Case #63

NAME Educational Activities Committee
Case submitted by: Dr. Nicole Croom, Assistant ME at WCMEO
Question crafted by: Dr. Melanie Bois, Cardiovascular Pathologist, Mayo Clinic
Histology photos courtesy of: Drs. Bois and Quinton, Mayo Clinic
1. This is a heart from an autopsy of a 53-year-old woman found unresponsive at home. Histologic sections of the area highlighted would most likely show:

- Non-caseating granulomas and multinucleated giant cells with occasional asteroid bodies
- Confluent mature adipocytes with nuclear hyperchromasia admixed with lipoblasts
- Unencapsulated mixture of mature adipocytes, multivacuolated fetal (brown) fat, and cardiomyocytes
- “Zellballen”, nested epithelioid cells surrounded by conspicuous fibrous stroma
- Confluent lymphoid cells with frequent mitoses and apoptotic debris
Answer...
C. Unencapsulated mixture of mature adipocytes, multivacuolated fetal (brown) fat, and cardiomyocytes (59.18% responses)

This is a microscopic description of lipomatous hypertrophy of the interatrial septum (LHAS) and corresponds to the observed macroscopic findings of “massive” (>2 cm thick) accumulation of fat in the interatrial septum. LHAS is a benign lesion with prevalence of 2.2% to 8% depending on the diagnostic modality. Patients are typically asymptomatic; however, they can experience atrial arrhythmias which, even more rarely, can induce sudden cardiac death. Unlike cardiac lipomas, LHAS is associated with older age, obesity, and female anatomical sex.
Other Responses -
A. Non-caseating granulomas and multinucleated giant cells with occasional asteroid bodies (12.96% responses)

This is a microscopic description of cardiac sarcoidosis (CS). The histologic features of cardiac sarcoid overlap with giant cell myocarditis (GCM) and there has been much debate as to whether CM and GCM are two distinct entities or represent a spectrum of a single disease. One study found that, after overreading of all available histological material by cardiac pathologists, 62% of GCM cases were reclassified as CS. These reclassifications were based on the identification of typical sarcoid granulomas that had either been misread or overlooked or were found in subsequent specimens. Overall, the repeatedly demonstrated differing clinical outcomes and responses to treatment provide evidence that GCM and CS are separate entities. GCM tends to have a much more acute presentation and worse prognosis with (fulminant) heart failure as the initial presentation. Sarcoidosis is a multisystem granulomatous disorder, with less than 10% of patients having cardiac involvement. In contrast to GCM, patients tend to have a more chronic presentation with insidious onset of restrictive (infiltrative) cardiomyopathy. Macroscopically, CS is characterized by yellow, white, or gray myocardial infiltrates with indistinct borders.
B. Confluent mature adipocytes with nuclear hyperchromasia admixed with lipoblasts (19.44% responses)

This is a microscopic description of a liposarcoma. Cardiac liposarcoma accounts for approximately 1% of all primary malignant cardiac tumors (“Fun” Fact: the most common type of cardiac sarcoma is angiosarcoma). Like cardiac lipomas, cardiac liposarcoma has no gender predominance and most occur in the right atrium. Cardiac liposarcoma tends to invade adjacent organs, and most clinical signs/symptoms are related to compression of those organs. Macroscopically, cardiac liposarcoma is similar to liposarcomas in other regions of the body: large, fatty yellow to firm white mass with areas of hemorrhage and necrosis depending on the grade
D. “Zellballen,” nested epithelioid cells surrounded by conspicuous fibrous stroma (5% responses)

This is a microscopic description of a cardiac paraganglioma (PGL), a rare, typically benign neuroendocrine tumor with significant morbidity due to (1) norepinephrine secretion leading to severe hypertension, palpitations, syncope, lethal tachyarrhythmias, and stroke and/or (2) compression of cardiovascular structures, depending on tumor size and location. Cardiac PGLs can originate from the visceral autonomic (atrium or interatrial septum) paraganglia or the branchiomeric (coronary, pulmonary, and aorticopulmonary) paraganglia. Macroscopically, these tumors present as rubbery nodules with homogeneous pink-gray parenchyma surrounded by a thin capsule.
E. Confluent lymphoid cells with frequent mitoses and apoptotic debris (3.52% responses)

This is a microscopic description of a cardiac lymphoma, a rare cardiac tumor. The histologic image above is actually a cardiac plasmacytoma, which can manifest as a solitary plasma-cell tumor (primary) or as part of multiple-myeloma disease (secondary), and the heart can be involved in either. Macroscopically, cardiac plasmacytoma typically presents as a large space-occupying tumor involving one or both atria. Pericardial effusion is the most common presentation.


