Cardiac Fibroma

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The cardiac fibroma is a rare tumour of the heart which often leads to sudden death though it is benign histologically. The pathological and clinical features of the case reported here are typical of this tumour.

Case Report

A supposedly healthy girl aged 12 years collapsed suddenly after taking part in a swimming race.

Necropsy. At necropsy a tumour measuring $8 \times 7$ cm. was found to have infiltrated and replaced the apex and lower half of the left ventricle producing nodular bulging of this region of the heart (Fig. 1). On cut section it had a whorled appearance very reminiscent of a uterine fibroid.

Histology. Histologically the tumour was a fibroma. In places it was cellular, fasciculi of elongated fibroblasts with spindle-shaped nuclei criss-crossing in an irregular fashion (Fig. 2). In some areas there was a myxomatous appearance, with much loose slightly basophilic ground substance being interposed between widely separated fibroblasts (Fig. 3). The benign nature of the neoplasm was revealed by the absence of overt infiltration of cardiac muscle (Fig. 3), though in a few places small groups of muscle fibres were cut off from the main mass of myocardium. Such appearances were more consistent with the tumour showing limited local infiltration of surrounding muscle than with striated muscle forming an integral part of the neoplasm. In other areas the tumour was much less cellular, consisting of dense bands of acellular fibrous tissue showing hyalinization. Frequently these bands had become confluent to form irregular masses of hyaline collagen which stained bright red with van Gieson's stain. Such areas showed focal calcification. There was no evidence of mitotic activity.

Discussion

The cardiac fibroma is a typical benign tumour forming a well-circumscribed mass in the myocardium. It is similar to the uterine fibroid on naked-eye examination (Fig. 1).

Histologically it is composed of fibrous tissue of various degrees of cellularity. Thus, both cardiac fibromas in infants reported by Clay and Shorter (1957) were cellular, consisting of interlacing bundles of elongated cells separated by a fine meshwork of reticulin and collagen fibres. In contrast,
Fig. 2.—A cellular area of the tumour consisting of fasciculi of elongated fibroblasts with spindle-shaped nuclei. (Haematoxylin and eosin. ×250.)

Fig. 3.—This section shows the edge of the tumour. Normal myocardium is seen above and a myxomatous area of the fibroma is seen below. At the junction of the two is a thin layer of compressed mature fibrous tissue (arrow). The benign nature of the neoplasm is revealed by the absence of overt infiltration of cardiac muscle. (Haematoxylin and van Gieson. ×100.)
the tumour in a middle-aged man reported by Svejda and Tomášek (1960) was composed of massive bundles of dense collagenous tissue which compressed interposed muscle fibres. In the present case some areas of the tumour were cellular (Fig. 2) while others were acellular, hyaline, and calcified.

In some reported cases of cardiac fibroma (Clay and Shorter, 1957; Kulka, 1949; Svejda and Tomášek, 1960) cardiac muscle fibres have been described among the fibrous tissue. Hudson (1965) points out that if such muscle forms an integral part of the tumour its most appropriate designation would be "rhabdomyofibroma". In most reports, however, as in the present case, the striated muscle is described as being at the periphery of the tumour, and in common with Kulka (1949) and Clay and Shorter (1957) I favour the view that such muscle is merely normal myocardium which has been infiltrated by the periphery of the tumour. Nevertheless, it must be kept in mind that the smooth muscle of uterine fibroids tends to die out with the passage of time, so that the absence of striated muscle in the centre of this type of cardiac tumour cannot be held to be incontrovertible evidence that such muscle did not exist there originally. Other authors have preferred to call this tumour a "fibrous hamartoma" of the heart (Svejda and Tomášek, 1960; Parks, Adams, and Longmire, 1962). At all events it must be distinguished from the rhabdomyoma which is characterized by "spider-cells" packed with glycogen, and which is probably a hamartoma of Purkinje cells.

The cardiac fibroma grows slowly and produces its deleterious effects physically. It does this by infiltrating and replacing the myocardium and protruding into the cavity of the heart. The tumour usually occurs in the anterior wall of the left ventricle or the interventricular septum; the right ventricle is rarely involved. If it obstructs the left ventricular outflow tract it may give rise to an erroneous diagnosis of congenital subaortic stenosis (McCue et al., 1955; Clay and Shorter, 1957).

The cardiac fibroma usually either kills the patient or causes symptoms early in life so that most of the reported cases have been in infants or young children (McCue et al., 1955; Clay and Shorter, 1957; Parks et al., 1962; Freeman et al., 1963). An exception in this respect was the case reported in a man of 51 years by Svejda and Tomášek (1960).

Most patients with this tumour die suddenly, as in the present case. Thus, 10 of the 12 cases reviewed by Clay and Shorter (1957) had died precipitately. So had the patient reported by Freeman et al. (1963).

Very rarely attempts have been made to remove the tumours surgically. The middle-aged man reported by Svejda and Tomášek (1960) was subjected to operation for a suspected aneurysm of the left ventricle. The tumour was found instead and its superficial portion was resected; there was considerable haemorrhage and the patient died three days later. On the other hand, Parks et al. (1962) reported successful removal of a large "fibrous hamartoma" from the posterior wall of the left ventricle in a boy aged 2 years.

Summary

The case is reported of a cardiac fibroma which led to the sudden death of a supposedly healthy 12-year-old girl. At necropsy the tumour was found to have infiltrated the apical region of the left ventricle; it resembled a uterine fibroid on naked-eye examination. Histologically the tumour was benign, consisting of fibrous tissue of various degrees of cellularity in different areas.

References