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Cardiovascular tumor-like conditions

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Tumors of the heart may be neoplastic or nonneoplastic. The latter are the topic of this section and include hamartomas, heterotopias, cysts and inclusions, age-related growths, reactive proliferations, thrombotic lesions, and infections or infestations. Their importance lies not only in the fact that they can mimic true neoplastic lesions but also that they can, of their own accord, produce obstructions, arrhythmias, embolization, heart failure, and sudden death.

Hamartomas

Cardiac hamartomas represent disorganized collections of cells or tissues that are indigenous to the heart. These may include mature cardiac myocytes, fetal cardiac myoblasts, specialized Purkinje cells, or interstitial components such as blood vessels, lymphatic channels, fibroblasts, adipocytes, and others. Lesions may be well-circumscribed or poorly demarcated, but none are truly encapsulated.

Hamartomas are considered to result from faulty differentiation during embryonic development, and as such to represent congenital lesions that are present at birth. It is important to emphasize that hamartomas often involute over time and are nonneoplastic in behavior.

Cardiac rhabdomyomas

Among infants and children, rhabdomyomas represent the most common cardiac mass. Most are detected before 5 years of age, and there is a slight male preponderance. One-third of patients have tuberous sclerosis, and nearly all patients with tuberous sclerosis have cardiac rhabdomyomas.[1]

Symptoms are related to cardiomegaly, obstruction, arrhythmias, heart failure, and sudden death. The latter can develop in utero and cause fetal hydrops. For patients who survive infancy, spontaneous regression of the rhabdomyomas is common.

Lesions are multiple in 80% of the cases and are generally 0.1 to 3.0 cm in diameter. Grossly, the nodules are pale pink–tan and have a predilection for the left ventricle, although this may simply reflect the site of greatest myocardial mass. Their location can be intramural or intracavitary (Figure 1).

Microscopically, sarcoplasmic vacuolization is the most striking feature and produces classic spider cells. Vacuoles result from excessive glycogen accumulation and are best demonstrated in alcohol-fixed tissue stained with PAS. Ultrastructurally, rhabdomyoma cells resemble fetal cardiac myoblasts.[1]

Fibromas

Cardiac fibromas are also congenital lesions thought to be present at birth. Over time, they can enlarge but the growth rate is extremely slow. Less commonly, involution or even complete regression can be seen. Consequently, large fibromas generally become symptomatic during infancy or early childhood, and smaller ones are typically

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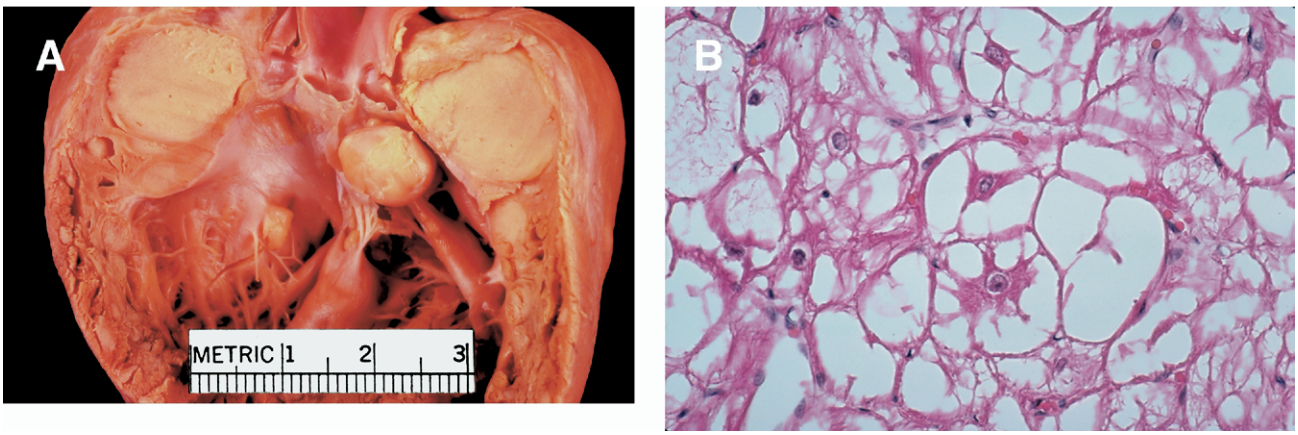

Figure 1 Cardiac rhabdomyoma. (A) Intramural and intracavitary lesions, causing left ventricular outflow tract obstruction in an infant (opened autopsy specimen). (B) Microscopy shows sarcoplasmic vacuolization and several spider cells (hematoxylin–eosin, 100×).

discovered only incidentally, sometimes late in adult life. There is no racial or gender predilection.

Cardiac fibromas primarily affect the ventricles (Figure 2A) and most commonly arise in the septum. Large fibromas in this location may be associated with appreciable cardiomegaly, chamber encroachment, heart failure, chest pain, arrhythmias, or sudden death. On cross-section, the firm white lesions appear striated or whorled and encapsulated (Figure 2B).

Microscopically, however, they resemble fibromatosis (Figure 2C). Their borders are indistinct and interdigitate with adjacent myocytes. Over time, cardiac fibromas show decreased fibroblastic cellularity and reciprocally increased collagen deposition. Although the extracellular

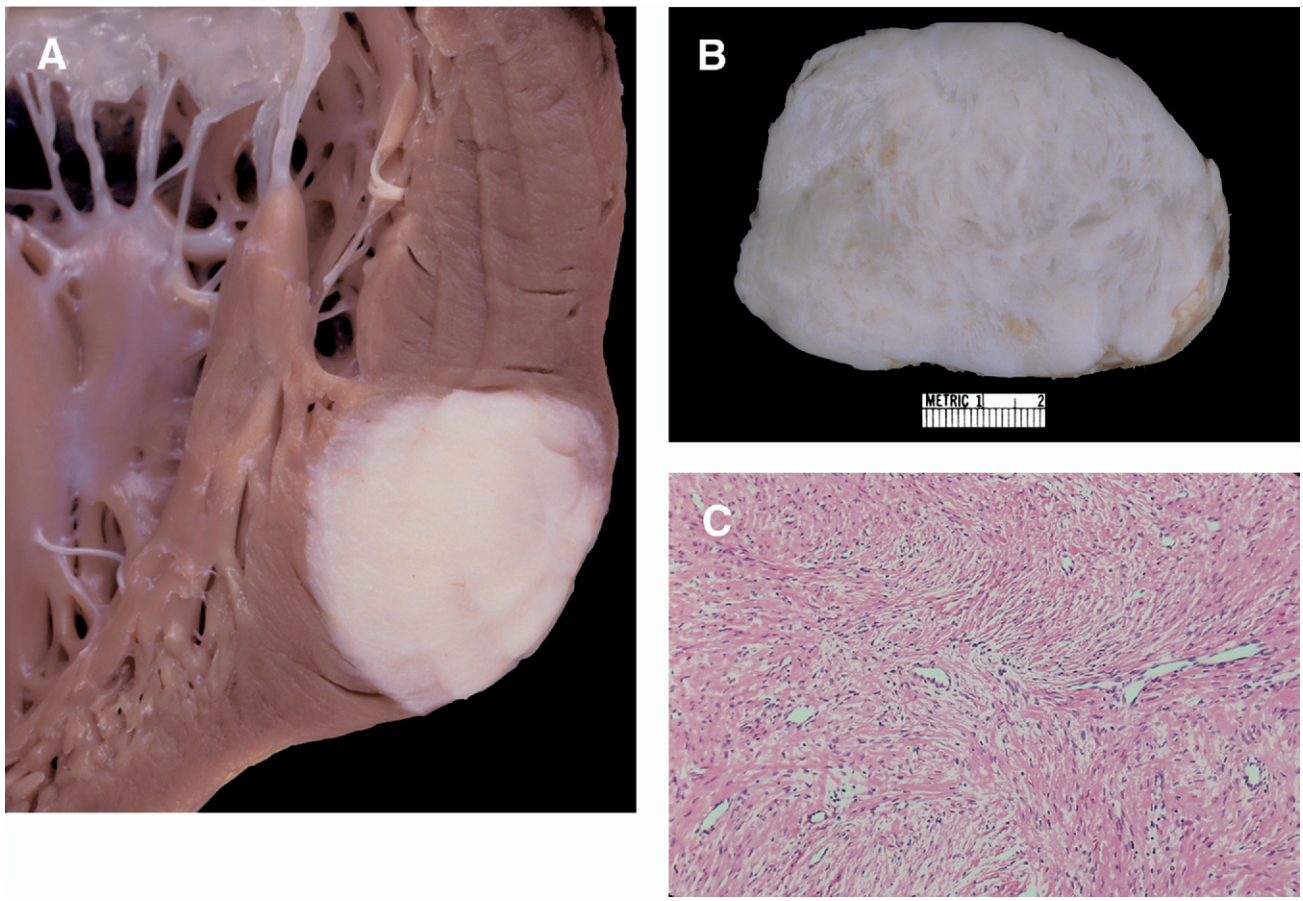

Figure 2 Cardiac fibroma. (A) Incidental tumor in the left ventricular free wall in a young adult (opened autopsy specimen). (B) Whorled cut surface, with small yellowish flecks of calcium, from a child (surgical specimen from the ventricular septum). (C) Microscopy reveals interlacing bundles of fibroblasts (hematoxylin–eosin, 25×).