Case #157

NAME Educational Activities Committee

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A 44-year-old man was found unresponsive in an outdoor public area, leaning against a wall. His medical history was notable for chronic alcohol use and previous surgical treatment of a hepatic lesion, for which the histopathologic diagnosis was not available. No recent cardiac symptoms were documented.

A medicolegal autopsy was performed. External examination demonstrated several minor superficial abrasions without significant traumatic injury. Examination of the heart revealed a prominent, rounded bulge involving the right ventricular wall (**Figure 1**). Sectioning revealed a well-circumscribed intramyocardial lesion, measuring 7.0 × 4.5 × 4.5 cm, with a thick, partially calcified wall and friable necrotic contents, filling more than half of the right ventricular cavity and extending toward the left ventricle (**Figures 2 and 3**). The cardiac valves and remaining myocardium were grossly unremarkable.

No significant alternative cause of death was identified, and death was attributed to the cardiac lesion. A representative histologic section is shown (**Figure 4**).

What is the most likely diagnosis?

A. Calcified cardiac fibroma

B. Cardiac hydatid cyst

C. Calcified amorphous tumor of the heart

D. Cardiac cavernous hemangioma with dystrophic calcification

Gross Heart, External Examination

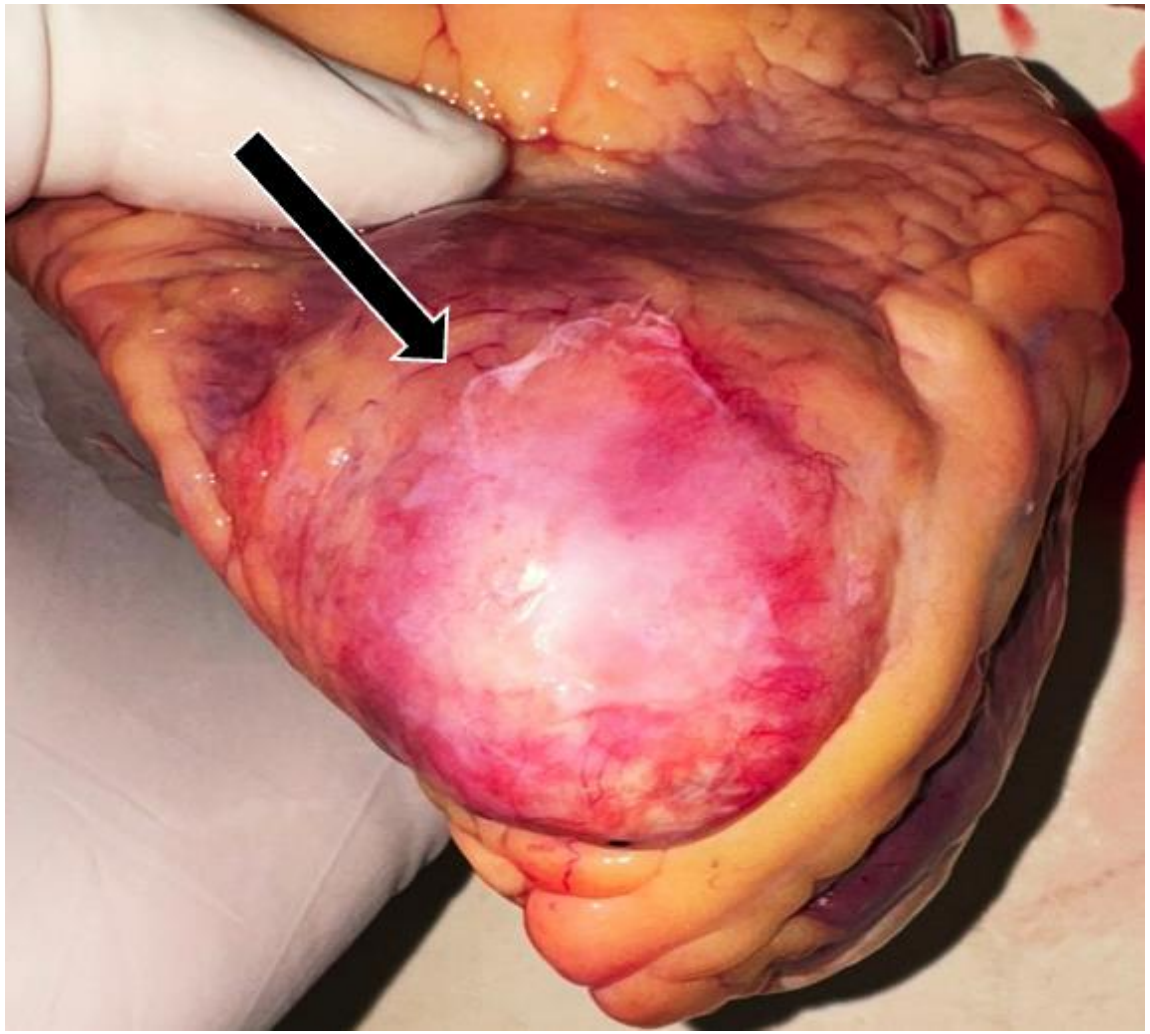


Figure 1. Prominent external bulging of the right ventricle (black arrow)

Intramyocardial Lesion, Internal Examination

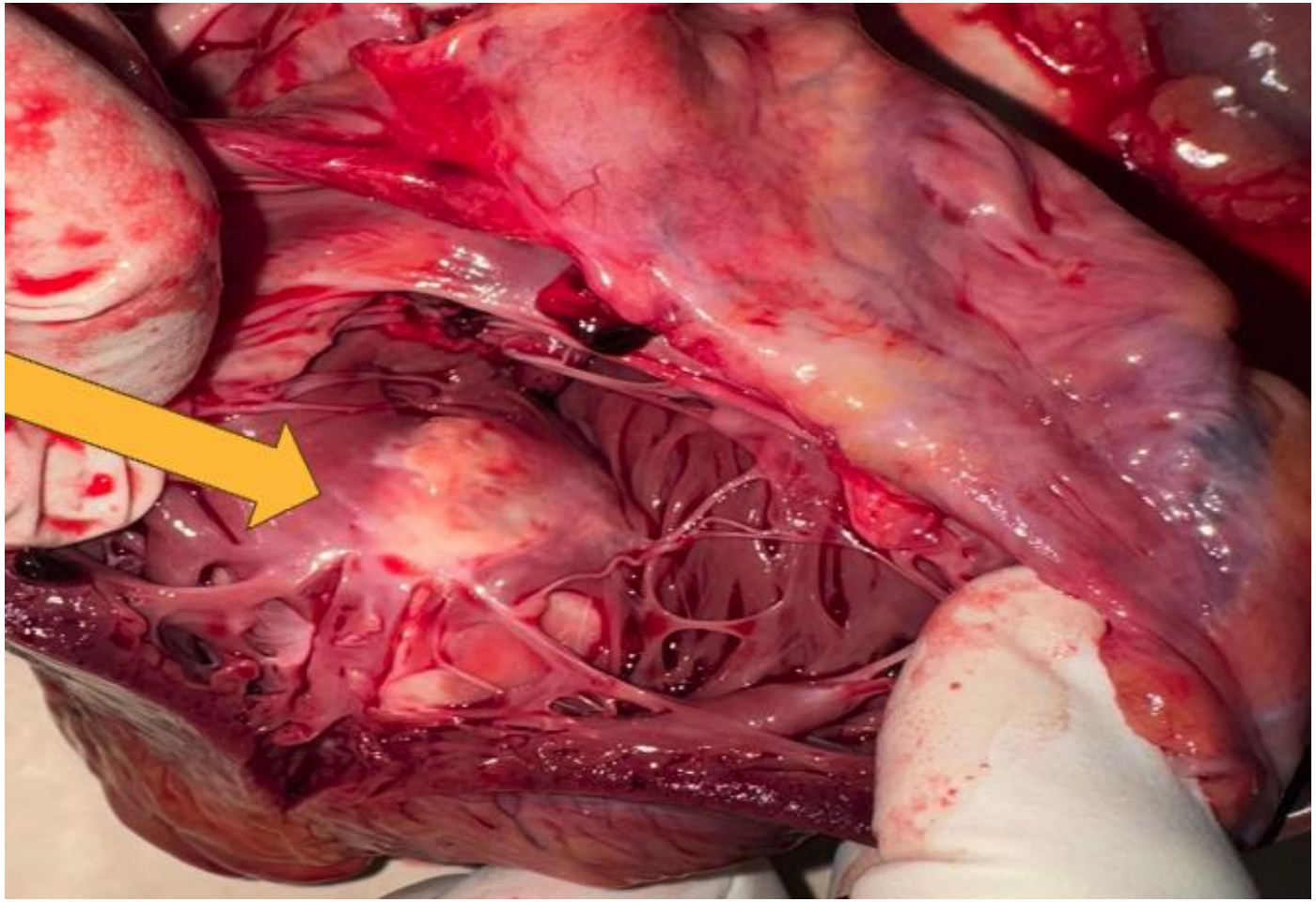


Figure 2. Intramyocardial lesion projecting into the right ventricular cavity (yellow arrow)

Intramyocardial Lesion, Internal Examination

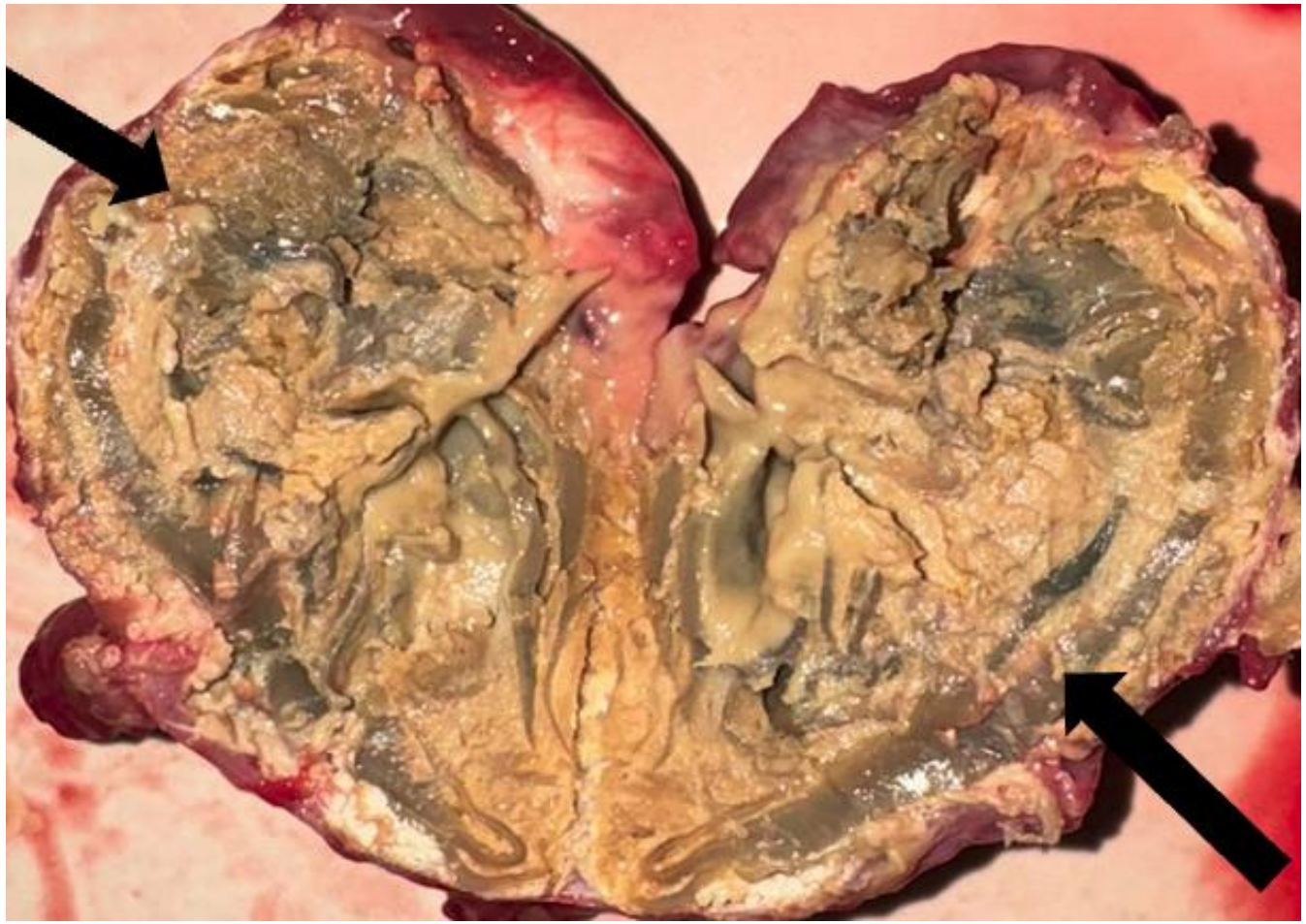
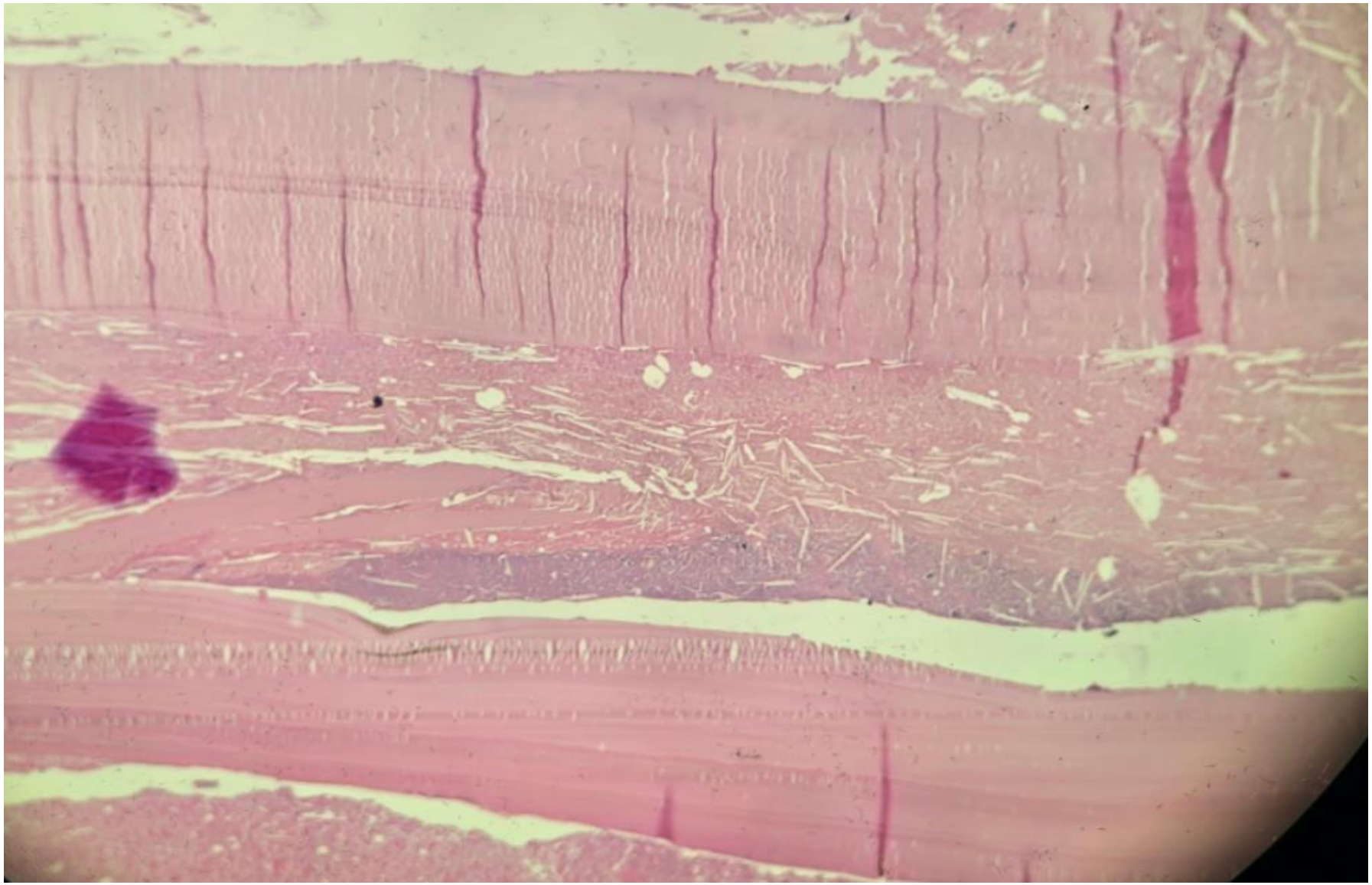


Figure 3. Calcified necrotic-hemorrhagic material within a myocardial lesion (black arrows)

Figure 4: Microscopic Examination

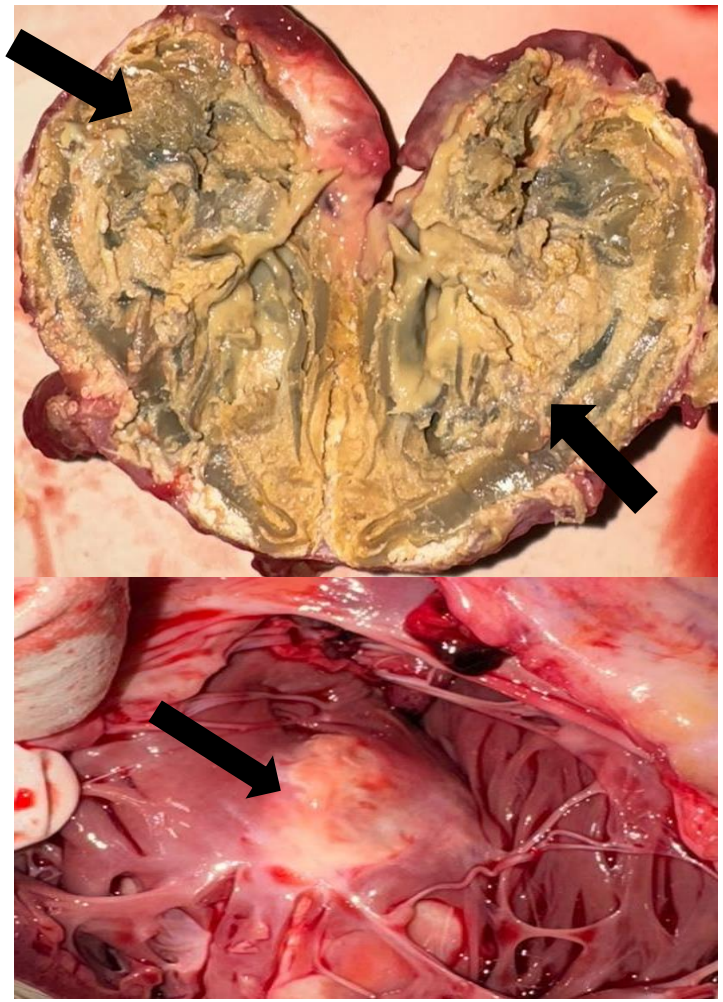


Answer...

Correct Answer: B. Cardiac hydatid cyst (58.47%)

Several findings converge on this diagnosis. At autopsy, the lesion is a large, well-defined intramyocardial cystic process centered in the right ventricle, with a firm, calcified wall and sufficient size to occupy more than half of the right ventricular cavity and extend toward the left ventricle. The patient's previous surgical treatment of a hepatic lesion is an important historical finding; however, because the histopathologic diagnosis of that lesion is unavailable, it cannot be definitively attributed to echinococcosis.

The geographic setting is also epidemiologically compatible with the diagnosis. Cystic echinococcosis is endemic in Morocco and remains an important zoonotic disease in the country[1]. A community-based ultrasound study conducted in endemic rural areas of the Middle Atlas reported an overall prevalence of abdominal cystic echinococcosis of 1.9%2. Nevertheless, cardiac involvement is exceptionally uncommon, accounting for approximately 0.02–2% of cystic echinococcosis cases[3].

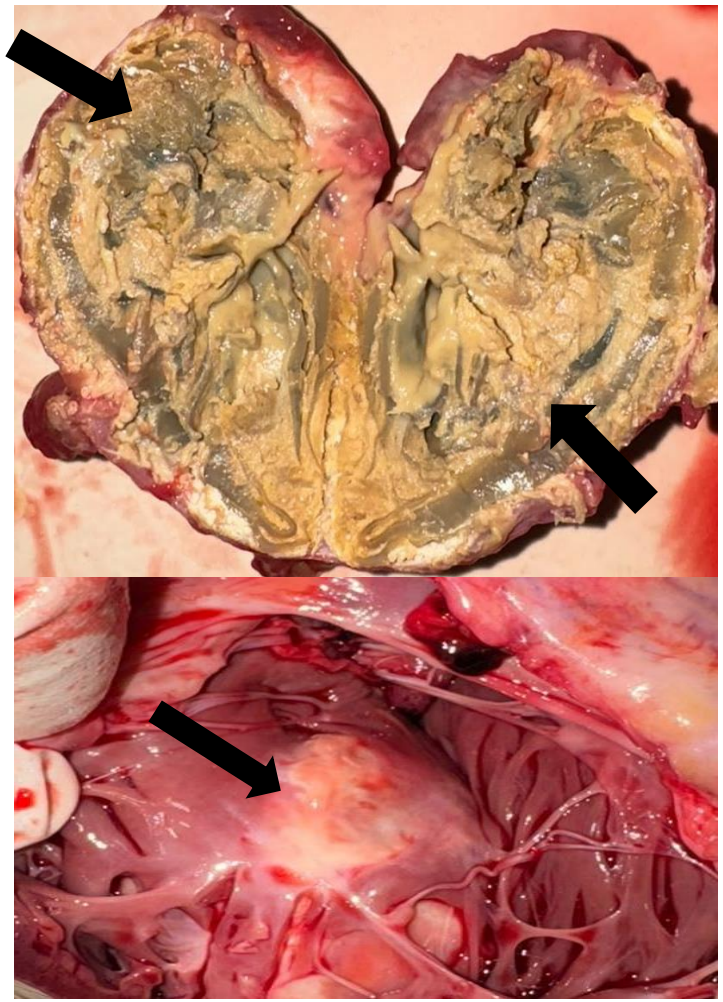


Intramyocardial lesion projecting into the right ventricular cavity, with Calcified necrotic-hemorrhagic material within a myocardial lesion

Correct Answer: B. Cardiac hydatid cyst

The decisive finding in the present case is microscopic: the cyst is surrounded by a fibrous host reaction with mixed inflammatory cells and contains necrotic-hemorrhagic material and focal calcifications, together with a thick eosinophilic, laminated, acellular membrane. This laminated membrane is a characteristic structural component of an Echinococcus hydatid cyst; CDC4 reference material similarly describes the hydatid cyst wall as containing a prominent acellular laminated layer surrounded by host tissue. Calcification does not argue against hydatid disease, particularly in a chronic or degenerating cyst.

Comparable autopsy cases of cardiac hydatidosis have demonstrated large right ventricular cysts with external ventricular bulging and histologic identification of the laminated acellular layer, with sudden death attributed to cardiac arrhythmia when no rupture or pulmonary hydatid embolism was identified[5].

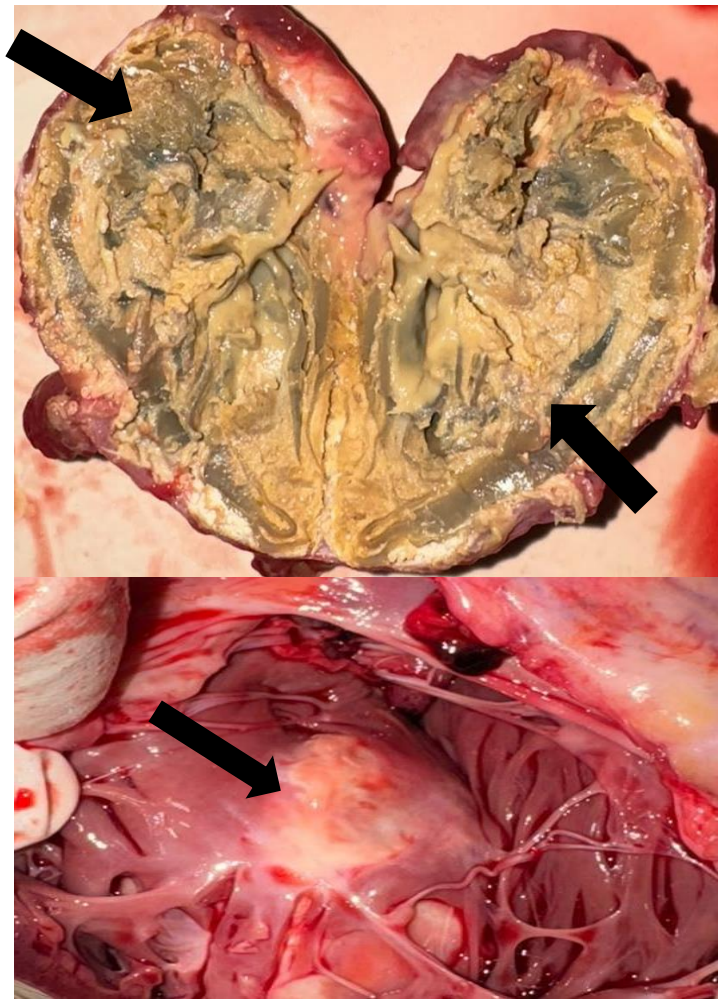


Intramyocardial lesion projecting into the right ventricular cavity, with Calcified necrotic-hemorrhagic material within a myocardial lesion

Correct Answer: B. Cardiac hydatid cyst

In the present case, the large size of the lesion and its intramyocardial right ventricular location could also have adversely affected cardiac function through marked distortion of the ventricular cavity and myocardial involvement, potentially providing an arrhythmogenic substrate. In the absence of rupture or pulmonary hydatid embolism, a fatal ventricular arrhythmia related to the cyst represents a plausible immediate mechanism of sudden death, although such an arrhythmia cannot be directly demonstrated postmortem.

Thus, the gross cystic architecture, chronic calcification, characteristic laminated membrane, and associated host inflammatory and fibrous response establish the diagnosis of a cardiac hydatid cyst.



Intramyocardial lesion projecting into the right ventricular cavity, with Calcified necrotic-hemorrhagic material within a myocardial lesion

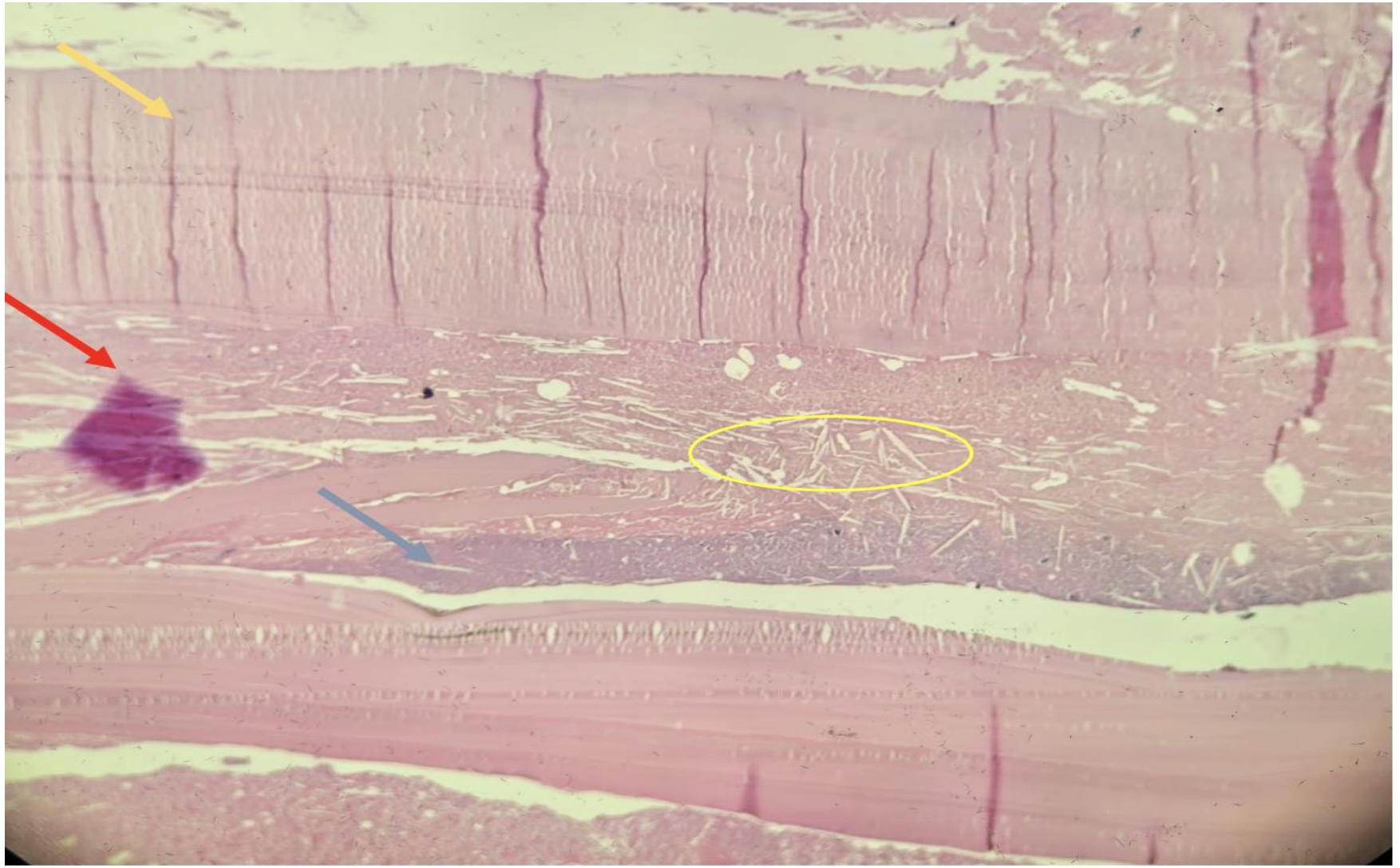


Figure 4. Histological features of a calcified and degenerative hydatid cyst (H&E stain) **(×20):** An outer acellular eosinophilic laminated membrane is identified (**yellow arrow**), bordered by a reactive fibrous pericyst (**red arrow**). The pericyst shows inflammatory changes (**blue arrow**) and clear spaces corresponding to cholesterol clefts (**yellow circle**).

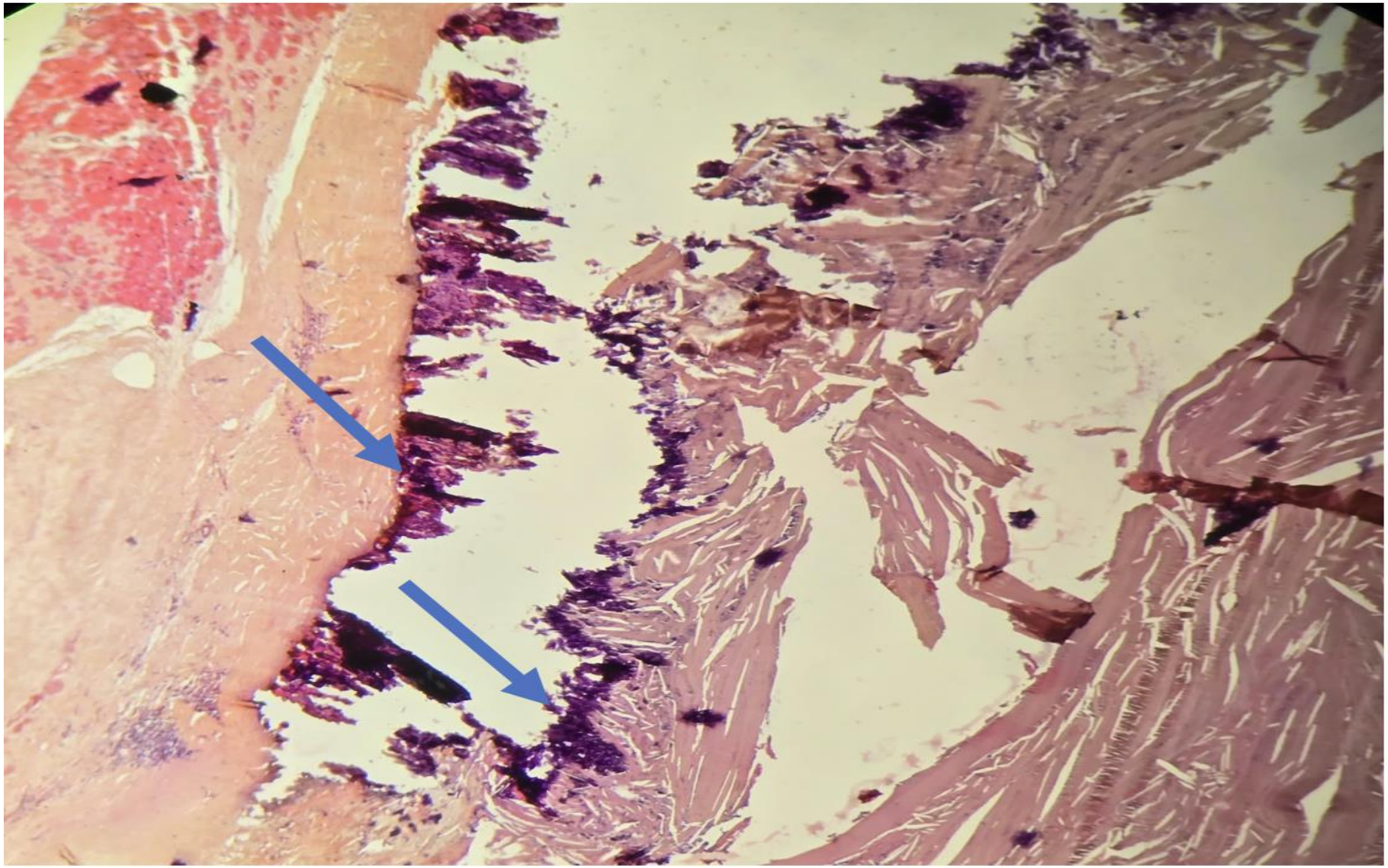


Figure 5. Histological features of a calcified and degenerative hydatid cyst (H&E stain) **(×40):** Dense basophilic dystrophic calcifications (**blue arrow**) are present adjacent to the laminated membrane, reflecting the chronicity of the lesion.

Other Responses...

A. Calcified cardiac fibroma (13.71%)

A calcified cardiac fibroma is a reasonable gross differential because fibromas arise within the ventricular myocardium, may be well circumscribed, can become heavily calcified, and may produce ventricular arrhythmias or sudden death. However, its fundamental morphology is different from that of the lesion in this case. Cardiac fibroma is a solid benign mesenchymal tumor, typically appearing grossly as a firm gray-white or tan-white, often whorled intramyocardial mass rather than a true cystic structure[6]. Microscopically, it is composed of relatively bland spindle-shaped fibroblasts[7] (**Figure X**), embedded within abundant dense collagen, with variable dystrophic calcification; older lesions may become particularly collagenized and hypocellular[6] (**Figure Z**). Although the firm consistency and calcification in the present case could initially suggest fibroma, the expected fibroblastic/collagenous tumor architecture is absent. Instead, the lesion demonstrates a cystic structure containing necrotic-hemorrhagic material and, most importantly, a thick laminated acellular membrane. A fibroma does not form such a membrane because it represents a proliferation of fibroblastic tissue rather than a parasitic cyst[6,7]. Therefore, the presence of the characteristic laminated cyst wall, together with the cystic rather than solid architecture, effectively excludes a calcified cardiac fibroma.

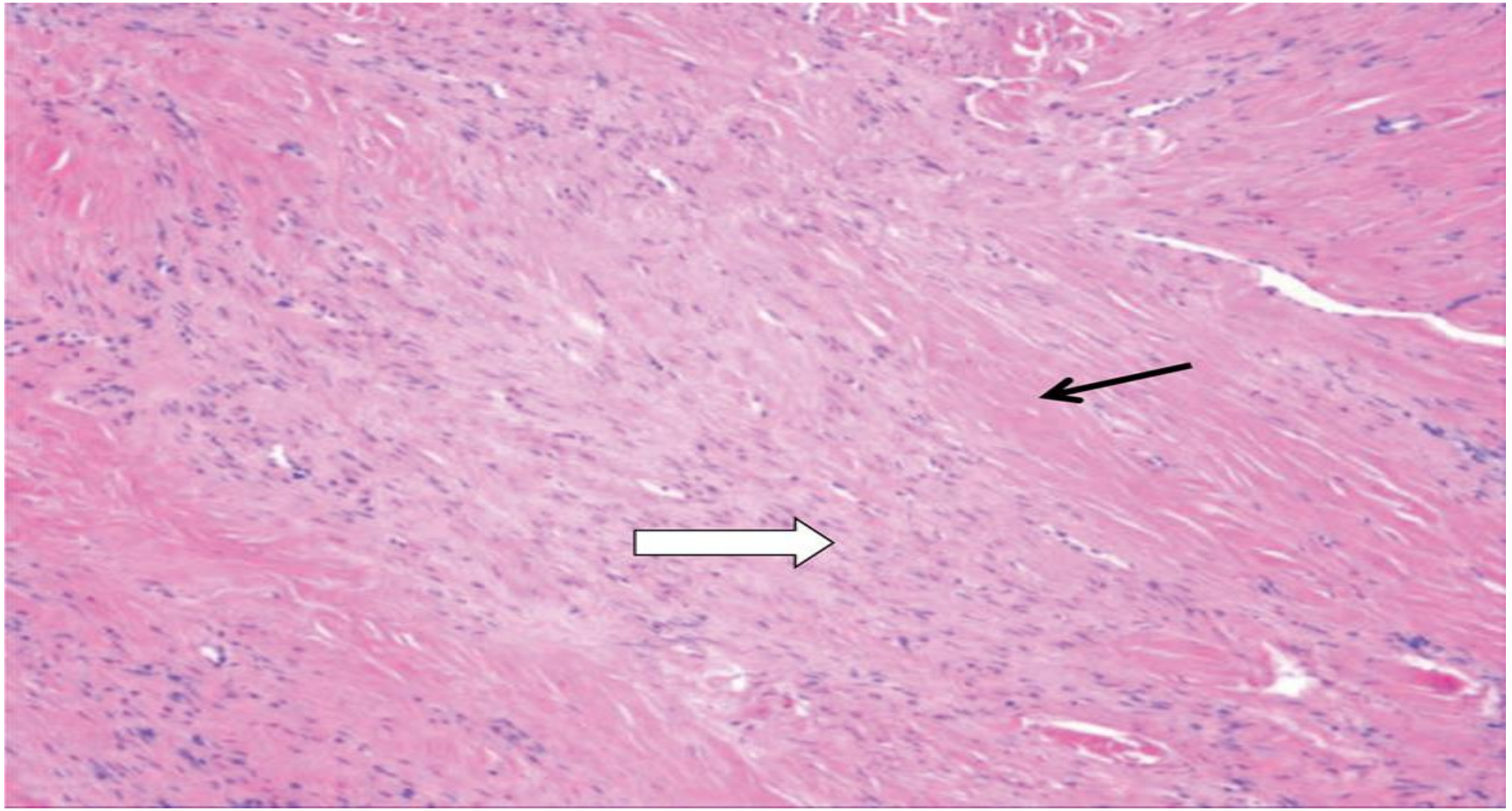


Figure X. Histologic appearance of tumor. Hematoxylin and eosin stain reveals fibroblastic proliferation (open arrow) and dense fibrosis (closed arrow)[7]*

[7]*Menon D, Dentel JN, Sanil Y, Lawrence D. Cardiac Fibroma with Asymptomatic Ventricular Arrhythmia in an Adolescent with Gorlin's Syndrome. J Pediatr Genet. 2021;12(2):171-174. Published 2021 Jan 12. doi:10.1055/s-0040-1722287*

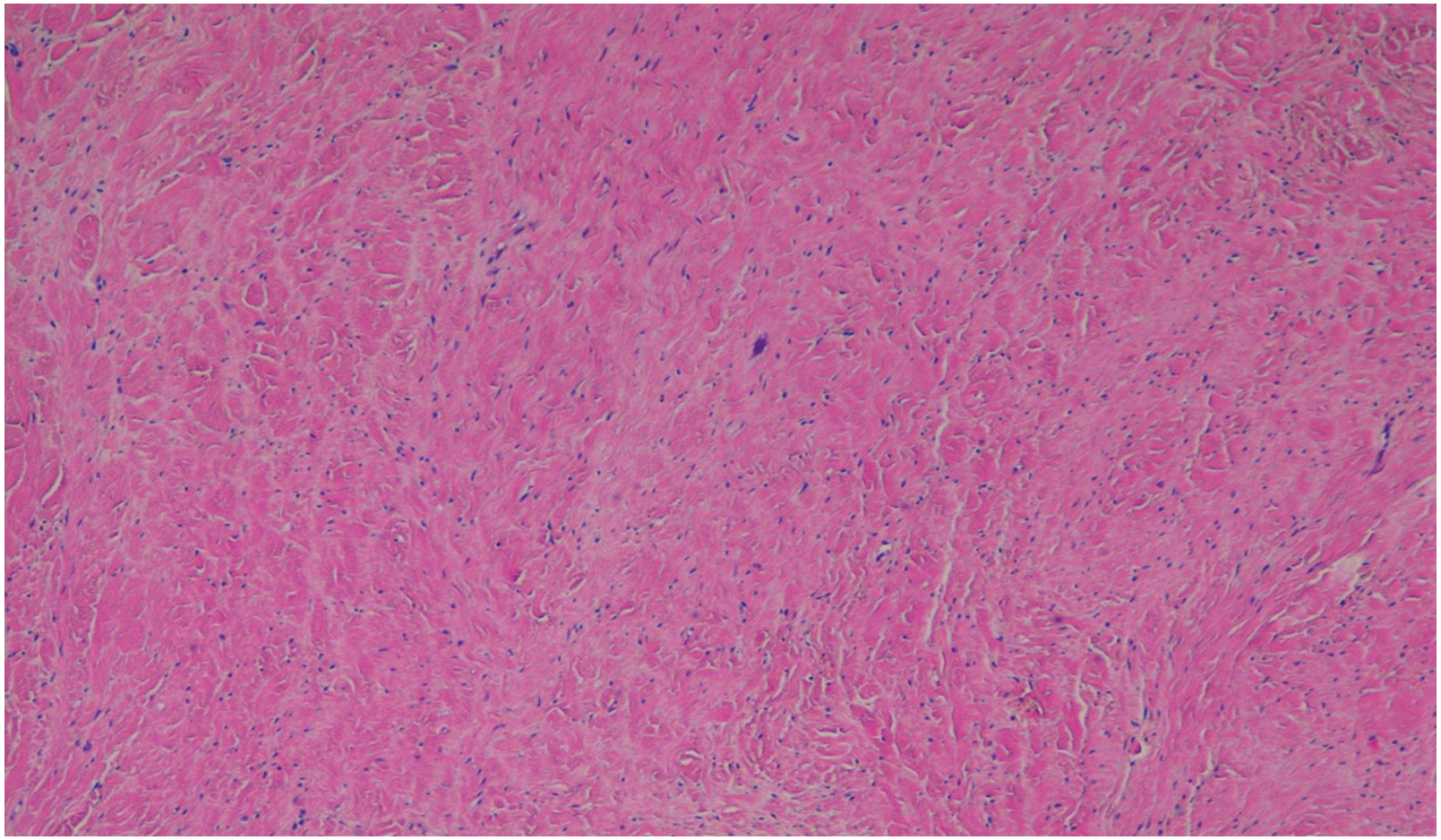


Figure Z. Low-power photomicrograph (hematoxylin-eosin stain) shows that the mass is composed of a variegated strongly and weakly eosinophilic matrix intermixed with a relatively small number of cells showing small nuclei and a lack of atypia. Separate examination with staining supported interpretation of this tissue as a mix of hyaline and fibrillar collagen and elastic fibers with interspersed fibroblastic cells*

[6]*Grunau G.L., Leipsic J.A., Sellers S.L., Seidman M.A. Cardiac Fibroma in an Adult AIRP Best Cases in Radiologic-Pathologic Correlation. Radiographics. 2018;38:1022–2026. doi: 10.1148/rg.2018170205

C. Calcified amorphous tumor of the heart (17.74%)

A calcified amorphous tumor (CAT) of the heart is also an important differential because it is a non-neoplastic cardiac mass that can be markedly calcified and may grossly resemble other calcified cardiac lesions. Histologically, however, CAT has a very specific pattern: it consists predominantly of nodular or dense calcium deposits embedded within amorphous eosinophilic, degenerating fibrinous material, often accompanied by dense collagen, organization, and variable chronic inflammatory cells. It is thought in at least some cases to represent an organized degenerative or thrombotic process rather than a true neoplasm[8]. Consequently, calcification, eosinophilic material, fibrosis, and inflammation alone could superficially overlap with portions of the present lesion. The distinction lies in the organization of the lesion and the nature of the eosinophilic material. In CAT, the eosinophilic material is amorphous degenerating fibrin associated with irregular calcified nodules; it does not form the thick, organized, laminated acellular membrane of a hydatid cyst. Published CAT specimens show calcific nodules surrounded by amorphous fibrin, collagenous fibers, and inflammatory cells, rather than a true parasitic cyst wall[9] (**Figures A&B**). In the present case, therefore, the presence of calcification is not sufficient to favor CAT. The large intramyocardial cystic configuration and, above all, identification of the laminated acellular membrane establish a hydatid cyst and exclude CAT.

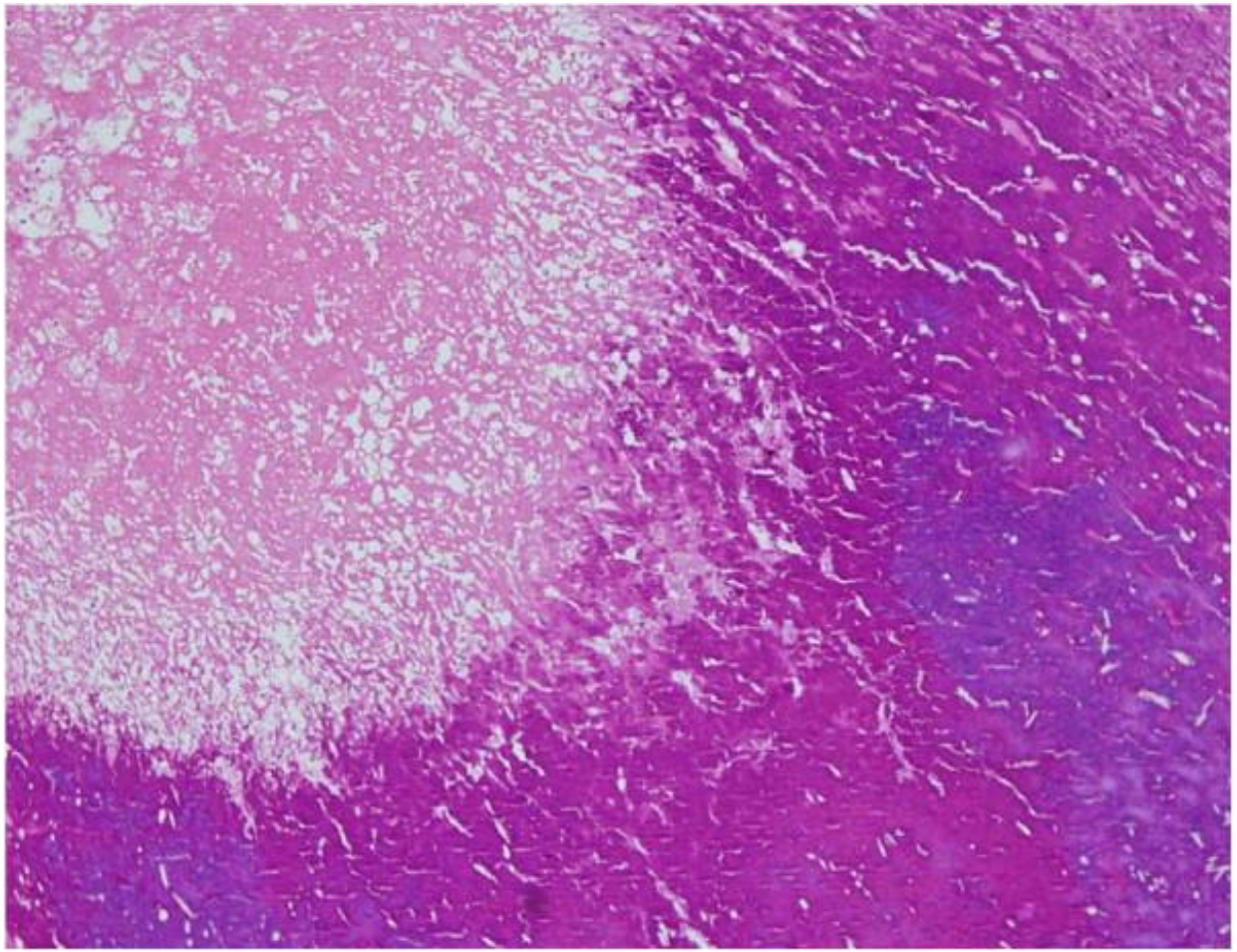


Figure A. Photomicrograph showing amorphous eosinophilic fibrinous material along with dense calcification (hematoxylin and eosin (H&E) ×100)*

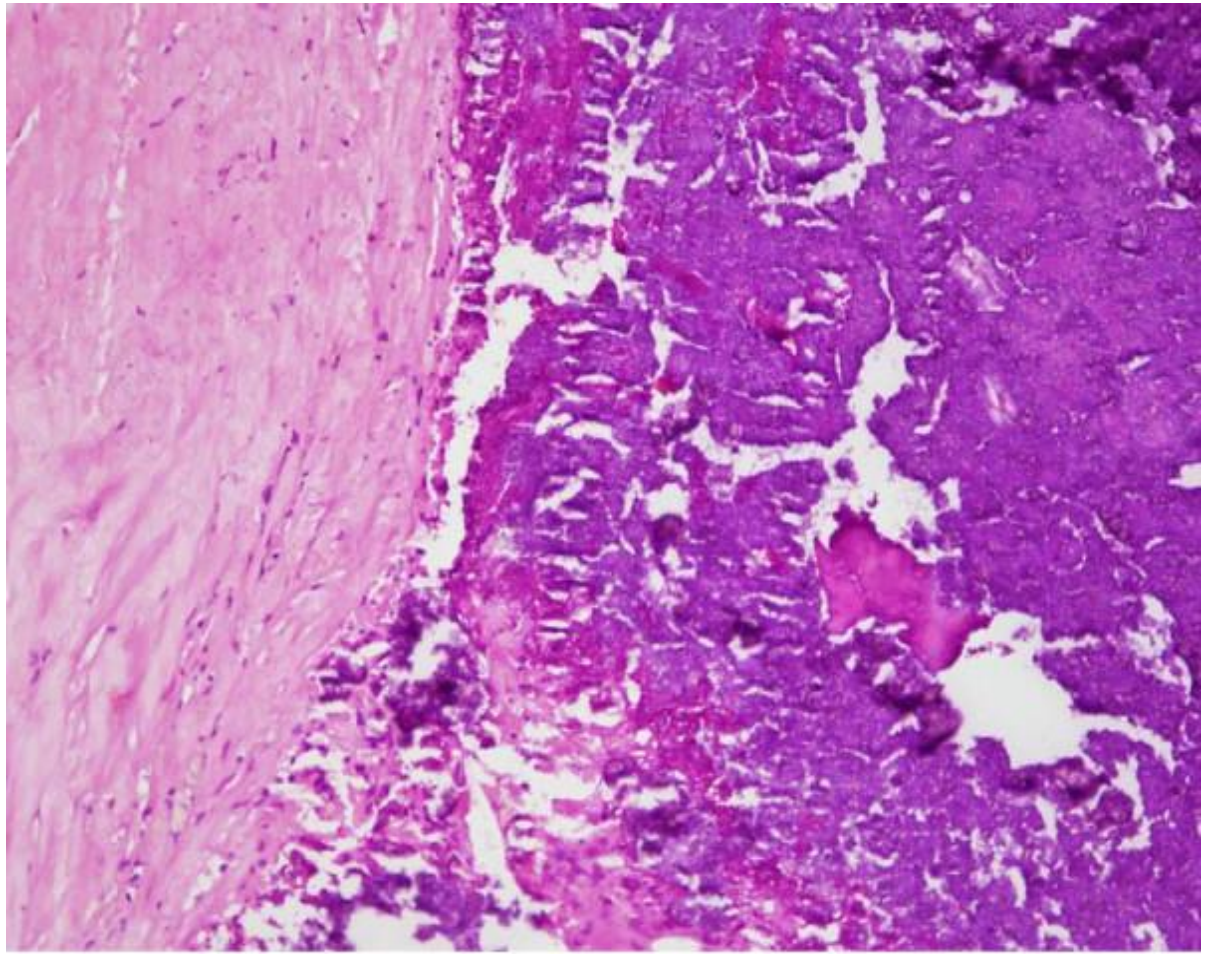


Figure B. Photomicrograph demonstrating hyalinized tissue with foca chronic inflammation and a focus of calcification (H&E ×200)*

[9]**Gupta R, Hote M, Ray R. Calcified amorphous tumor of the heart in an adult female: a case report. J Med Case Rep. 2010;4:278. Published 2010 Aug 19. doi:10.1186/1752-1947-4-278*

D. Cardiac cavernous hemangioma with dystrophic calcification (10.08%)

A cardiac cavernous hemangioma with dystrophic calcification can also enter the differential of an intramyocardial mass because cardiac hemangiomas may arise within the ventricular myocardium, reach substantial size, and undergo secondary thrombosis, fibrosis, degeneration, or calcification[10]. Some may even appear grossly cystic or hemorrhagic. Nevertheless, the defining microscopic feature of a cavernous hemangioma is a proliferation of numerous variably sized, markedly dilated vascular channels containing blood and lined by flattened, cytologically bland endothelial cells[11], with the vascular spaces interspersed among myocardium, fibrous tissue, or adipose tissue[11] (**Figure Y**). Dystrophic calcification or organizing thrombus within such a lesion would be a secondary alteration and would not eliminate the underlying vascular architecture. In the present case, there is no described proliferation of endothelial-lined, blood-filled vascular spaces. Instead, the lesion is a true intramyocardial cyst with a fibrous inflammatory host response, necrotic-hemorrhagic contents, focal calcification, and a thick eosinophilic laminated acellular membrane. That membrane cannot be explained by thrombosis, hemorrhage, or dystrophic calcification within a hemangioma. Thus, although the hemorrhagic and calcified components could superficially suggest a degenerated vascular lesion, the absence of cavernous vascular channels and presence of the characteristic hydatid membrane exclude a cardiac cavernous hemangioma.

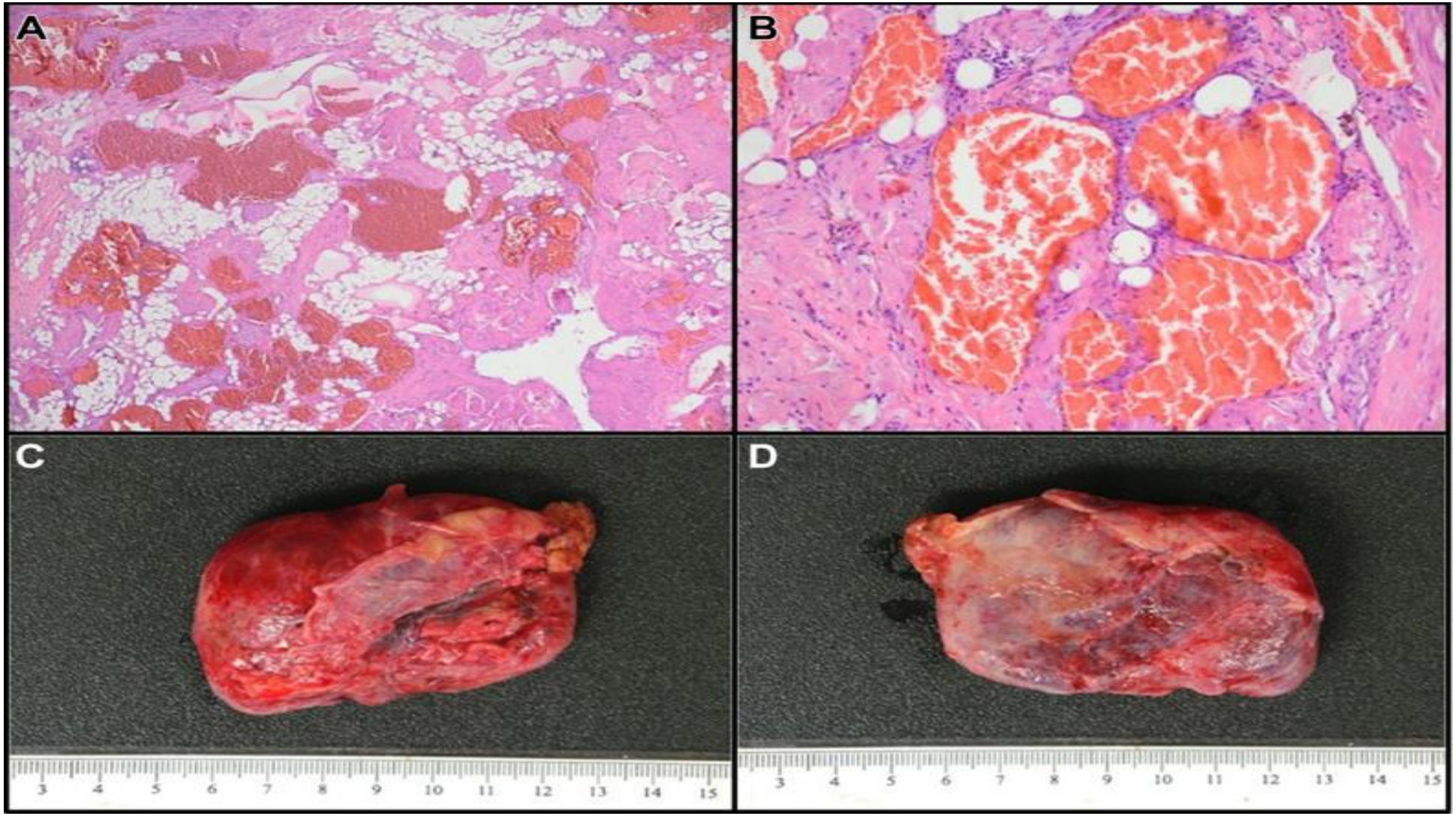


Figure Y: (A) Low power view with dilated vascular spaces, muscle, and adipose tissue. (B) High power view with flattened, bland, lining of vascular channels. (C and D) Gross appearance of the excised cavernous hemangioma tumor*

[11]*Qamar F, Kharsa C, Letham P, et al. Cardiac Cavernous Hemangioma. JACC Case Rep. 2025;30(4):102956. doi:10.1016/j.jaccas.2024.102956*

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11. Qamar F, Kharsa C, Letham P, et al. Cardiac cavernous hemangioma. *JACC Case Rep.* 2025;30(4):102956. doi:10.1016/j.jaccas.2024.102956. PMID: 39972709; PMCID: PMC11862176.