Chronic or Healed Pericarditis. In some cases organization merely produces plaque-like fibrous thickenings of the serosal membranes ("soldier's plaque") or thin, delicate adhesions that rarely cause impairment of cardiac function. In other cases, fibrosis in the form of mesh-like stringy adhesions completely obliterates the pericardial sac. In most instances, this *adhesive pericarditis* has no effect on cardiac function.

Adhesive mediastinopericarditis may follow infectious pericarditis, previous cardiac surgery, or mediastinal irradiation. The pericardial sac is obliterated, and adherence of the external aspect of the parietal layer to surrounding structures strains cardiac function. With each systolic contraction, the heart pulls not only against the parietal pericardium but also against the attached surrounding structures. Systolic retraction of the rib cage and diaphragm, pulsus paradoxus, and a variety of other characteristic clinical findings may be observed. The increased workload causes occasionally severe cardiac hypertrophy and dilation.

In *constrictive pericarditis* the heart is encased in a dense, fibrous or fibrocalcific scar that limits diastolic expansion and cardiac output, features that mimic a restrictive cardiomyopathy. A prior history of pericarditis may or may not be present. The fibrous scar can be up to a centimeter in thickness, obliterating the pericardial space and sometimes calcifying; in extreme cases it can resemble a plaster mold *(concretio cordis)*. Because of the dense enclosing scar, cardiac hypertrophy and dilation cannot occur. Cardiac output may be reduced at rest, but more importantly the heart has little if any capacity to increase its output in response to increased systemic demands. Signs of constrictive pericarditis include distant or muffled heart sounds, elevated jugular venous pressure, and peripheral edema. Treatment consists of surgical resection of the shell of constricting fibrous tissue (pericardiectomy).

Heart Disease Associated with Rheumatologic Disorders

The heart (vessels, myocardium, valves, or pericardium) can be significantly impacted by chronic rheumatologic diseases (e.g., rheumatoid arthritis, SLE, systemic sclerosis, ankylosing spondylitis, and psoriatic arthritis). Indeed, as improved therapies lead to longer life expectancies, the cardiovascular manifestations of such systemic inflammation are increasingly recognized. In addition, ischemic heart disease can be accelerated in the setting of systemic inflammation.

Although rheumatoid arthritis is primarily a joint disorder, it also has several extra-articular manifestations, including subcutaneous rheumatoid nodules, vasculitis, and neutropenia (Chapter 26). The heart is also involved in 20-40% of severe cases. The most common finding is a *fibrinous pericarditis* that may progress to fibrous thickening of the visceral and parietal pericardium and dense adhesions. Granulomatous rheumatoid nodules resembling the subcutaneous nodules may also occur in the myocardium, endocardium, valves, and aortic root. *Rheumatoid valvulitis* can lead to marked fibrous thickening and secondary calcification of the aortic valve cusps, producing changes resembling those of chronic rheumatic valvular disease. The Libman-Sacks valvular lesions associated with SLE were discussed previously.

Tumors of the Heart

Primary tumors of the heart are rare; in contrast, metastatic tumors to the heart occur in about 5% of persons dying of cancer. The most common primary cardiac tumors, in descending order of frequency (overall, including adults and children) are myxomas, fibromas, lipomas, papillary fibroelastomas, rhabdomyomas, and angiosarcomas. The five most common tumors are all benign and collectively account for 80% to 90% of primary tumors of the heart.

Primary Cardiac Tumors

Myxomas are the most common primary tumor of the adult heart (Fig. 12-38). These are benign neoplasms thought to arise from primitive multipotent mesenchymal cells. Although sporadic myxomas do not show consistent genetic alterations, familial syndromes associated with myxomas have activating mutations in the *GNAS1* gene, encoding a subunit of G protein (Gsα) (in association with McCune-Albright syndrome) or null mutations in *PRKAR1A*, encoding a regulatory subunit of a cyclic-AMP-dependent protein kinase (*Carney complex*). About 90% of myxomas arise in the atria, with a left-to-right ratio of approximately 4:1.

MORPHOLOGY

The tumors are usually single, but can rarely be multiple. The region of the fossa ovalis in the atrial septum is the favored site of origin. Myxomas range from small (<1 cm) to large (≥10 cm), and can be sessile or pedunculated lesions (Fig. 12-38*A*). They vary from globular hard masses mottled with hemorrhage to soft, translucent, papillary, or villous lesions having a gelatinous appearance. The pedunculated form is often sufficiently mobile to move during systole into the atrioventricular valve

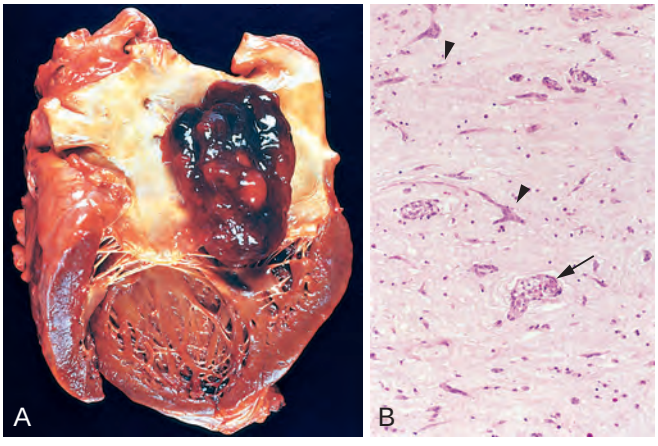

Figure 12-38 Atrial myxoma. **A,** A large pedunculated lesion arises from the region of the fossa ovalis and extends into the mitral valve orifice. **B,** Abundant amorphous extracellular matrix contains scattered multinucleate myxoma cells (*arrowheads*) in various groupings, including abnormal vessel-like formations *(arrow)*.