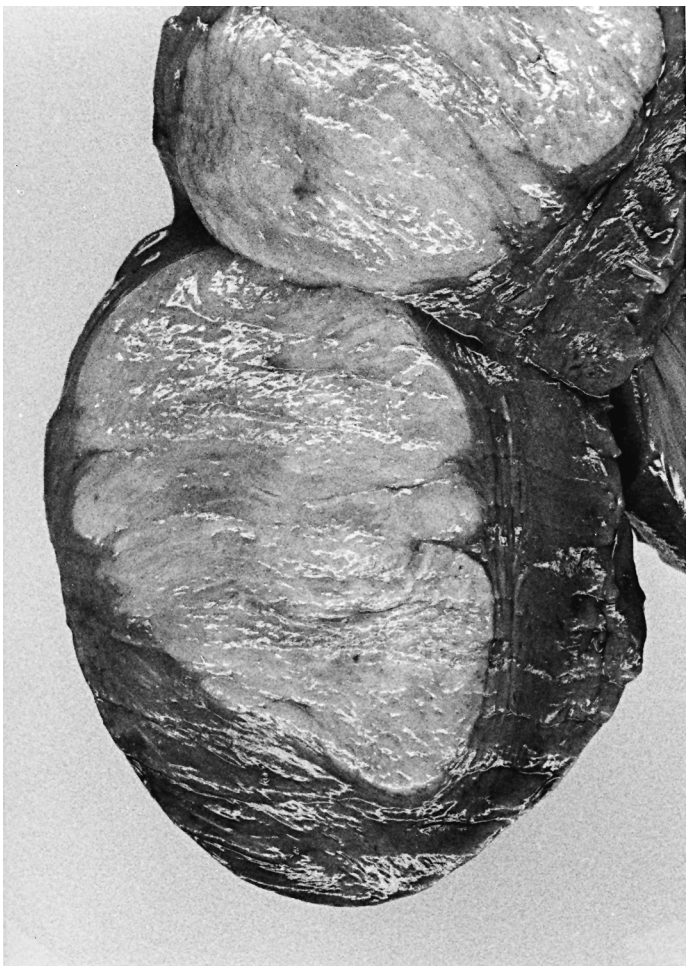

FIG. 2—*Gross photograph of the cut surface of the fibroma in the septum/posterior wall of the left ventricle.*

The lungs contained no iron-positive macrophages. There was no evidence of cerebral or cerebellar anoxia. The macroscopic diagnosis of aspirated chyme was confirmed microscopically.

Discussion

The advent of new imaging techniques has led to an increase in the detection rate of primary cardiac tumors in children. Echocardiography and sonography even allow diagnosis of these tumors in the unborn fetus (5,6). Probably because of better diagnostic techniques, a large study covering a period of 15 years showed a rise in the incidence of these tumors from 0.06% between 1980 and 1984 to 0.32% between 1990 and 1995. Among all primary heart tumors in children, rhabdomyomas comprised 78%, fibromas 11%, teratomas 2% (2). Whereas small rhabdomyomas are especially liable to spontaneous regression, fibromas tend to massive growth relative to the size of a child's heart, which can render complete surgical excision impossible (3,4). Even if the tumor is diagnosed early, excision can be very difficult and does not always lead to long-term success (4,7). About one fourth of all such patients require surgical intervention to relieve constriction of the pulmonary artery or ascending aorta, or to treat arrhythmias not responsive to medication (3,8–10). Even if complete or partial resection succeeds, a favorable long-term prognosis is not assured (2,3,11).

In the present case, up to the time of death neither the parents nor the family pediatrician observed any signs pointing to the severity of the child's disease. This accords with the findings of others that heart tumors in some patients produce no clinical symptoms (1); in other patients tachycardia or arrhythmia constitute the principal clinical finding (2,8,10). Histologically, we detected no signs of long-term diminished output by the left ventricle as evidenced by iron-positive macrophages in the lungs or neuronal hypoxia. Given the tumor's localization in the septum, it can thus be assumed that death resulted from asystole secondary to acute disruption of the conduction system. This is supported by the fact that cardiac rhythm disturbance is one of the leading signs of heart tumors (2,8,10).

For the parents, it was important that the differential diagnostic possibility of a rhabdomyoma/rhabdomyosarcoma was ruled out by the definitive diagnosis of fibroma. Thirty percent of all rhabdomyomas are associated with tuberous sclerosis, which is known to have an autosomal dominant inheritance and thus would have put siblings of the victim at risk of the same fate (12–15).

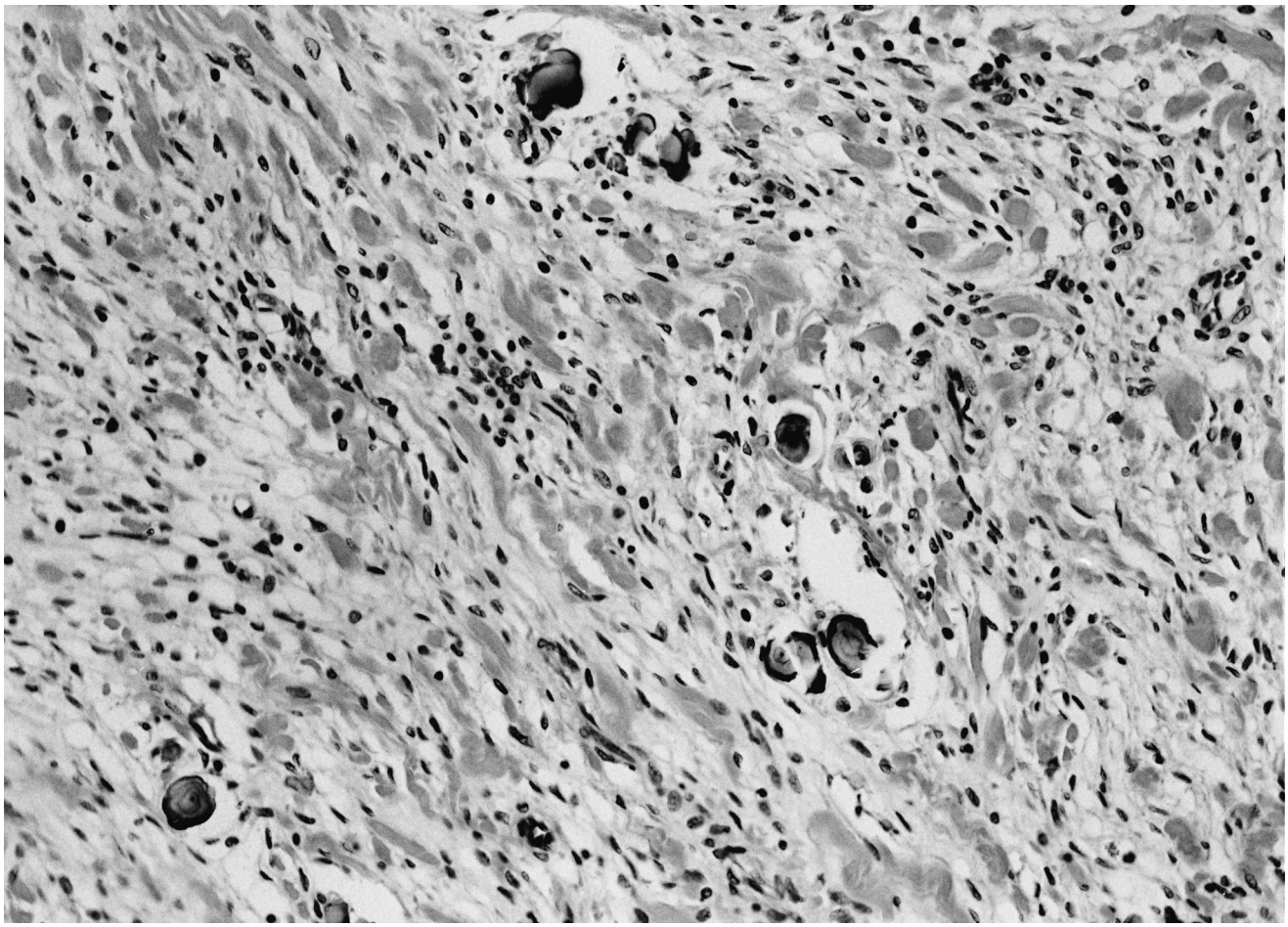

FIG. 3—*Photomicrograph of the fibroma, which is composed of fibrocytes, collagen and elastic fibers as well as of some foci of calcification. (Trichrome stain; original magnification ×100).*

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