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FIT Clinical Decision-Making

Uhl's anomaly: An uncommon cause of fetal cardiomegaly

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BACKGROUND

Uhl's anomaly is a rare congenital heart defect associated with right ventricular and tricuspid valve dysplasia. Prenatal differentiation from Ebstein anomaly is challenging but is important for prognostication.

CASE

A 29 year old woman underwent fetal echocardiography at 22 weeks gestation due to fetal cardiomegaly. Imaging revealed an abnormal cardiac axis (80 degrees), a cardiothoracic ratio of 71%, severe dilation of the right atrium and right ventricle (Fig 1) and severe tricuspid valve regurgitation. The entire pulmonary outflow tract was moderately hypoplastic. The left heart was normal in size and without dysfunction. There was no evidence of hydrops.

RESULTS

Although Ebstein anomaly was initially considered, severe right ventricular dilation and thinning combined with a normal location of the tricuspid annulus at the AV groove supported a diagnosis of Uhl's anomaly. Parents were advised of the high probability of fetal demise and elected to continue the pregnancy. Single ventricle repair was deemed unfeasible, and exit to ECMO as a bridge to cardiac transplantation was considered. However, intrauterine fetal demise was diagnosed at 39 weeks. Pathology revealed massive right atrial and right ventricular dilation (Fig 2), a thin, floppy right ventricular wall and tricuspid valve dysplasia (Fig 3). Pulmonary valve atresia and main pulmonary artery hypoplasia were present. Histology revealed abnormal right ventricular wall dimensions ranging from 81-451 μm (Fig 4).

CONCLUSION

Uhl's anomaly should be considered in cases of fetal cardiomegaly with significant right ventricular and right atrial dilation. A thin-walled right ventricle and non-displaced tricuspid valve are the differentiating characteristics. When combined with severe fetal cardiomegaly and significant tricuspid regurgitation, this condition has a grim prognosis.



SCAN ME

Questions?
Contact Bethany Runkel MD
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For more information on Uhl's anomaly,
scan the QR code

FIGURE 1

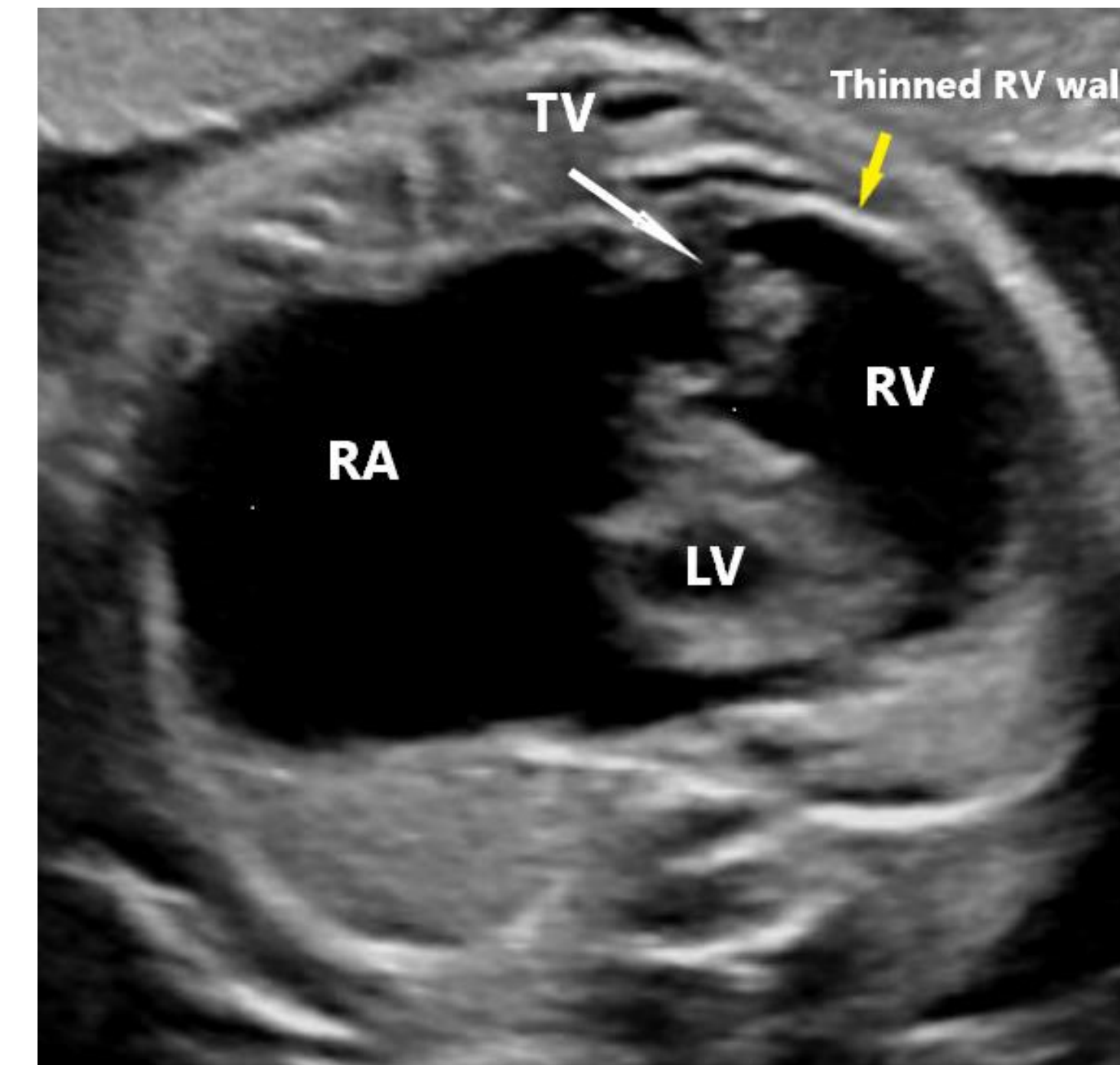


FIGURE 2

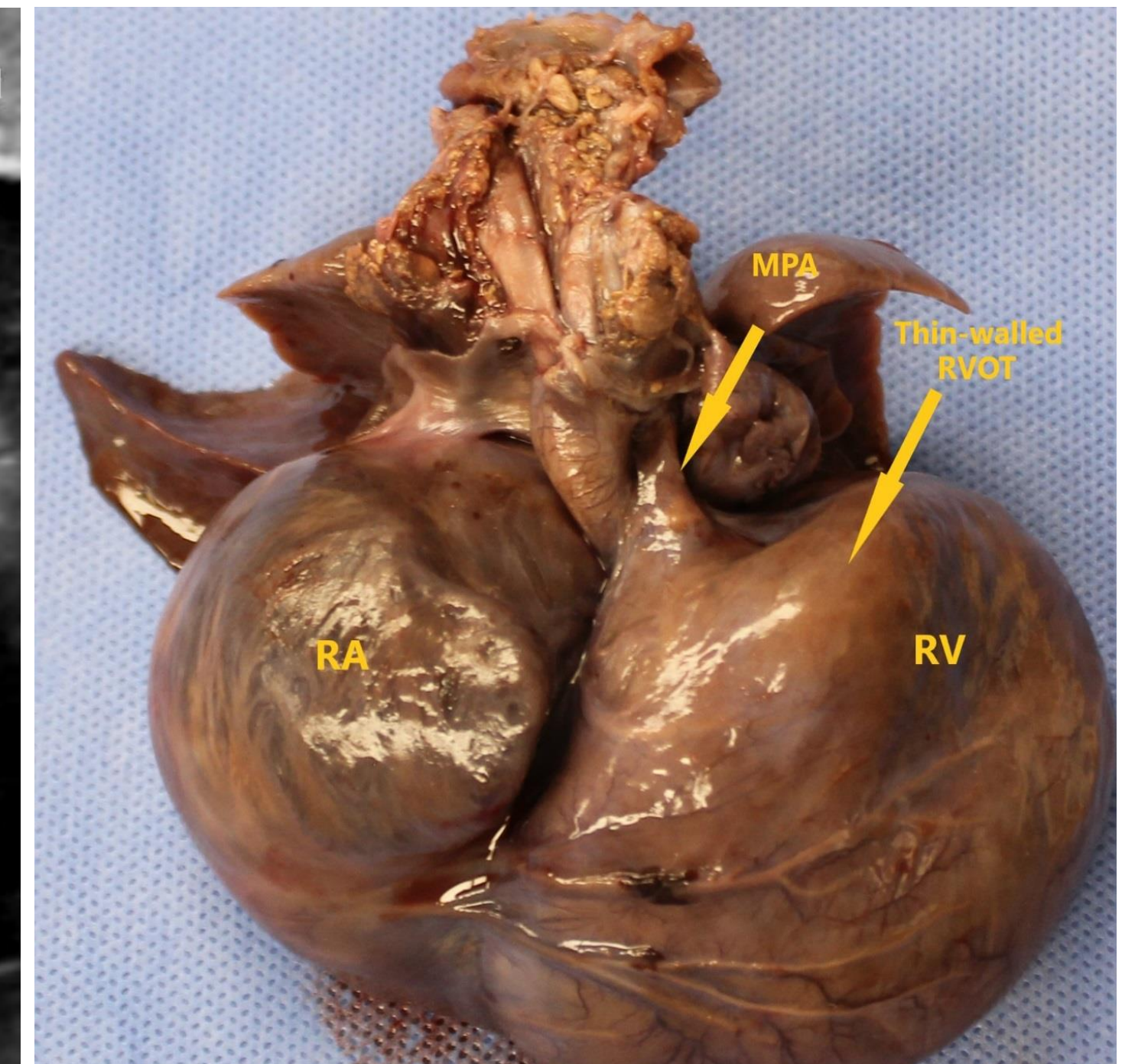


FIGURE 3

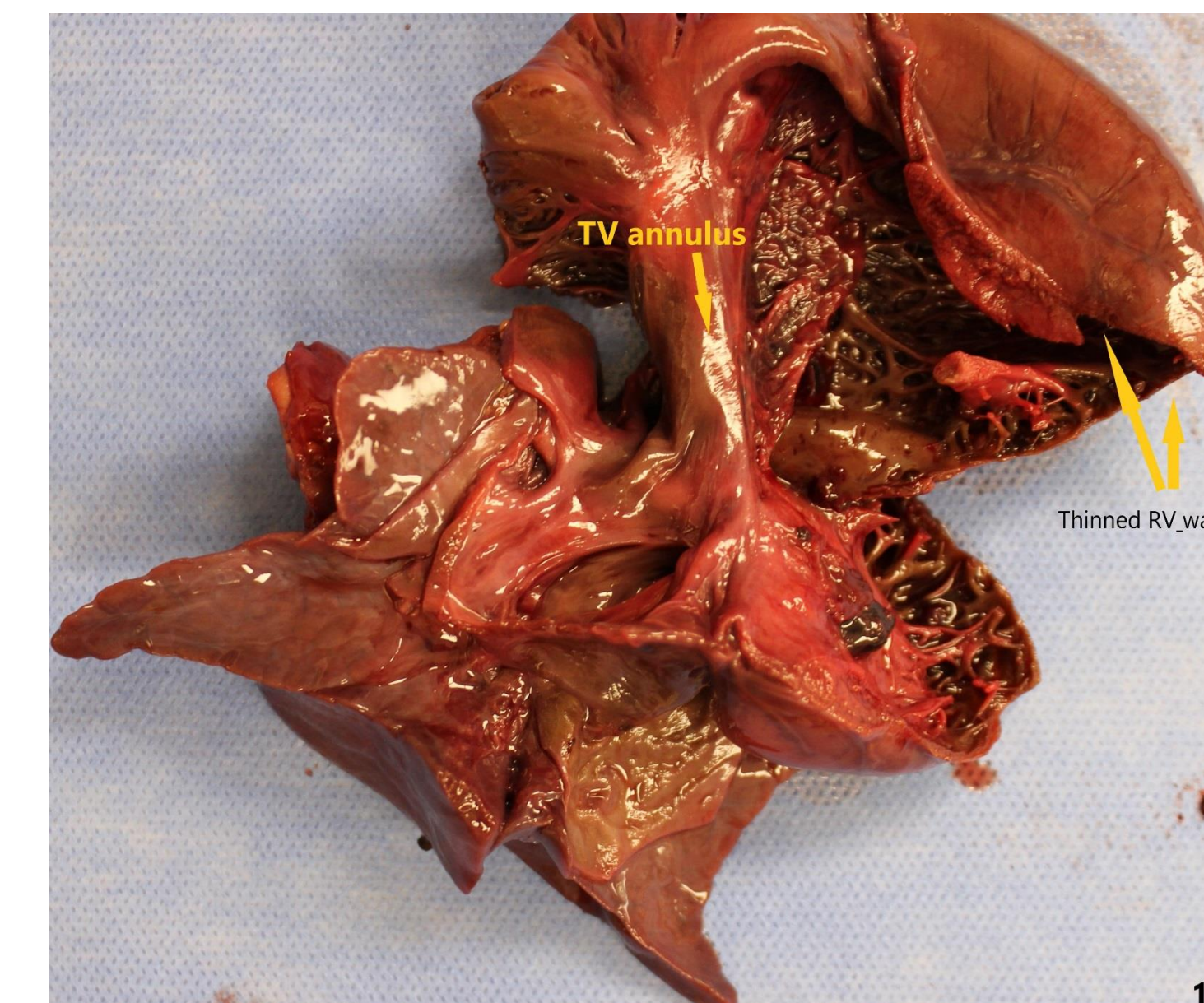
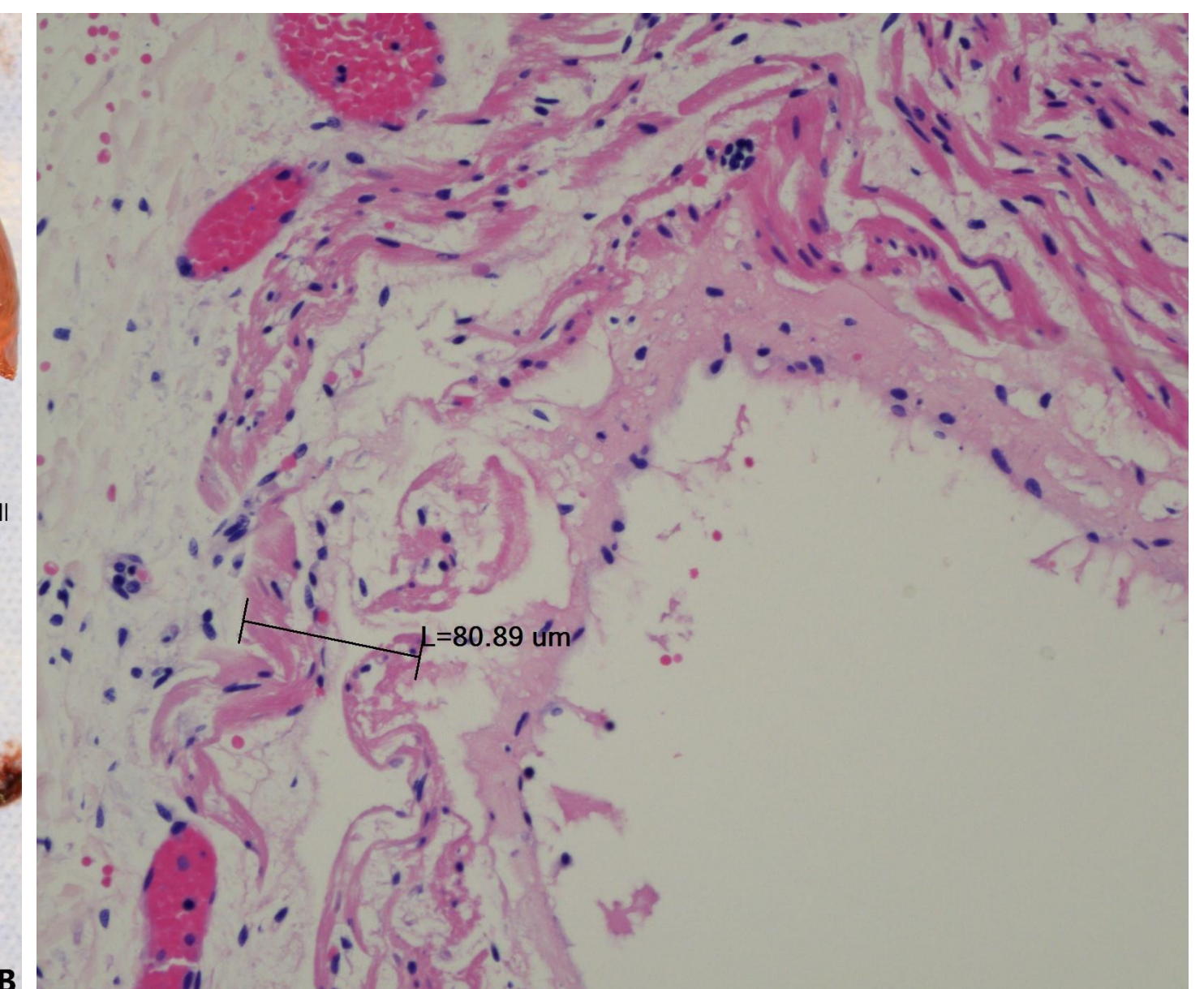


FIGURE 4



DISCLOSURE INFORMATION

The authors have no relevant disclosures.