



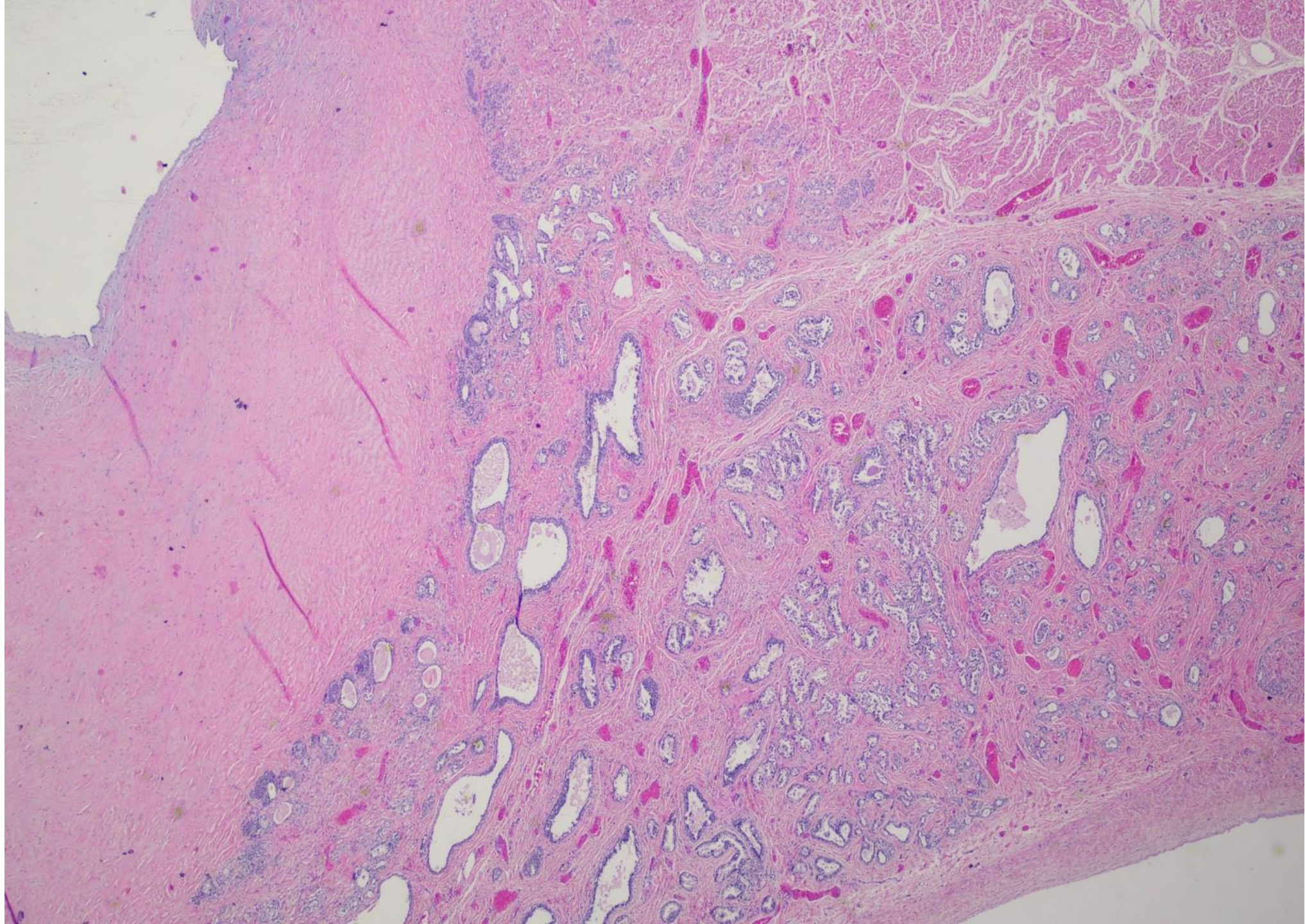
Case #68

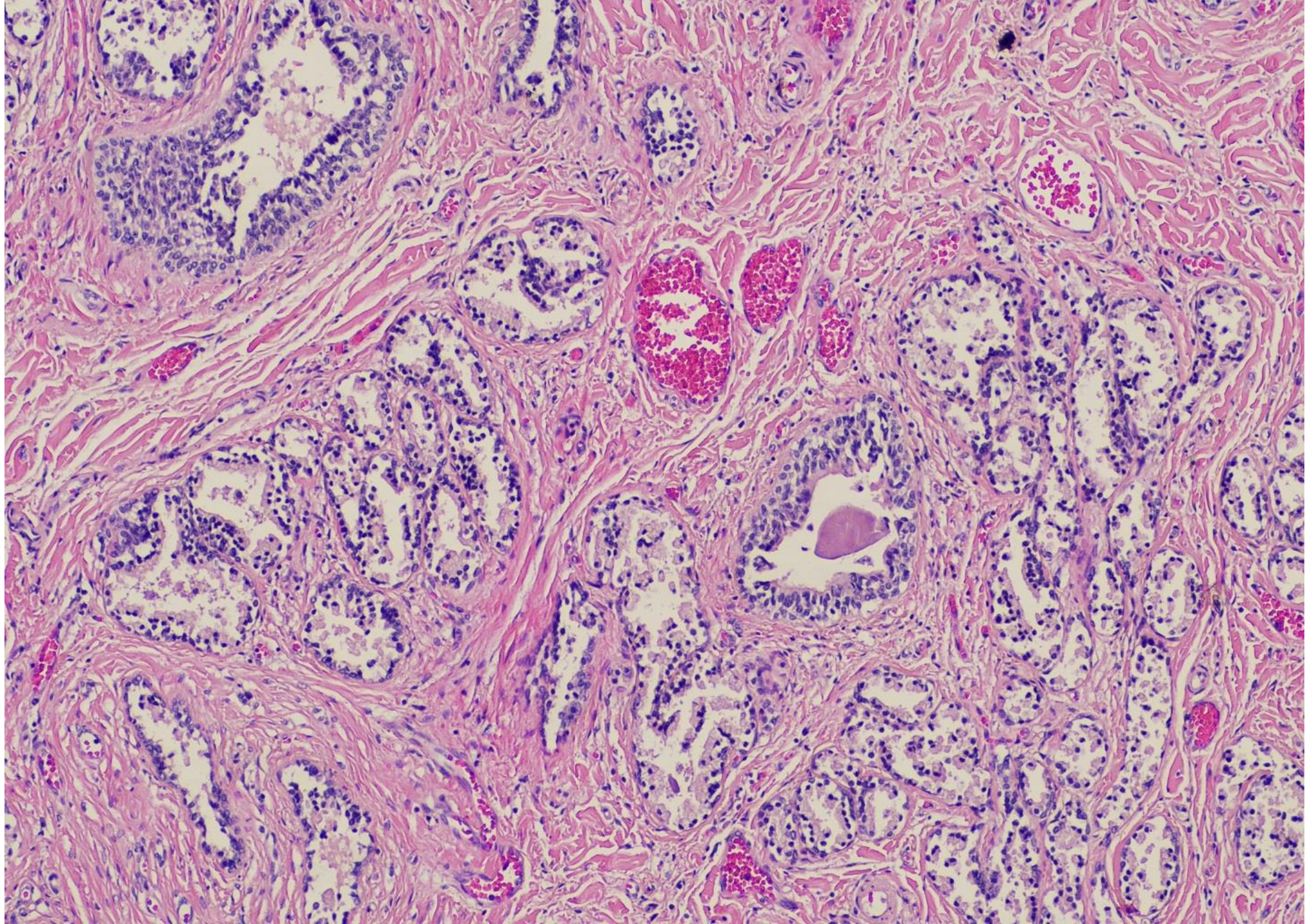
NAME Educational Activities Committee

Case provided by:

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1. A 16-year-old female with a medical history of congenital heart block and implanted pacemaker was found unresponsive in bed and pronounced dead at the scene. The autopsy showed a grossly normal heart with no structural abnormalities. Interrogation of the pacemaker showed a normally paced rhythm with an abrupt episode of ventricular fibrillation. A genetic panel for inherited arrhythmias and cardiomyopathies was negative. Histological examination of the cardiac conduction system identified the lesion depicted above in the Triangle of Koch. What is the correct diagnosis?

- Cystic tumor of the AV node
- Mature cystic teratoma
- Mesothelial cyst
- Metastatic carcinoma
- Bronchogenic cyst

Answer...

A. Cystic tumor of AV node (CORRECT RESPONSE, 64.86% of responses)

Previously known by many names, including mesothelioma of AV node, the cystic tumor of the AV node is a benign entity which may nonetheless cause varying degrees of heart-block or fatal arrhythmia. Its pathogenesis is not understood, but immunohistochemical studies support it being of endodermal origin and it may represent endoderm trapped in the heart during embryogenesis. Pacemakers may correct heart block, but fatal arrhythmias may still develop. The size of the tumor is not predictive of arrhythmia development in cases to date. It occurs within the Triangle of Koch in the atrioventricular node and may extend locally to include other elements of the cardiac conduction system and is often grossly inapparent. It should be considered in cases of sudden cardiac death when other causes have been excluded.

Other responses...

B. Mature cystic teratoma (7.92% responses)

A cystic teratoma should have cells from more than one germ cell layer, as is commonly seen in cystic teratomas elsewhere.

C. Mesothelial cyst (3.86% responses)

Mesothelial cyst should have mesothelial cells and should be epicardial rather than within the myocardium.

D. Metastatic carcinoma (9.65% responses)

Metastatic carcinoma would have malignant cytology and/or invasive growth. The location and bland cytology are characteristic of a cystic tumor of AV node.

E. Bronchogenic cyst (13.71% responses)

Bronchogenic cysts have respiratory type epithelium with smooth muscle, +/- cartilage or sero-mucinous type glands.

REFERENCES

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