

Varix of the heart causing outflow tract obstruction

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Summary. We report a case about a neonate who died of severe subaortic stenosis due to a giant vascular dilation of the left ventricular outflow tract.

We emphasize the fatal result of this benign lesion and make differential diagnosis with haemangiomas and valvular blood cysts.

Key words: Varix of the heart, Heart neoplasms, Blood cyst, Haemangioma

Introduction

The varix of the heart is a vascular dilation of a venous type which is most frequently located in the subendocardium (McAllister and Fenoglio, 1978).

The incidence of cardiac varices is unknown. Reported cases are chance finds in necropsy studies, that is why this kind of lesion does not usually appear in the series about tumours of the heart (McAllister and Fenoglio, 1978; Silverman, 1980; Poole et al., 1984).

Heggtveit (1966) found 25 cases of cardiac varices in a series of 1,000 consecutive post-mortem prospective studies. Histologically, the vessels consisted of irregular venous spaces with deficient musculo-elastic components in their walls, and appeared to have no clinical significance.

Materials and methods

Case report

A 26-day-old baby girl, weighing 2,600 g was admitted with a 24-hour history of dyspnea, cyanosis and fever coinciding with a catarrh. Since 18 days before she had shown an arrested weight although at birth physical

examination had been unremarkable.

On admission she had fever (38° C). Physical examination showed a grade 4/5 pansystolic murmur and hepatomegaly. The chest X-ray showed cardiomegaly and the electrocardiogram revealed signs of right cavity dilation.

Treatment with oxygen, digital drugs and antibiotics was started resulting in a slight improvement in the patient's condition. However, 72 hours after admission sudden deterioration happened and the baby girl died.

Results

A necropsy study was carried out. There were no signs of visceral malpositions and malformations. A diffuse visceral congestion and dense lungs appeared macroscopically. We carried out a systemic opening of the heart: a 0.5 cm pediculated tumor-like lesion was found in the left ventricle which completely obstructed the subaortic space (Fig. 1). This lesion was connected to the septum just below the aortic valve without communication with heart cavities (Fig. 2). On cutting, the lesion showed a coagulated haematic content. The heart did not present any other lesion except for a discrete partial aortic coarctation of the foetal type.

Microscopically, the related lesion was formed by a single dilation with a thin fibrous-venous type wall and haematic content inside (Fig. 3).

Discussion

Varicose dilations of cardiac vessels are benign lesions of unknown incidence because they are usually postmortem findings (McAllister and Fenoglio, 1978). They are easily distinguishable from haemangiomas: the most common vascular tumors of the heart (Silverman, 1980). In the angiomas a true vascular neoformation can be observed instead of a simple dilation of preexisting vessels.

Although the varix can be found in any part of the

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Fig. 1. Varix of the heart (arrow) in the left ventricle which completely obstructs the subaortic space. L = Left ventricle. R = Right ventricle. S = Interventricular septum. Arrowheads = Partial aortic coarctation of the foetal type.

miocardium, it most often appears in the septum (Heggveit, 1966). In the case presented here, the varix was found dependent on the wall, below the aortic valve.

The location near to the valve means that it has to be differentiated from valvular blood cysts. These cysts depend on valvular surface and are comprised of a single blood-filled space and a layer of endothelial cells without basal layer (Silverman, 1980). In the case reported here the vascular dilation with haematic content had a thin fibrous wall with basal layer. The absence of elastic fibres in the wall of the cysts suggests that the pathogenesis is a diverticulum of the valve, probably as a result of a localized structural defect (Galluci et al., 1976); the varices have their origin in dilation on of thebesian veins (Begg, 1964).

Although, the lesions reported here was connected to the septum, the possibility of an aneurysm of the membranous septum with sequestered blood was abandoned because there was a fibrous-venous type wall. Moreover, no communication between vascular dilation and right cavities has been proved. The mode of presentation of cardiac varices, haemangioma and valvular blood cysts varies depending on the size and

location of the lesion. They produce symptoms, like other benign tumors, by their mass effect, embolization and local connection with the conduction system (Silverman, 1980; Rajs et al., 1983; Poole et al., 1984).

The most common complication produced by these kinds of tumors consists of cardiac out-flow tract obstruction (Galluci et al., 1976; Novitzky et al., 1984; Soberman et al., 1988). However, arrhythmias (Rajs et al., 1983) and cerebral embolisms have been reported (Albers et al., 1987).

Blood-filled cysts in the heart valves usually measure less than 1 mm, and no pathological significance is recognised, but in cases of giant blood cysts (Galluci et al., 1976) or large haemangiomas (Novitzky et al., 1984; Soberman et al., 1988) out-flow tract obstruction has been reported. In the case of cardiac varices, they appear to have no clinical significance, but in the patient reported in this paper, the varix size (0.5 cm) and location, near the aortic valve, produced left ventricular out-flow tract obstruction.

The fatal fast evolution in the patient's condition was probably due to a sharp dilation of the venous vessel.

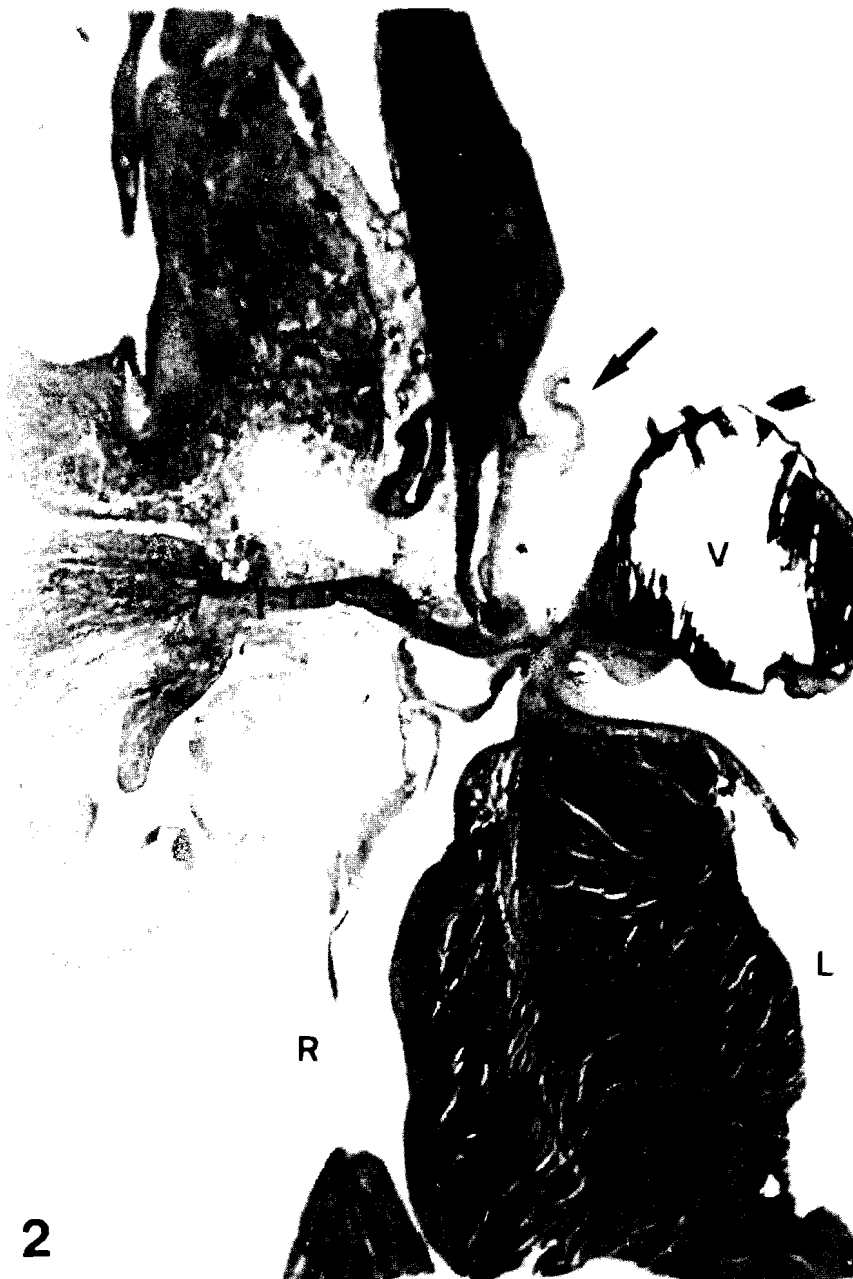


Fig. 2. Varix of the heart connected to the interventricular septum just below aortic valve. V = varix. L = left ventricle. R = Right ventricle. S = interventricular septum. A = Aortic wall. Arrow = Aortic valve. Elastin-van Gieson $\times 10$

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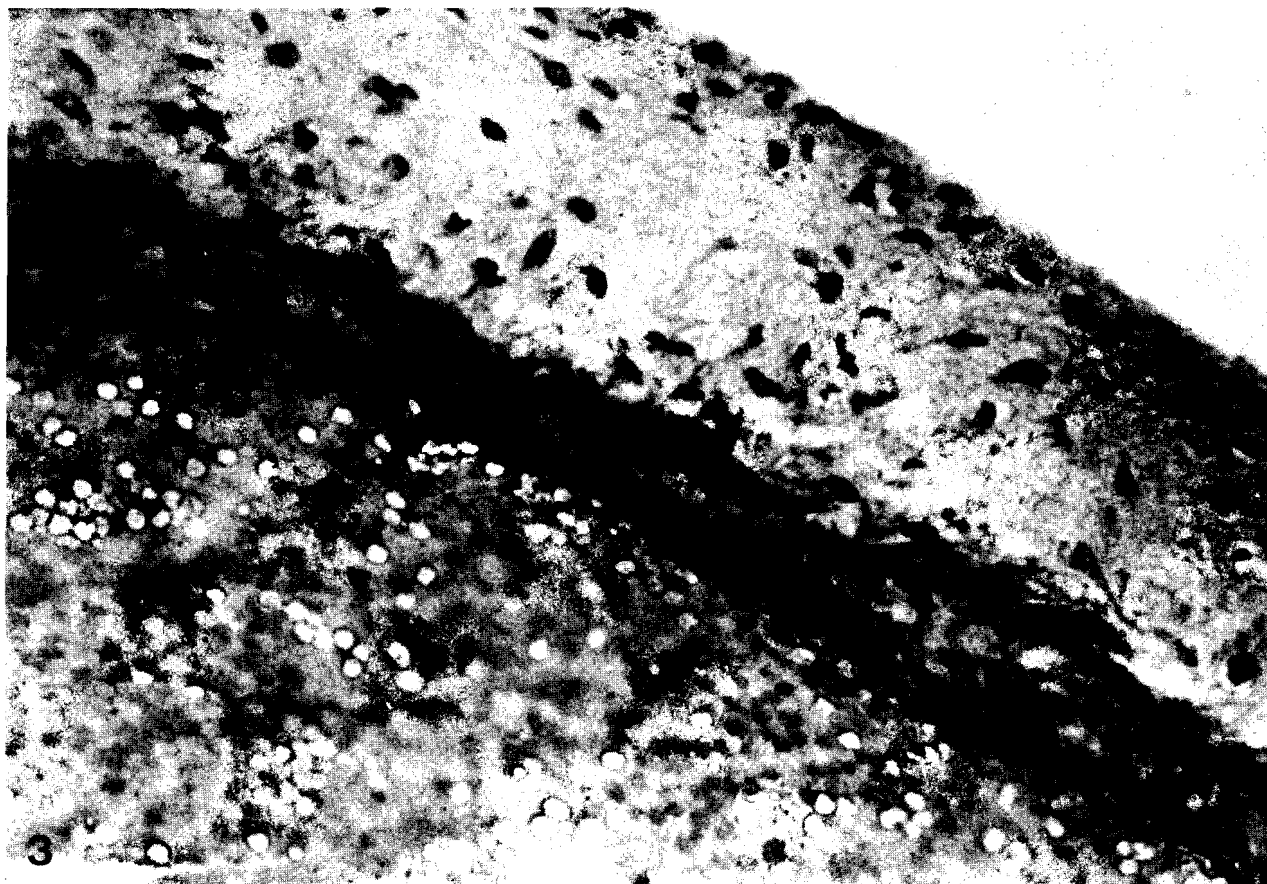


Fig. 3. Wall of the varix venous-type and vascular cavity with a haematic content. Elastin-van Gieson $\times 100$

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