Case #126

NAME Educational Activities Committee

Case provided by:

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Oklahoma City, OK

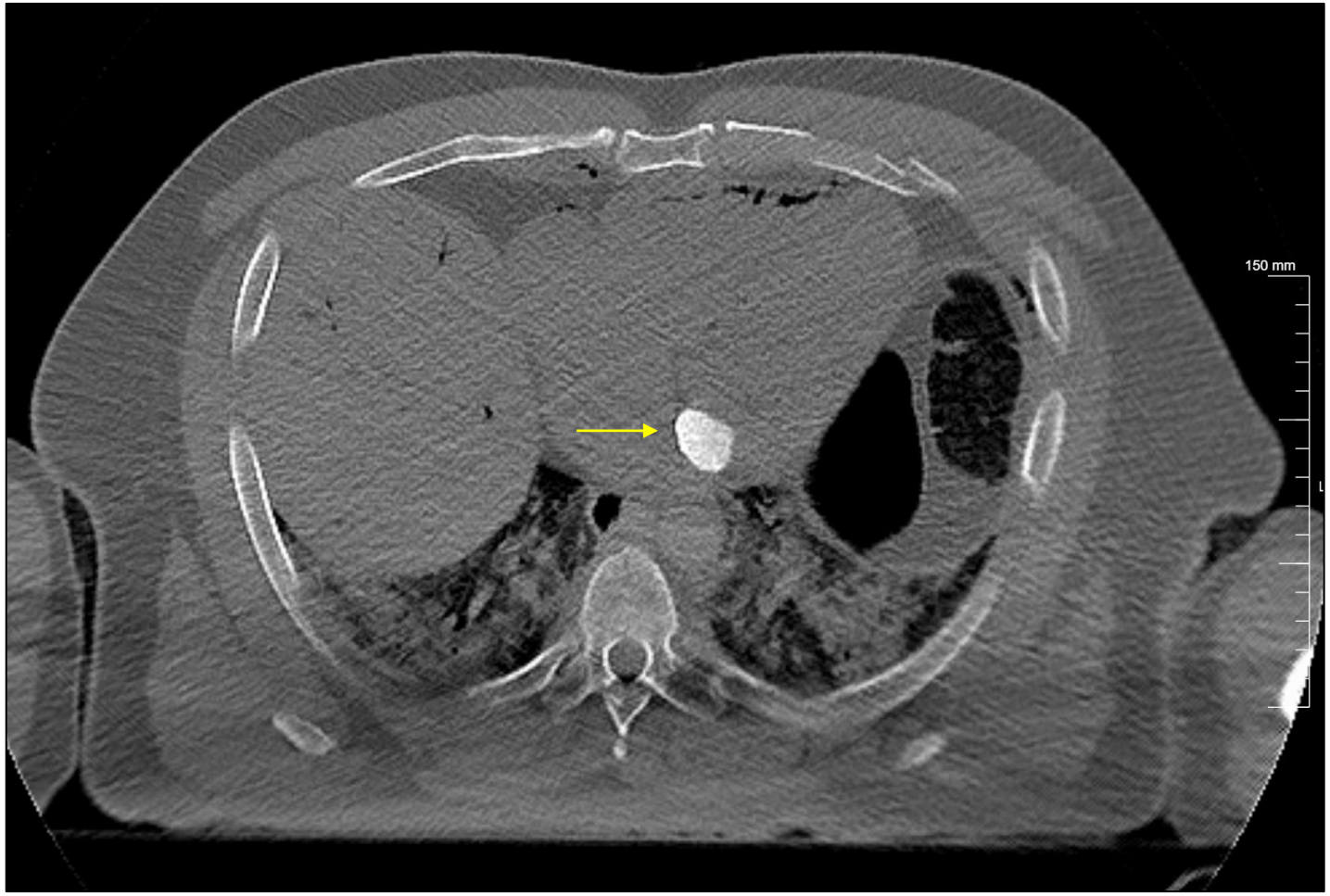
1. A 63-year-old man with a medical history of hypertension, hyperlipidemia, and coronary artery disease was witnessed to collapse while coaching football. The recorded outdoor temperature was 87 °F. Emergency medical services were called, and cardiopulmonary resuscitation was attempted, however, resuscitative efforts were ultimately unsuccessful.

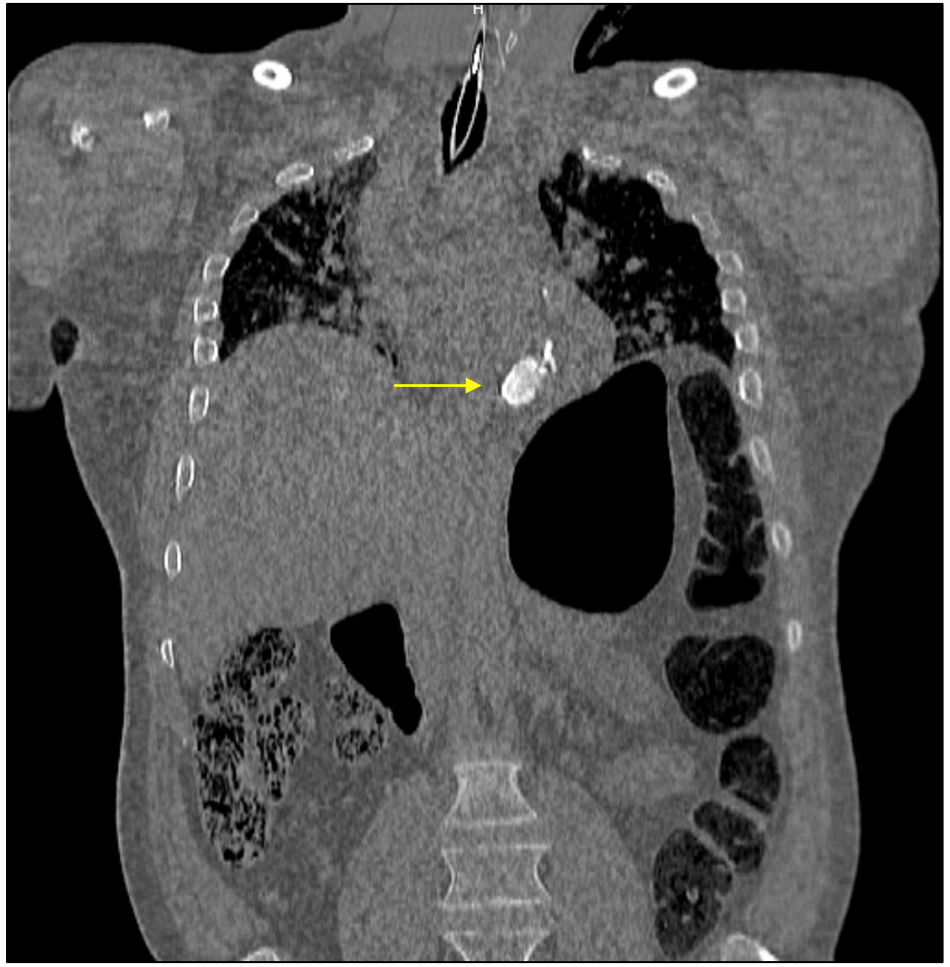
A postmortem computed (CT) scan revealed an oval to round hyperdense nodule near the mitral valve. Autopsy showed the following gross and subsequent microscopic findings.

What is the most likely diagnosis for the observed cardiac findings in this case?

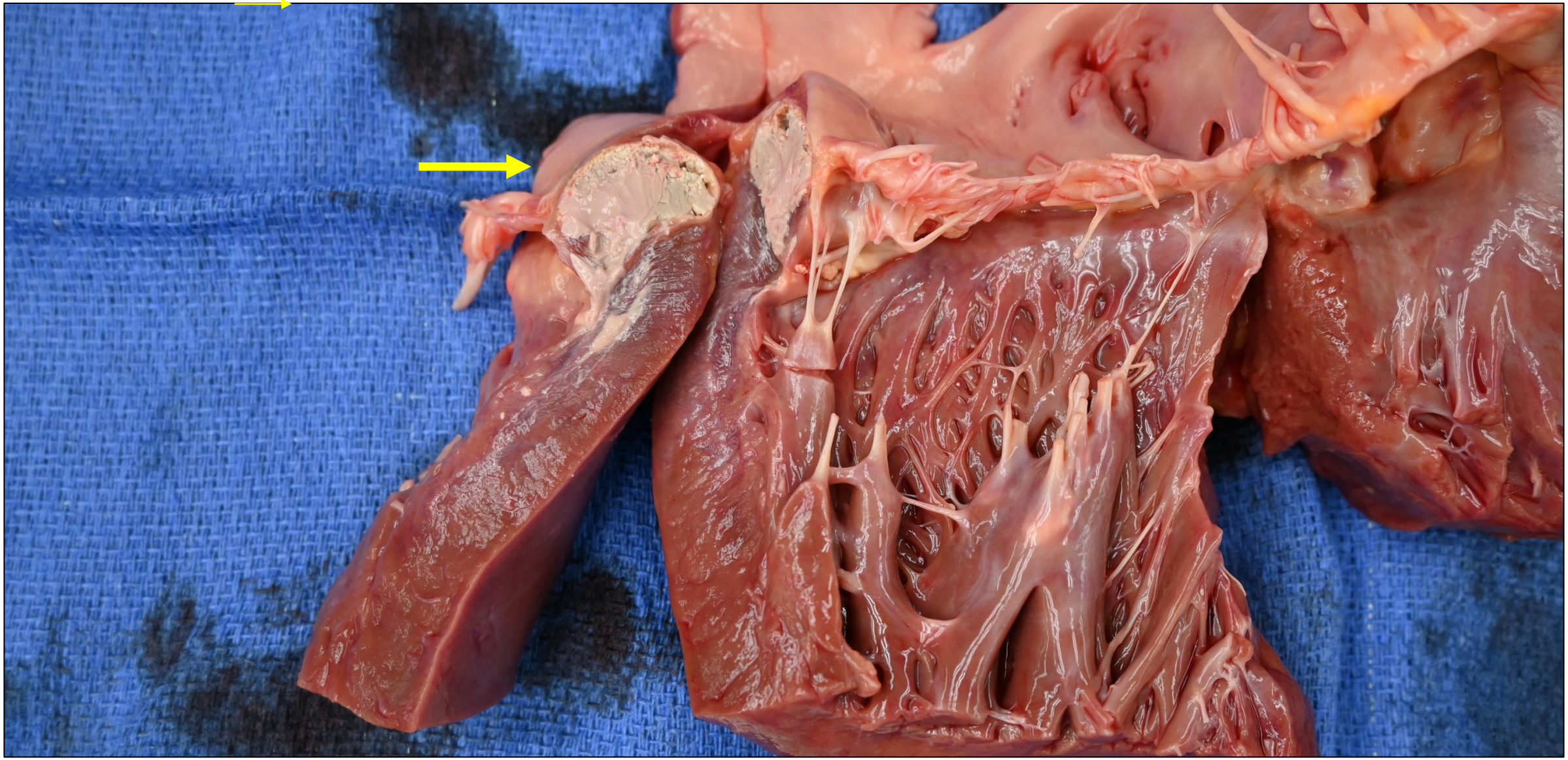
- ◯ Myxoma
- ◯ Hydatid Cyst
- ◯ Infective Endocarditis with abscess
- ◯ Caseous calcification of mitral annulus
- ◯ Cystic tumor of the atrioventricular nodal region

Postmortem Body CT (Axial and Coronal)

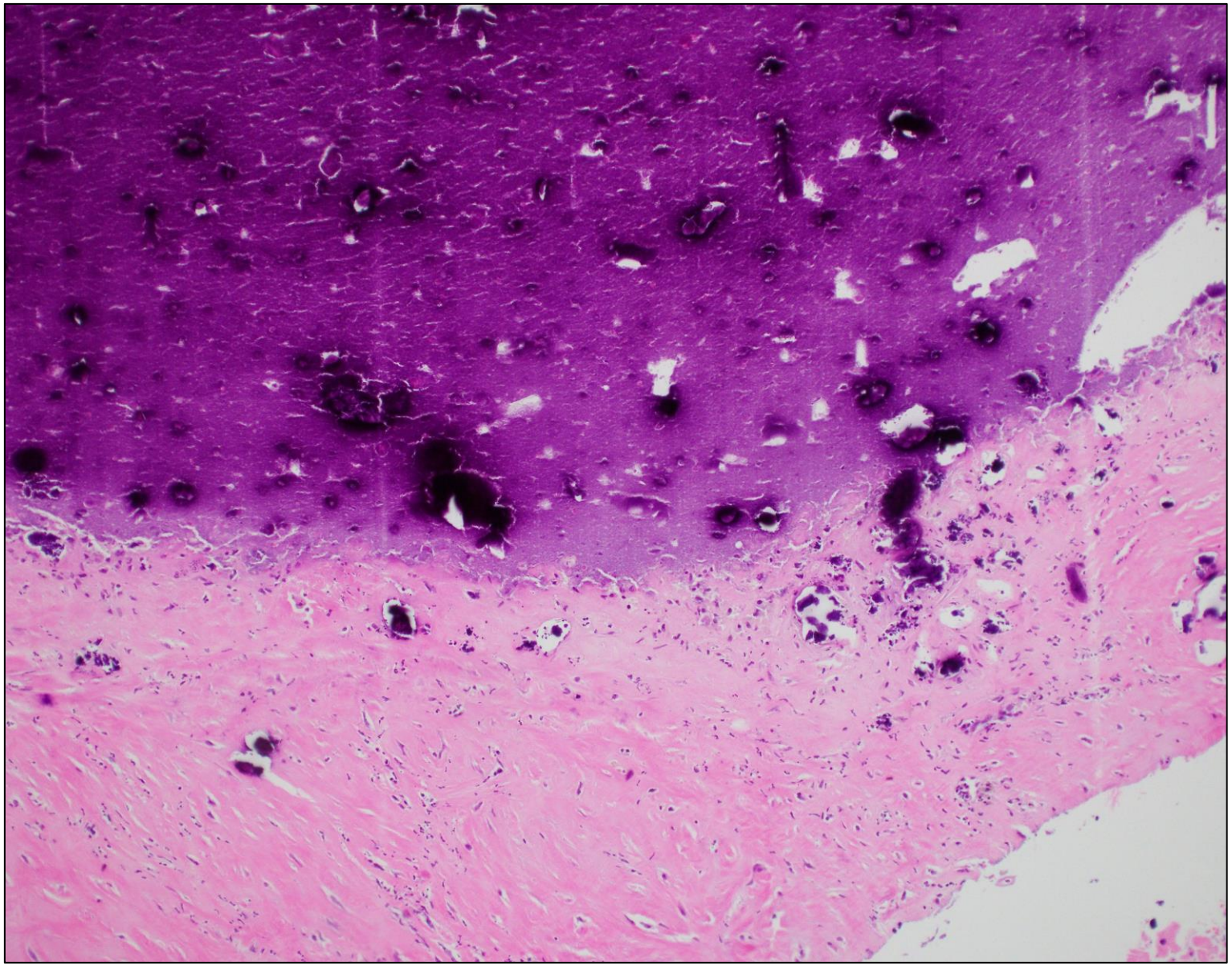




Autopsy Findings:



Microscopic Findings:



Answer.....

(D) Caseous calcification of mitral annulus (CORRECT ANSWER, 83.00 % of responses)

This case involves a caseous calcification of the mitral annulus (CCMA) which refers to a calcified cardiac mass and a rare variant of a mitral annular calcification. CCMA is a classic tumor mimicker, often mistaken for myxoma or abscess both radiologically and grossly. Most commonly, it occurs in individuals > 60 years and is associated with end-stage renal disease, hypertension, hypercholesterolemia, and atrial fibrillation.

CCMAs characteristically consist of a liquefactive, caseous, milky substance (often described as “toothpaste-like”) that is enveloped within a calcific, fibrous shell. They’re most often found in the periannular region of the ventricular or atrial side of the mitral annulus adjacent to the posterior leaflet.

Microscopically, CCMAs consist of abundant amorphous basophilic material with myxoid areas, hyaline degeneration, calcifications, and zones of necrosis with surrounding inflammatory infiltrates usually involving macrophages and lymphocytes.

The exact etiology and pathogenesis is still unknown. Some hypotheses postulate that the development of CCMAs is a direct result from altered calcium phosphate metabolism.

These “toothpaste tumors” overall are considered benign entities that can resolve spontaneously on their own or transform back into a normal mitral annular calcification. If these tumors elicit mass effect by impeding ventricular filling of chamber blood, they can be surgically excised. However, in the absence of emboli, valve dysfunction, or conduction system involvement most of these tumors are found incidentally at autopsy in approximately ~2.7% of cases involving mitral annular calcification.

Other responses:

(A) Myxoma (7.28 % of responses)

Myxomas are the most common primary tumor of the heart and arise most often in the left atrium with attachment to the atrial septum near the foramen ovale. They can occur in the sporadic or familial setting and are associated with the heritable Carney complex (*PRKAR1A* gene mutation). Grossly, myxomas are soft, polypoid, lobulated masses and can often have areas of hemorrhage or calcification. Microscopically, the tumor cells are round, polygonal, or stellate surrounded by an abundant myxoid stroma rich in acid mucopolysaccharides. The hallmark microscopic findings for these tumors are tumor cells that show a concentric arrangement around vessels.

In rare cases, myxomas can become extremely calcified and are referred to as “petrified” myomas. However, the location and microscopic findings are not supportive of a myxoma for this case.

(B) Hydatid Cyst (2.43 % of responses)

Hydatid disease is caused by a cestode (tapeworm) that occurs on all continents with *Echinococcus granulosus* being the most frequent. The pathophysiology involves mucosal attachment of an adult tapeworm to the small intestine of the definitive host (i.e., dogs or jackals). Embryonated eggs are shed in the feces and then ingested by the intermediate host where the eggs hatch and the larva oncospheres pass to the liver via the portal vein. Subsequently, in some cases, the larva oncospheres can enter the heart through the liver and inferior vena cava with cardiac involvement occurring through the invasion of the myocardium primarily through the coronary artery circulation. The left ventricle is the most frequently involved (55–60% of cases), followed by the right ventricle (15%) and interventricular septum (5–9%).

Microscopically, the cyst wall has 3 structural components: outer acellular laminated membrane, inner germinal membrane, and protoscolices (hooklets are birefringent under polarized light).

(C) Infective Endocarditis with abscess (3.31 % of responses)

Infective endocarditis with abscess formation is a complication of bacterial infection involving the endocardial surface, most commonly affecting the heart valves. It is frequently seen in individuals with pre-existing valvular heart disease, prosthetic valves, or a history of intravenous drug use. These conditions predispose to endothelial damage, allowing bacterial colonization and vegetation formation. Grossly, the affected valve may show friable, tan-white vegetations and surrounding areas of erosion or tissue destruction. An abscess typically appears as a localized area of necrosis or purulence, often in the valve ring or annular region, and may extend into adjacent myocardium.

Microscopically, vegetations consist of fibrin, acute inflammatory cells, and bacterial colonies. Abscesses show central necrosis with dense neutrophilic infiltration and tissue destruction. Involvement of the conduction system or annular extension can lead to heart block, heart failure, or sudden death.

(E.) Cystic tumor of the atrioventricular nodal region (3.97 % of responses)

Cystic tumor of the atrioventricular nodal region (CTAVNR) is a rare tumor that was historically regarded as a mesothelioma. It is now understood that these tumors are not mesothelial in origin and may represent a congenital nests of endodermal origin or ultimobranchial heterotopia analogous to solid nests of thyroid gland.

Grossly, they may appear as a multicystic lesion in the area of the atrioventricular (AV) node and membranous septum. Microscopically, these tumors consist of ductular structures, cysts, and solid nests of epithelial appearing cells. Diagnosis requires histologic sampling of the conduction system. Due to its critical location near the AV node, CTAVNR is often associated with complete heart block and may lead to sudden death.

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