

Endocardial fibroelastosis

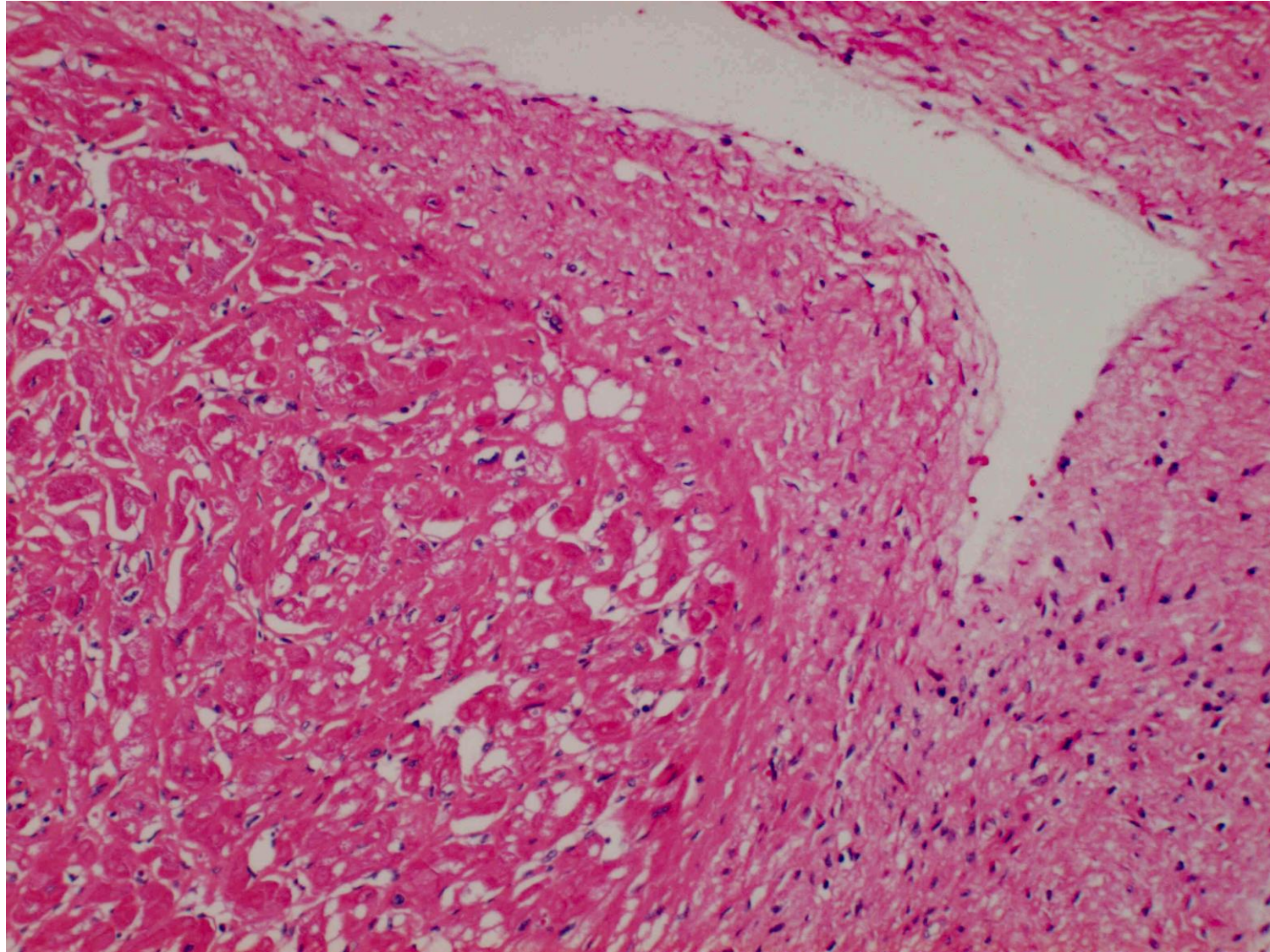
Endocardial fibroelastosis (EFE), an infantile form of restrictive cardiomyopathy, is an uncommon cause of unexplained heart failure in children two years old and younger. EFE is featured by thickening of the endocardium due to an increase in the amount of supporting connective tissue and elastic fibers. Grossly, the endocardium shows glistening white appearance. Mural thrombus formation is common. Heart complications of EFE include thromboembolism, arrhythmia, mitral regurgitation, dilated cardiomyopathy, congestive heart failure and sudden cardiac death. Right heart involvement leads to pulmonary hypertension. There are both inherent and acquired forms. The acquired form is related to the reaction to stress, such as viral infections, cardiomyopathies, immunologic diseases, congenital malformations and electrocution by lightning strike. The exposure to mumps virus in the fetal life may be closely related to EFE, and a skin test for mumps is positive in the affected infant. The mumps viral genome is detectable in the heart tissues of the patient. Mumps immunization can prevent the disease.

Ref.-1: Noren GR, et al. Positive skin reactivity to mumps virus antigen in endocardial fibroelastosis. J Pediatr 1963; 62: 604-606. doi: 10.1016/S0022-3476(63)80021-9

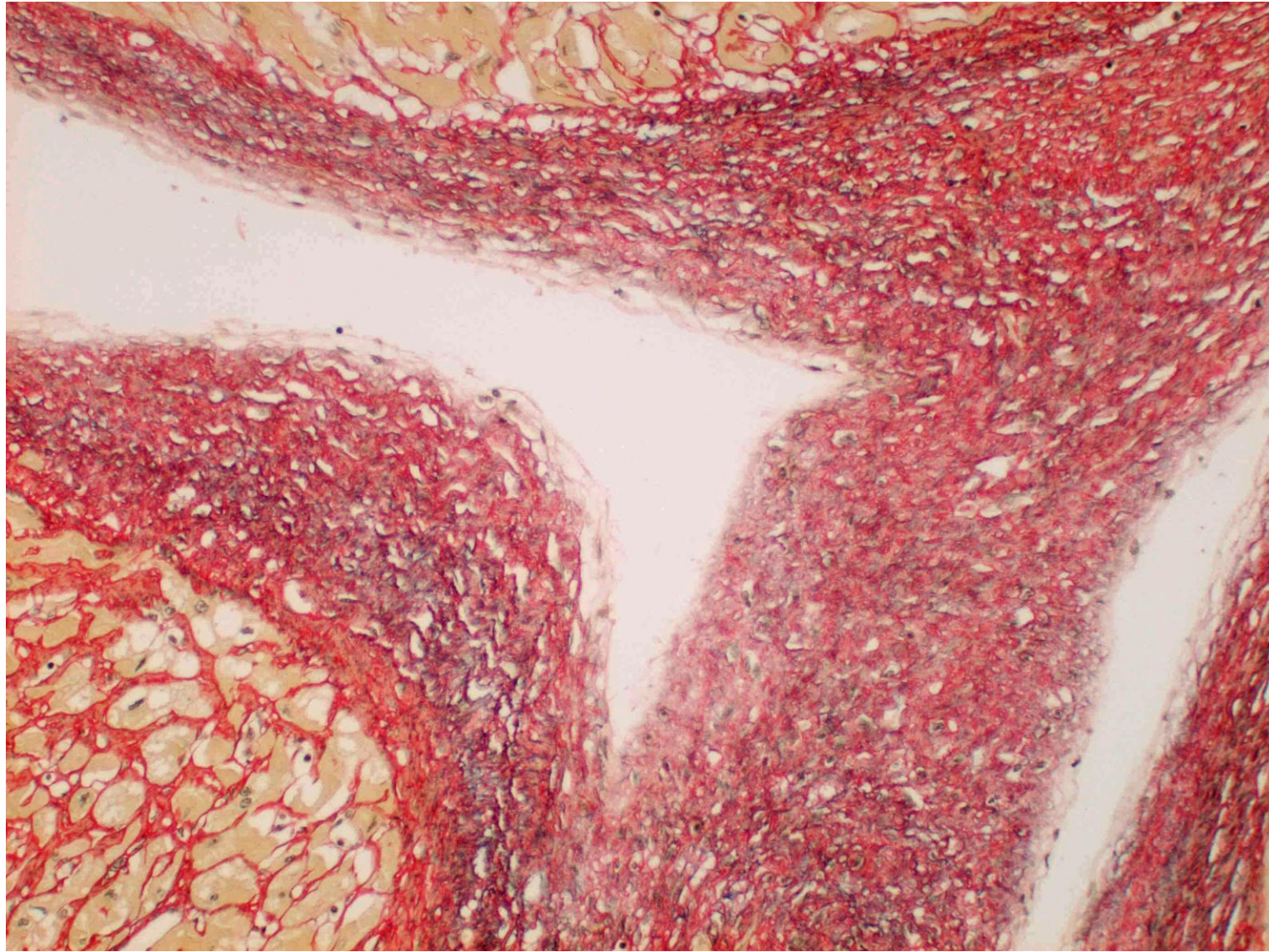
Ref.-2: Luca AC, et al. Endocardial fibroelastosis and dilated cardiomyopathy: the past and future of the interface between histology and genetics. Rom J Morphol Embryol 2020; 61(4): 999-1005. doi: 10.47162/RJME.61.4.02



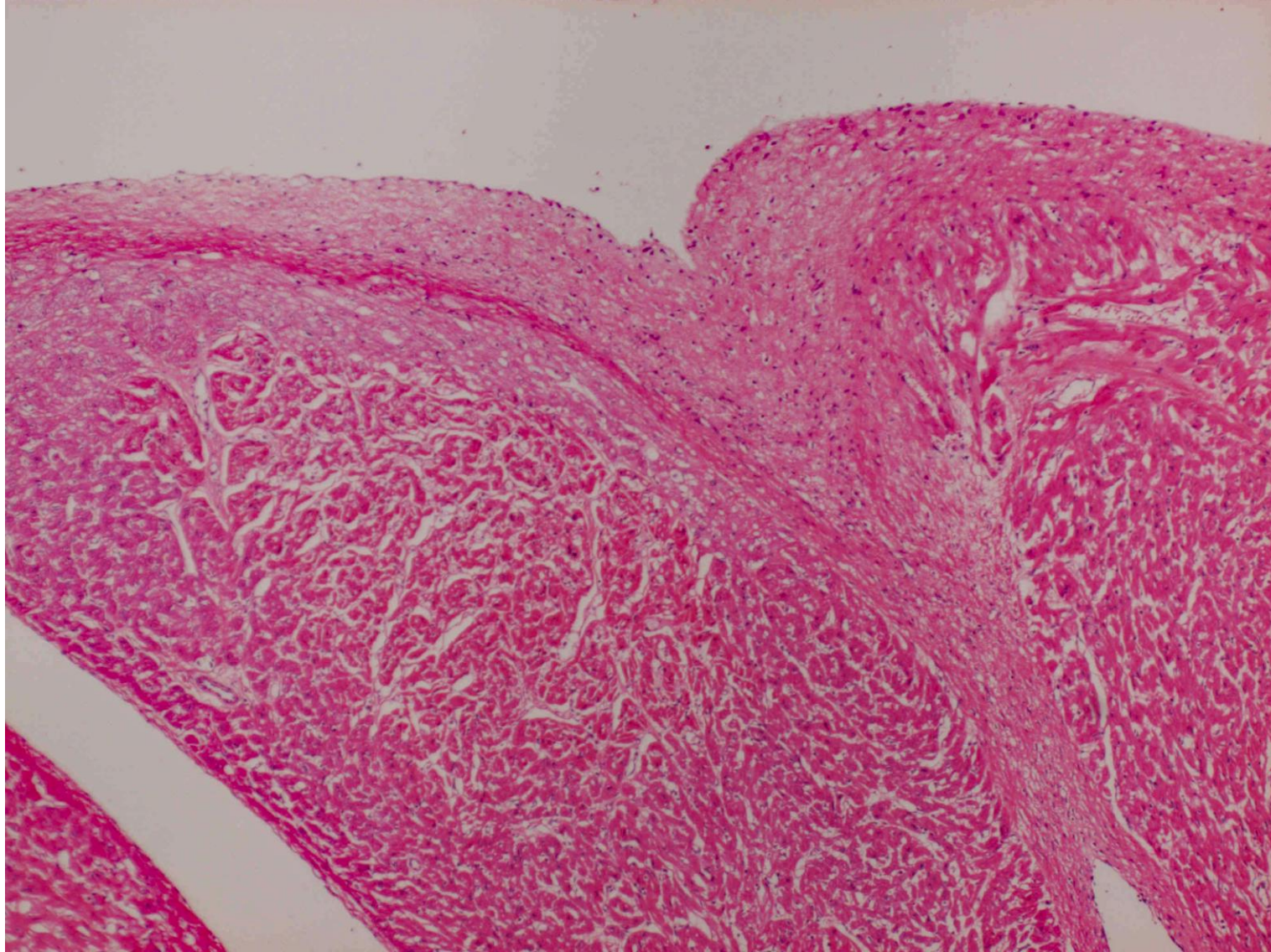
Endocardial fibroelastosis seen in a 7 months-old girl. A horizontal section of the autopsied heart reveals diffuse thickening of the endocardium of the left ventricle with glistening white appearance. Left heart failure led the patient to death.



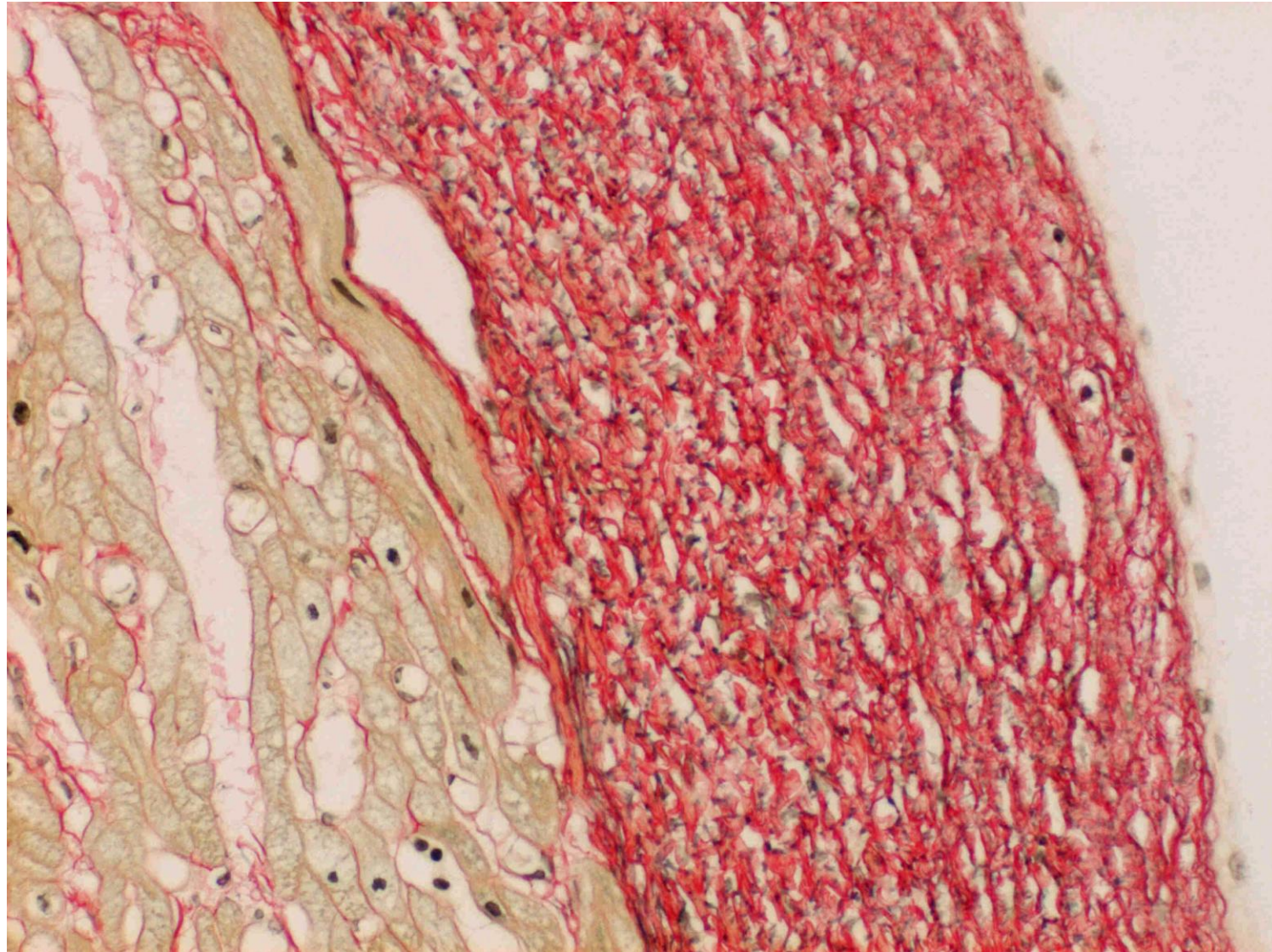
Endocardial fibroelastosis seen in a 7 months-old girl. The endocardium of the left ventricle reveals diffuse fibrous thickening. Neither inflammatory change nor substance deposition is discerned (H&E-1).



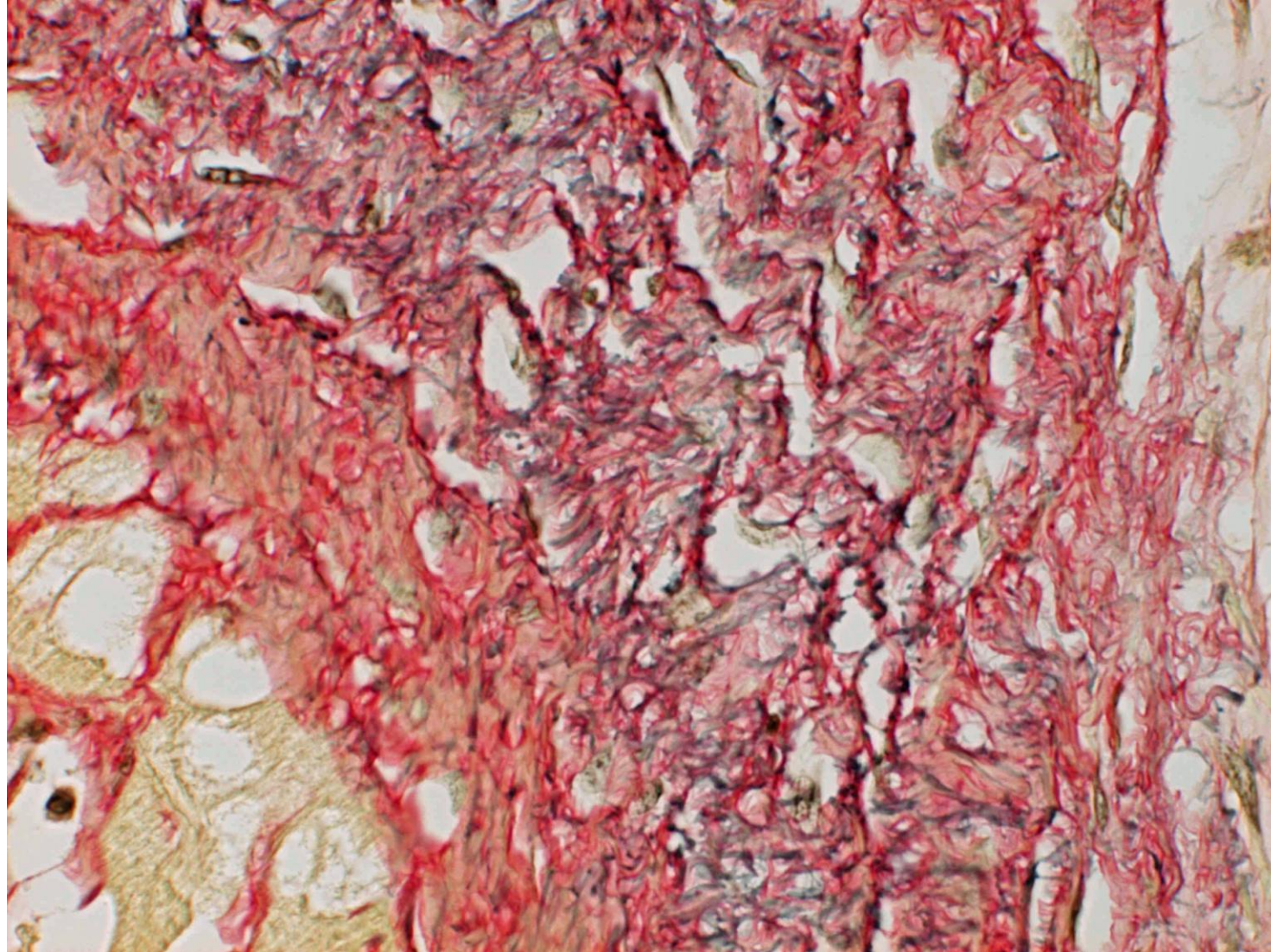
Endocardial fibroelastosis seen in a 7 months-old girl. The endocardium of the left ventricle reveals diffuse fibrous thickening. Elastic fibers are intermingled with the collagenous fibrosis (EVG-1).



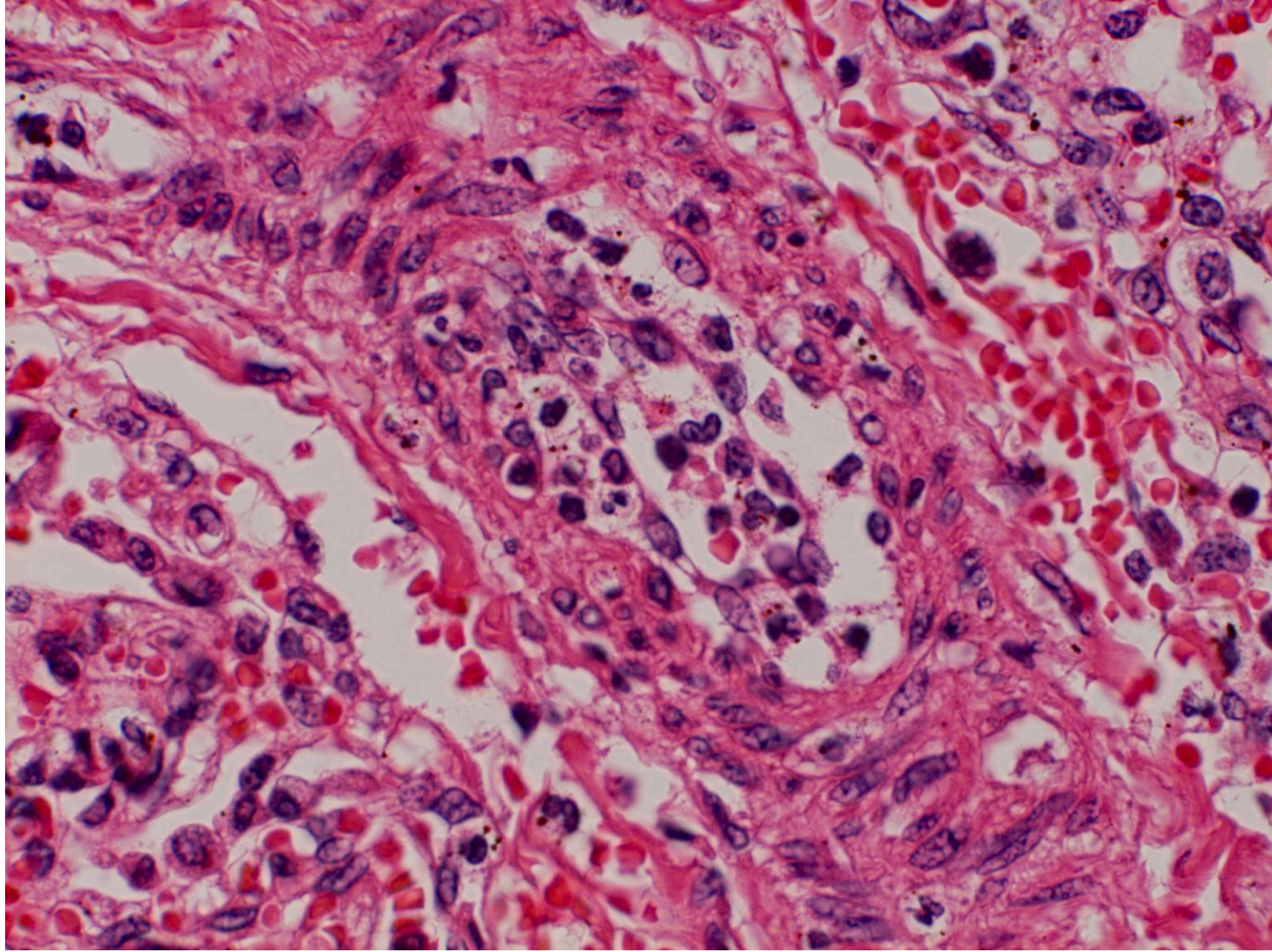
Endocardial fibroelastosis seen in a 7 months-old girl. The endocardium of the left ventricle reveals diffuse fibrous thickening. Neither inflammatory change nor substance deposition is discerned (H&E-2).



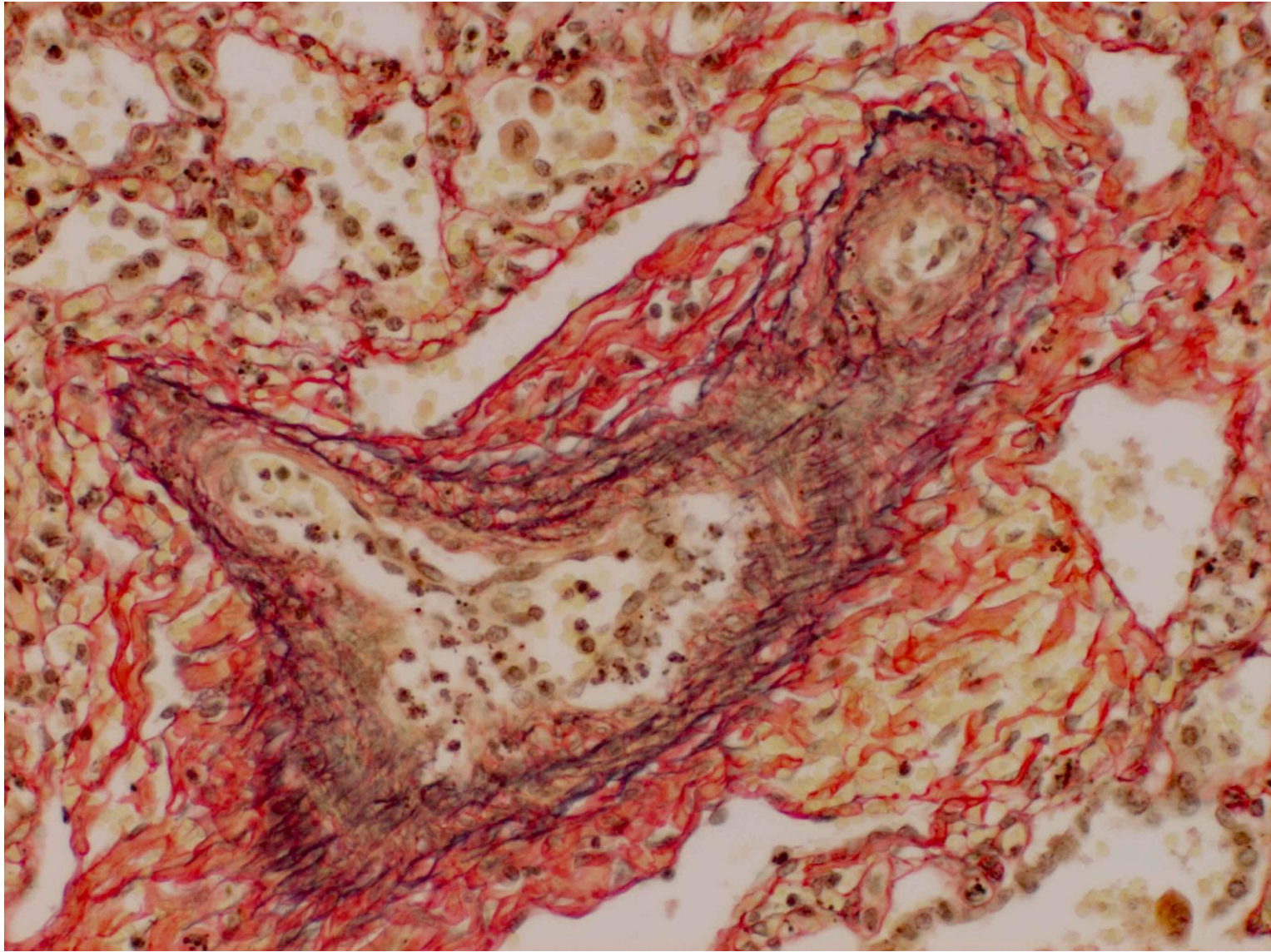
Endocardial fibroelastosis seen in a 7 months-old girl. The endocardium of the left ventricle reveals diffuse fibrous thickening. Elastic fibers are intermingled with the collagenous fibrosis (EVG-2).



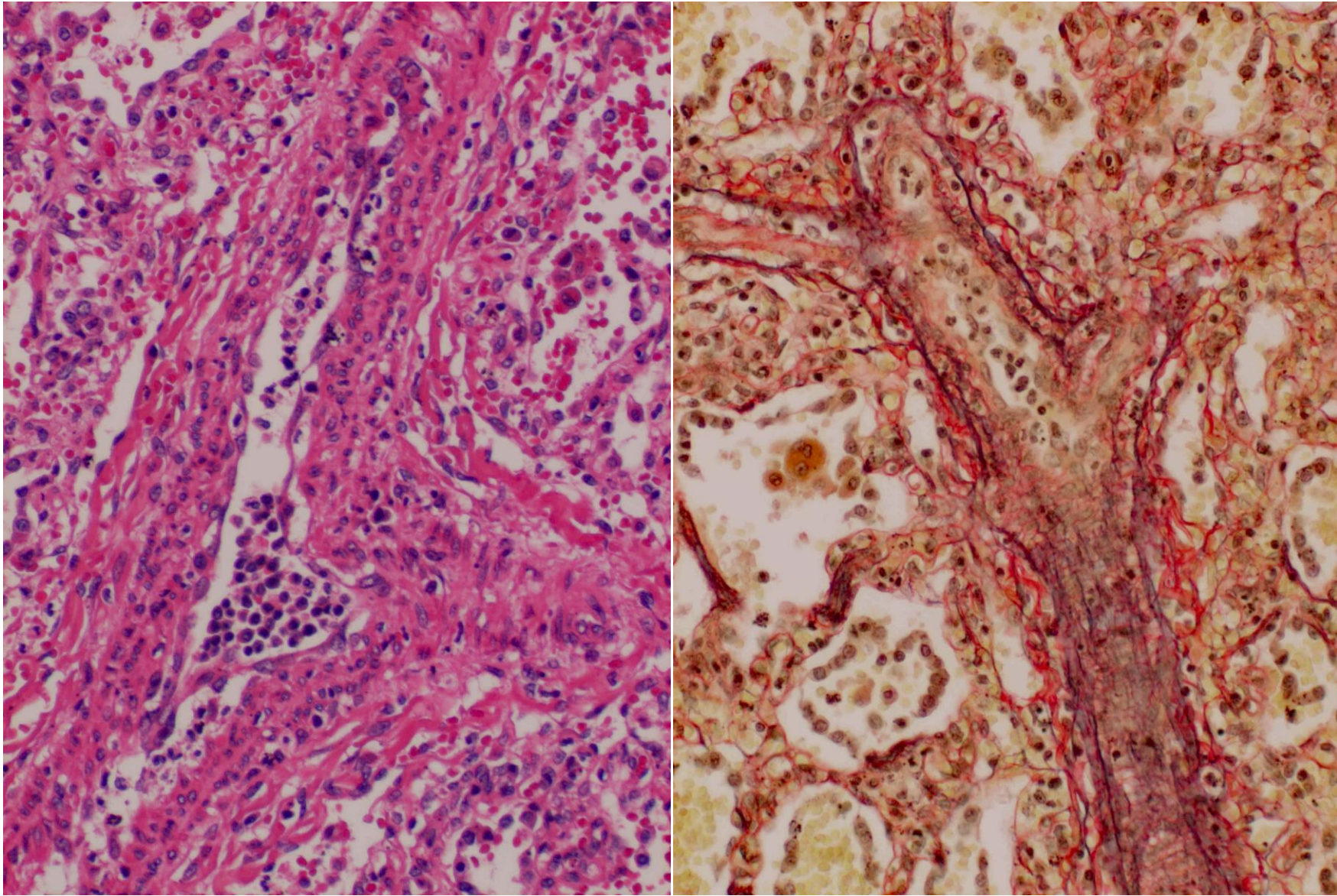
Endocardial fibroelastosis seen in a 7 months-old girl. The endocardium of the left ventricle reveals diffuse fibrous thickening. Elastic fibers are intermingled with the collagenous fibrosis (EVG-3).



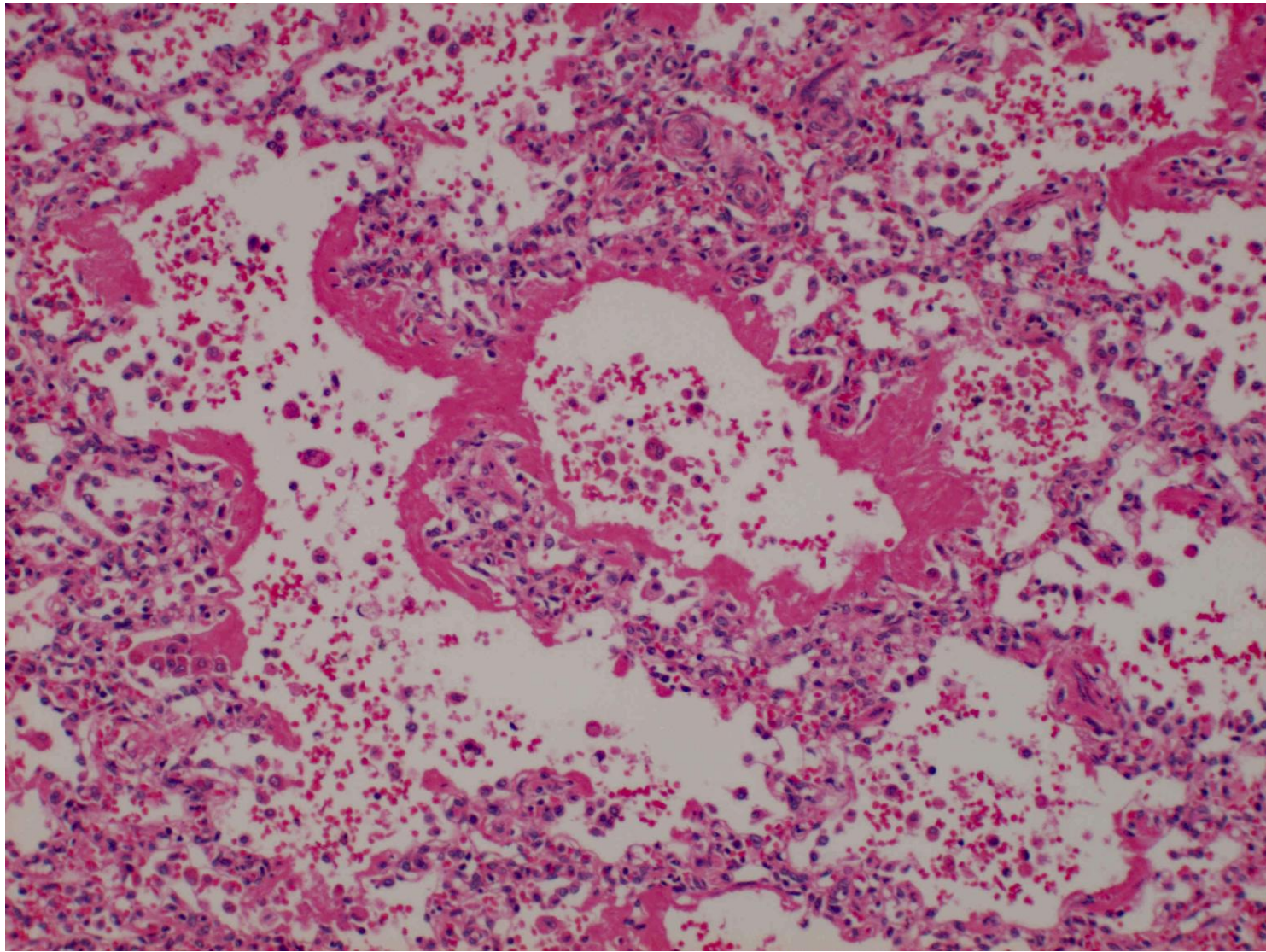
Endocardial fibroelastosis seen in a 7 months-old girl. The pulmonary vein branch reveals intimal thickening with stasis of leukocytes, provoked by increased back pressure due to left heart failure (H&E-a).



Endocardial fibroelastosis seen in a 7 months-old girl. The pulmonary vein branch reveals intimal thickening with stasis of leukocytes, provoked by increased back pressure due to left heart failure (EVG-a).



Endocardial fibroelastosis seen in a 7 months-old girl. The pulmonary vein branch reveals intimal thickening with stasis of leukocytes, provoked by increased back pressure due to left heart failure (left: H&E-b, right: EVG-b).



Endocardial fibroelastosis seen in a 7 months-old girl. The congested lung reveals hyaline membrane formation (H&E-c).