Case #96

NAME Educational Activities Committee

Case provided by:

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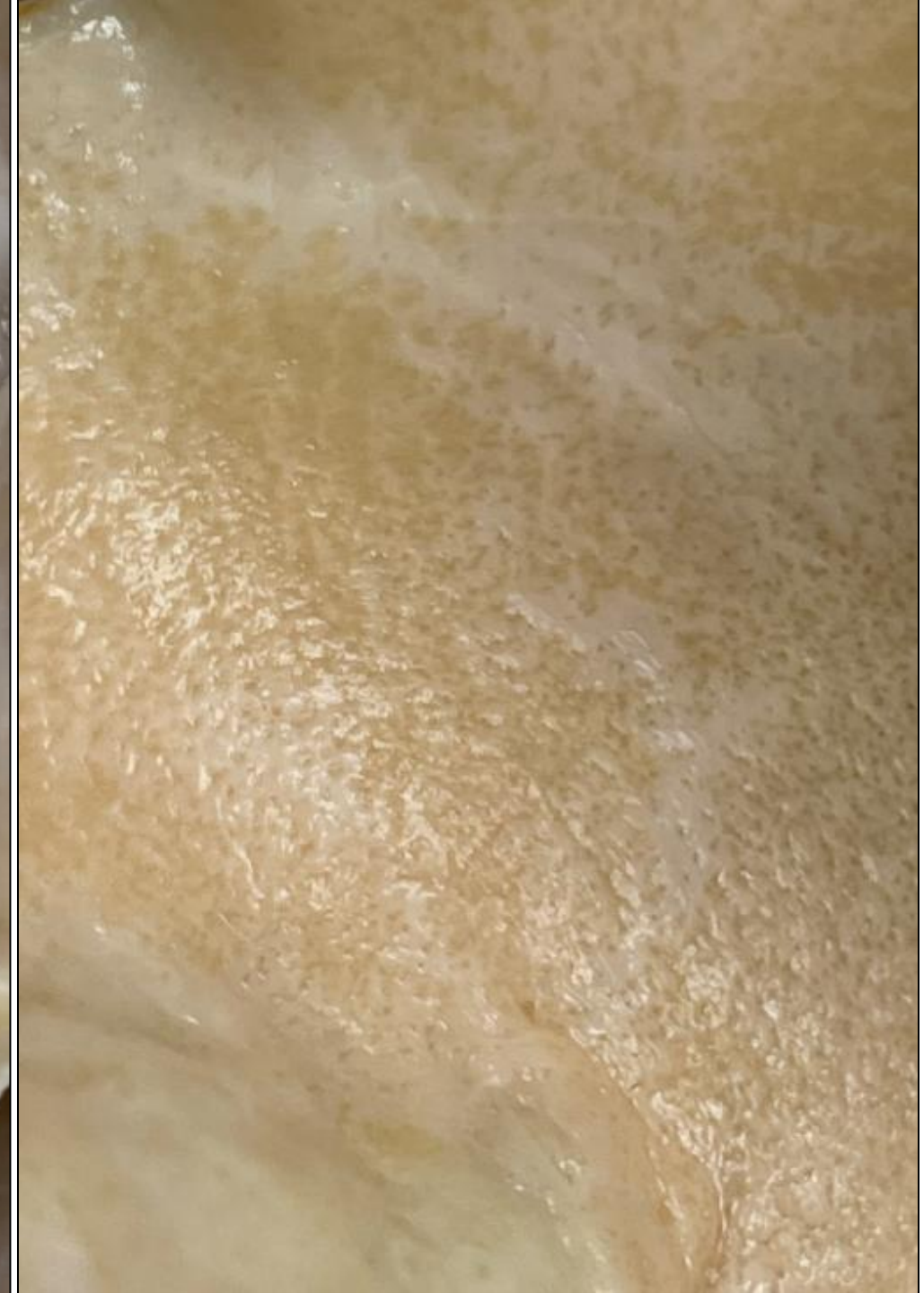


Examination of the left atrium of a 55-year-old man who experienced sudden cardiac death revealed the findings depicted in the images.

What is the most likely diagnosis?

A. Cardiac amyloidosis
B. Hypertrophic cardiomyopathy
C. Microvesicular steatosis of the heart
D. Fabry disease

Answer...

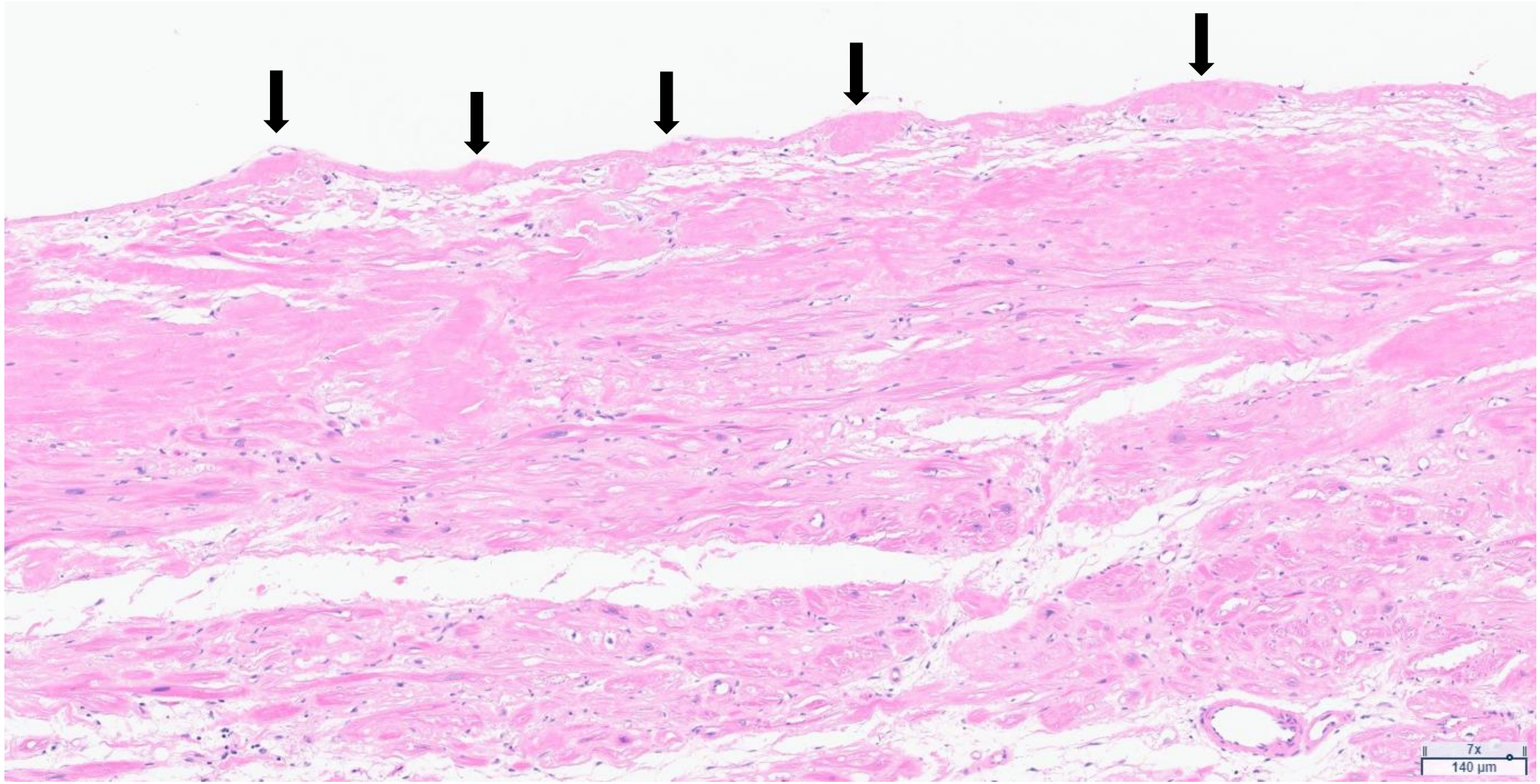
C. Cardiac amyloidosis

Amyloid deposition can occur in various regions of the heart, encompassing the myocardium, vessels, endocardium, valves, epicardium, and parietal pericardium.

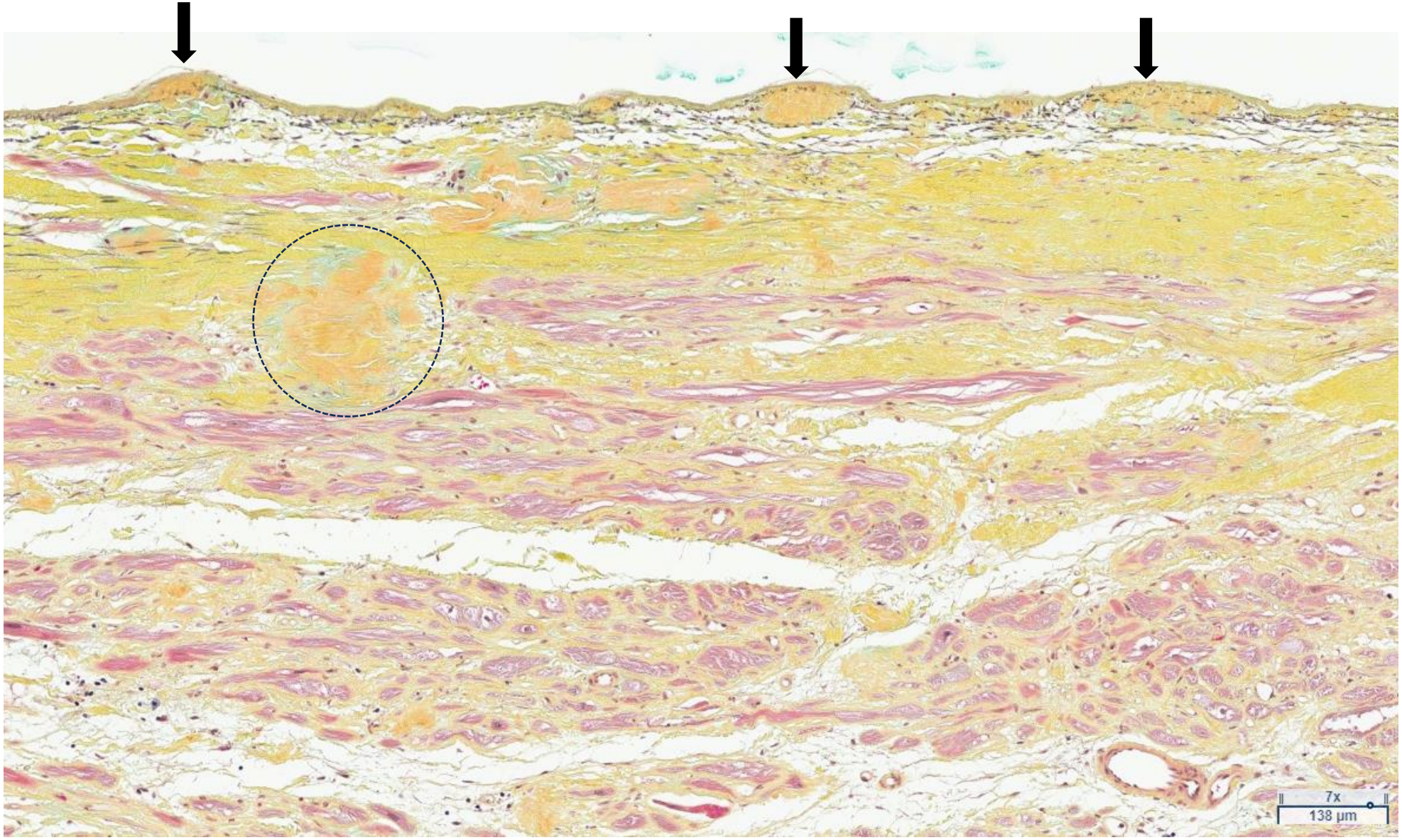
Macroscopically, the myocardium affected by amyloid infiltration typically exhibits a tan, firm, and rubbery texture. The degree of infiltration displays significant heterogeneity, manifesting through three primary patterns:

- Diffuse
- Segmental (with frequent involvement of the septum and inferolateral walls).
- Subendocardial

In cardiac amyloidosis, the atria are mildly enlarged and thickened. Subendocardial amyloid deposits (see next image) can impart a gritty or sandpaper-like appearance to the heart lining like seen in the question image, especially noticeable following formalin fixation. This visual appearance can also be seen on the atrioventricular valves in cases where endocardial involvement is present.



Photomicrograph showing subendocardial amyloid deposits in the left atrium (arrows). Hematoxylin and eosin staining. Digitally acquired image, scale bar as shown.



This photomicrograph depicts subendocardial amyloid deposits highlighted in orange within the left atrium using a Movat pentachrome stain (arrows). Additionally, amyloid deposits are observed within the myocardium (circle). Digitally acquired image, scale bar as shown.

Other responses…

A. Microvesicular steatosis
Cardiac microvesicular steatosis refers to the presence of many lipid droplets inside the myocytes due to excessive fat accumulation in the cardiac muscle.

B. Hypertrophic cardiomyopathy
HCM is characterized by regional and often asymmetric myocardial hypertrophy, ranging from mild (13–15 mm) to massive (30 mm).

D. Fabry disease
Fabry disease is an X-linked disorder that results from deficiency of the lysosomal enzyme α-galactosidase. It typically manifests as concentric LV hypertrophy, with only 5% of cases showing asymmetric septal hypertrophy. The main histologic finding is the vacuolization of both working and conduction myocytes. These deposits can cause a significant increase in individual cardiac myocyte cross-sectional area, thereby contributing to the cardiac hypertrophy.

None of these disorders are typically associated with a gritty or sandpaper-like appearance of the endocardial surface.

REFERENCES

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