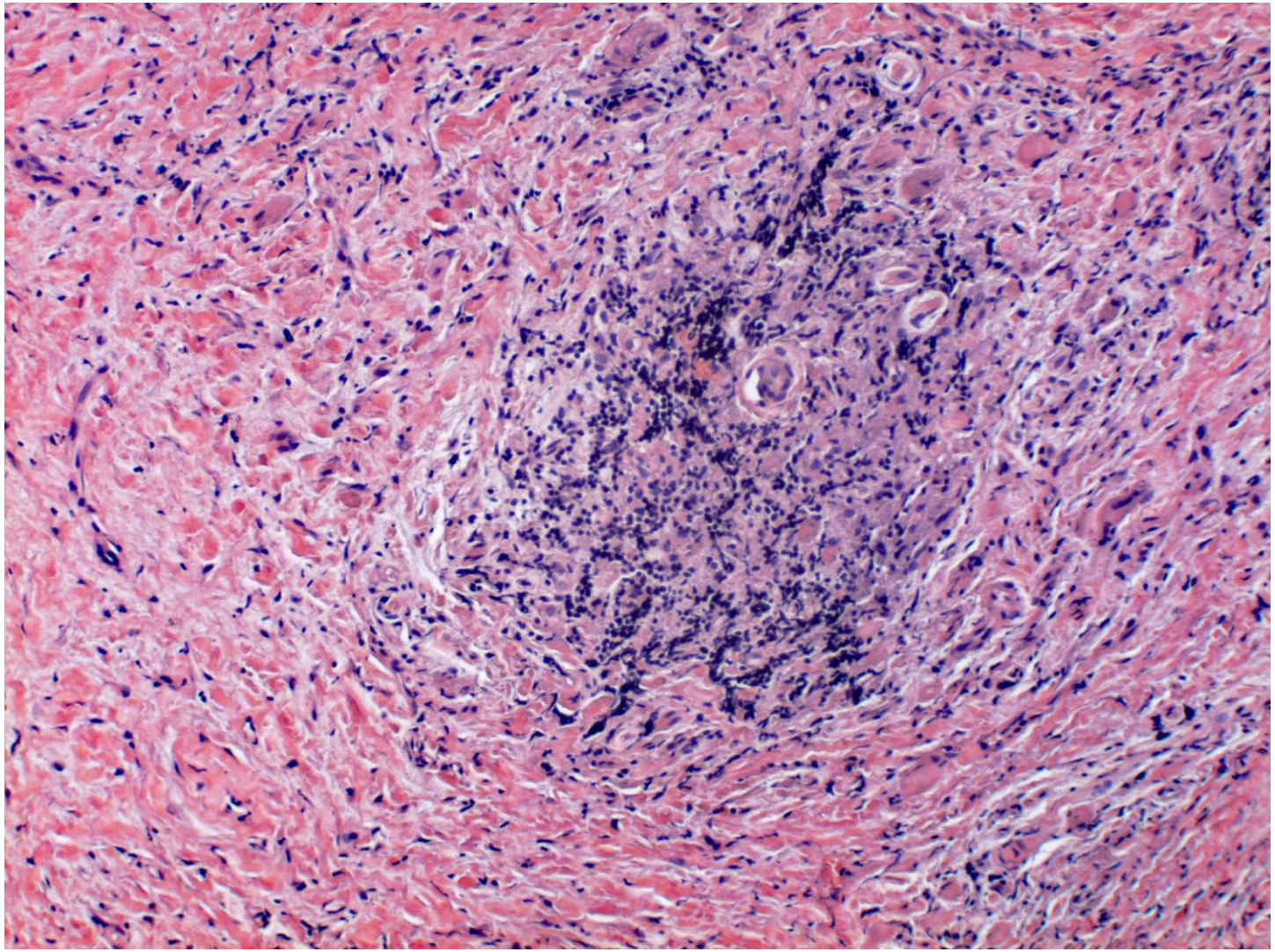
Orbital myositis (ocular myositis)

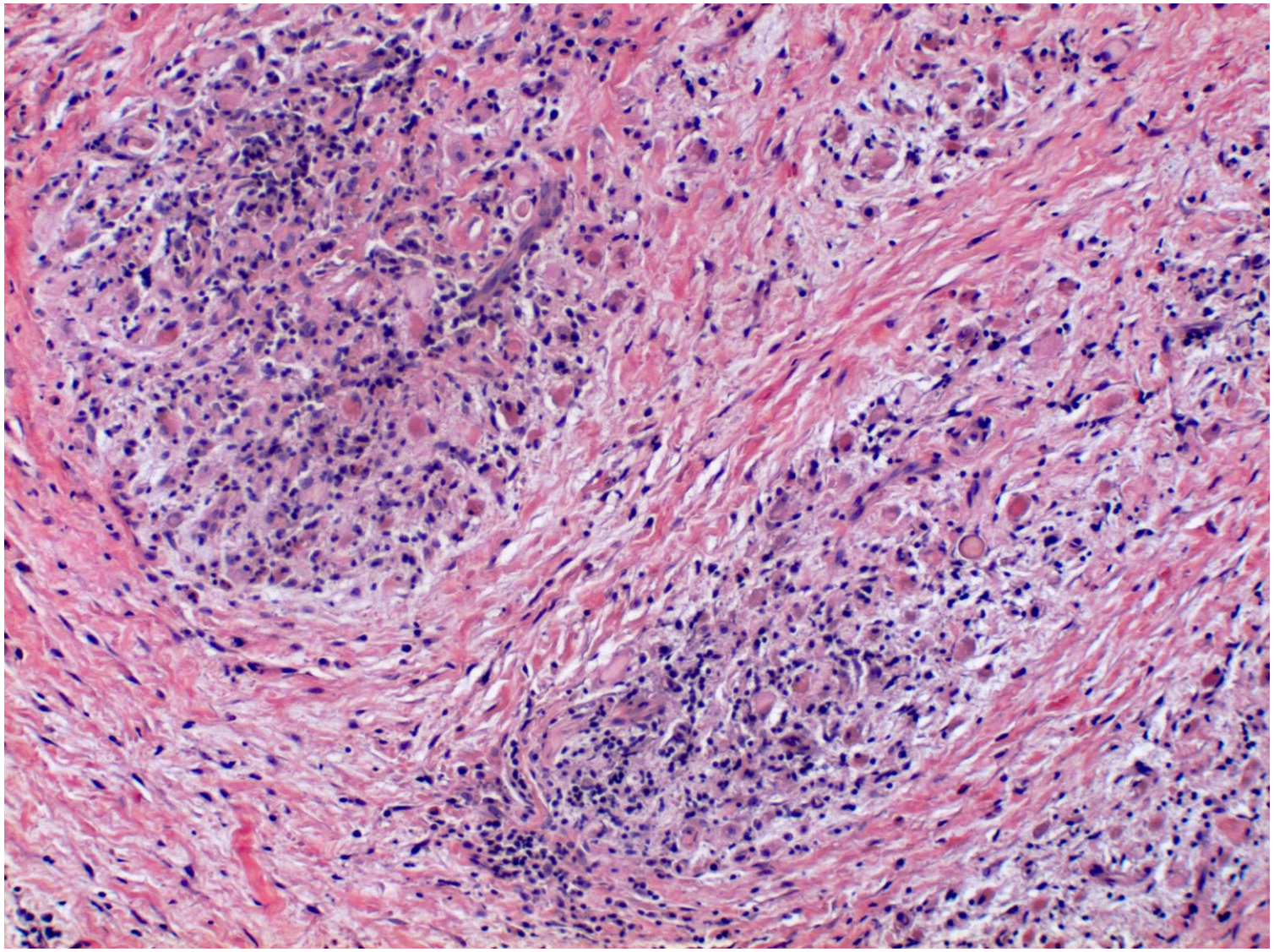
Orbital (ocular) myositis, formerly termed as orbital pseudotumor in the category of idiopathic orbital inflammatory syndrome, is an autoimmune disorder affecting the extraocular muscles of the eye. Unilateral single muscle inflammation with tendon involvement is the most common presentation. Superior, lateral and medial rectus muscles are more often affected than inferior rectus muscle. Orbital myositis manifests with orbital pain and diplopia. It may be complicated with SLE, Crohn's disease and infections of the upper respiratory tract and sinus. Now, orbital myositis is categorized in orbital IgG4-related disease. Microscopically, dense lymphoplasmacytic infiltrate, storiform fibrosis and obliterative phlebitis are observed, and IgG4-positive plasma cells should be richly distributed in the lesion. There should be>10 IgG4+ plasma cells/hpf, or the ratio of IgG4+ plasma cells/IgG+ plasma cells should be >40%. Systemic corticosteroid treatment is quite effective to control the disease.

Ref.-1: Schoser BG. Ocular myositis: diagnostic assessment, differential diagnoses, and therapy of a rare muscle disease. Five new cases and review. Clin Ophthalmol 2007; 1(1): 37-42. PMID: 19668464

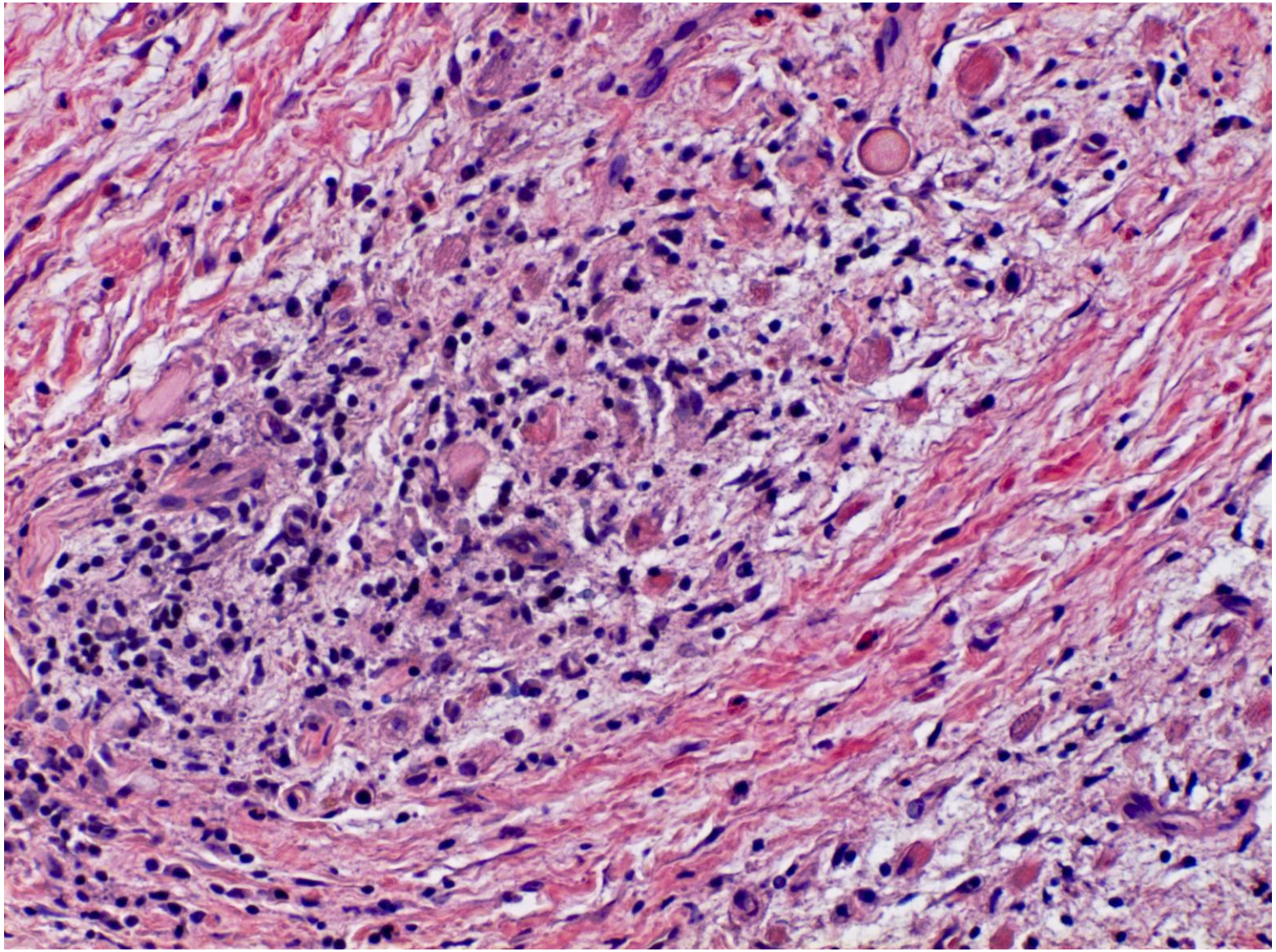
Ref.-2: Kubota T, Moritani S. Orbital IgG4-related disease: clinical features and diagnosis. ISRN Rheumatol 2012; 2012: 412896. doi: 10.5402/2012/412896



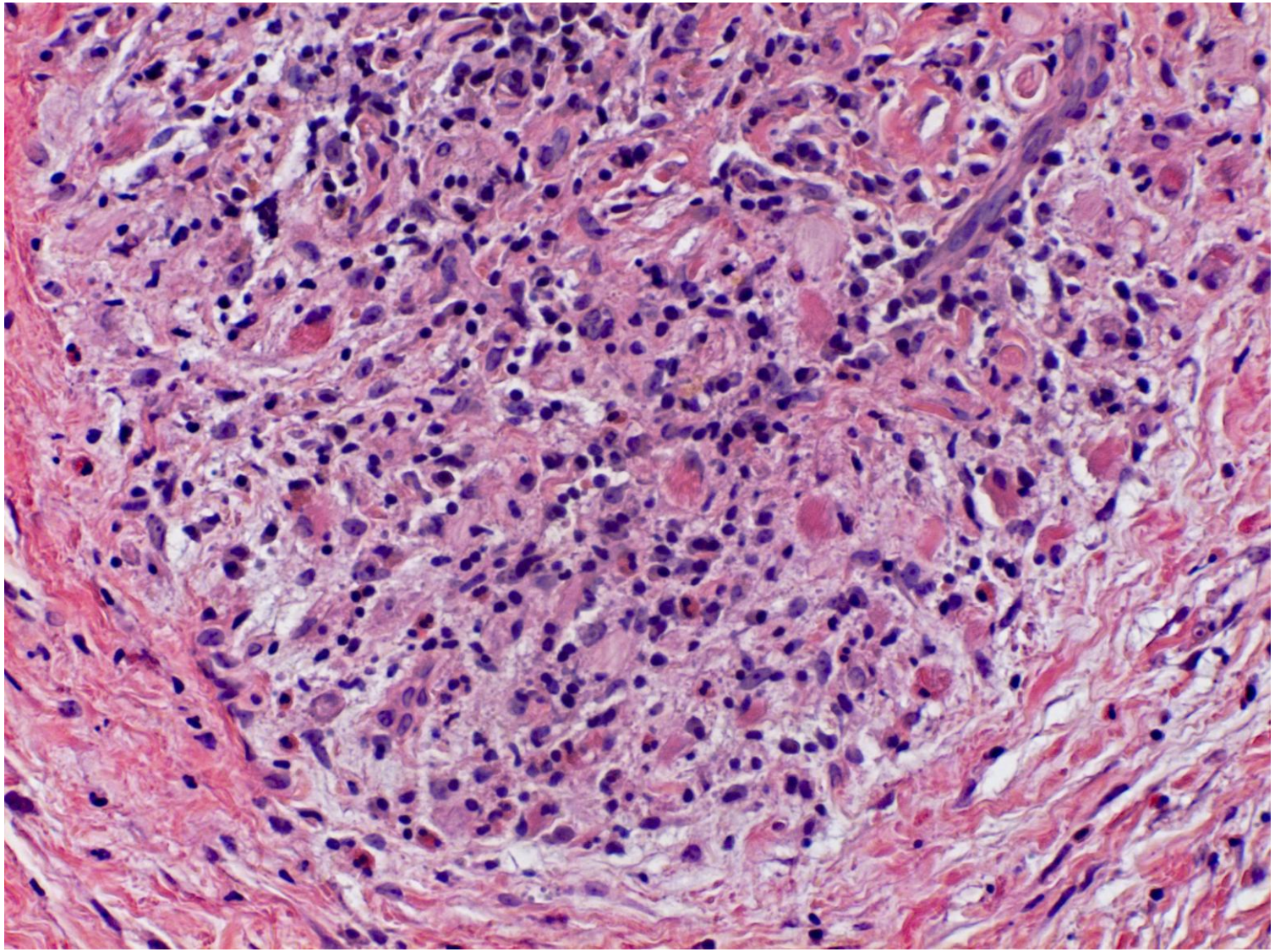
Orbital (ocular) myositis seen in a 59 y-o female patient. The patient presented with an orbital mass with diplopia. Biopsy reveals fibrosing chronic inflammation with lymphoid clustering in the striated muscle tissue (H&E-1).



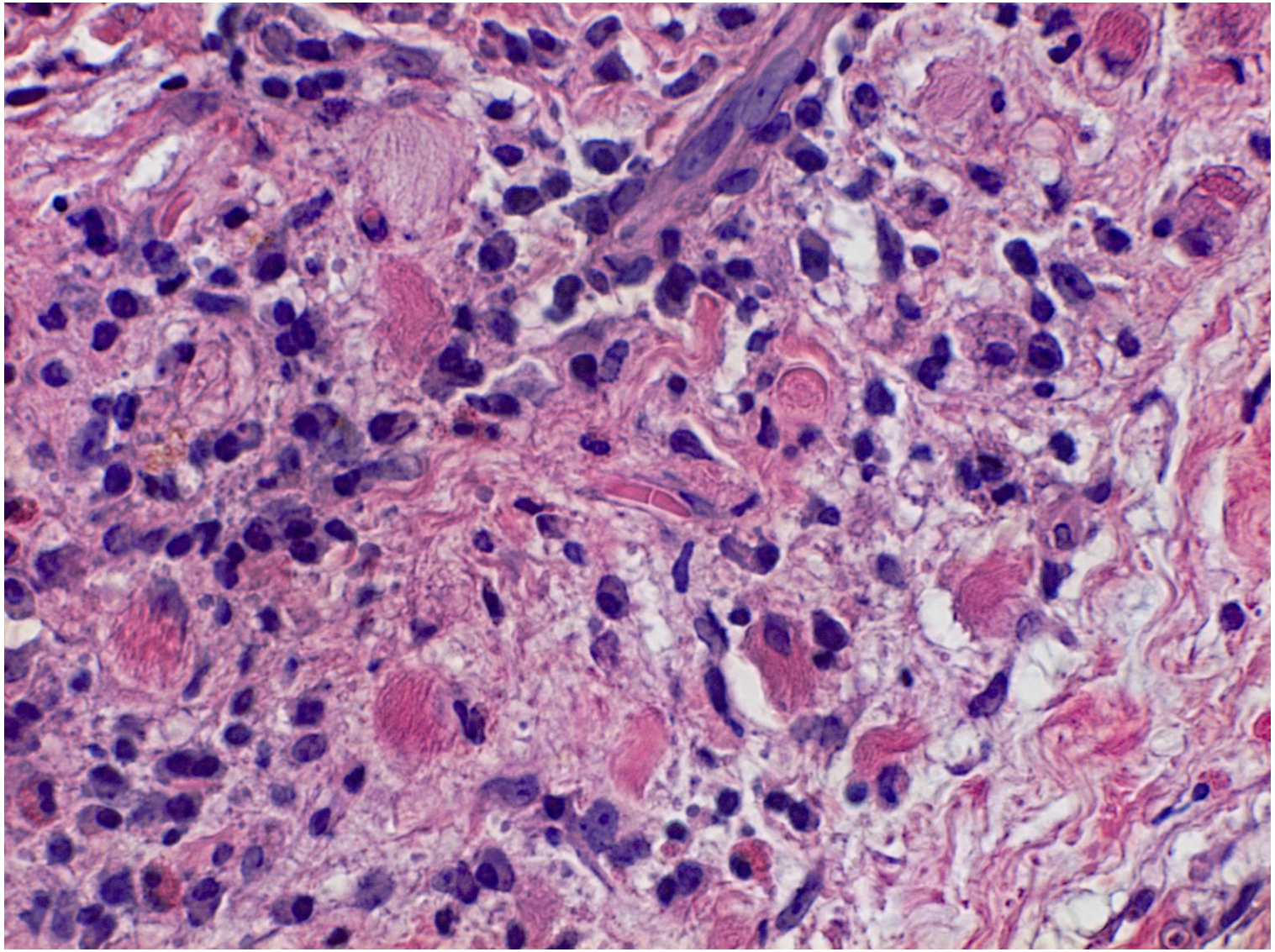
Orbital (ocular) myositis seen in a 59 y-o female patient. The patient presented with an orbital mass with diplopia. Biopsy reveals fibrosing chronic inflammation with lymphoid clustering in the striated muscle tissue (H&E-2).



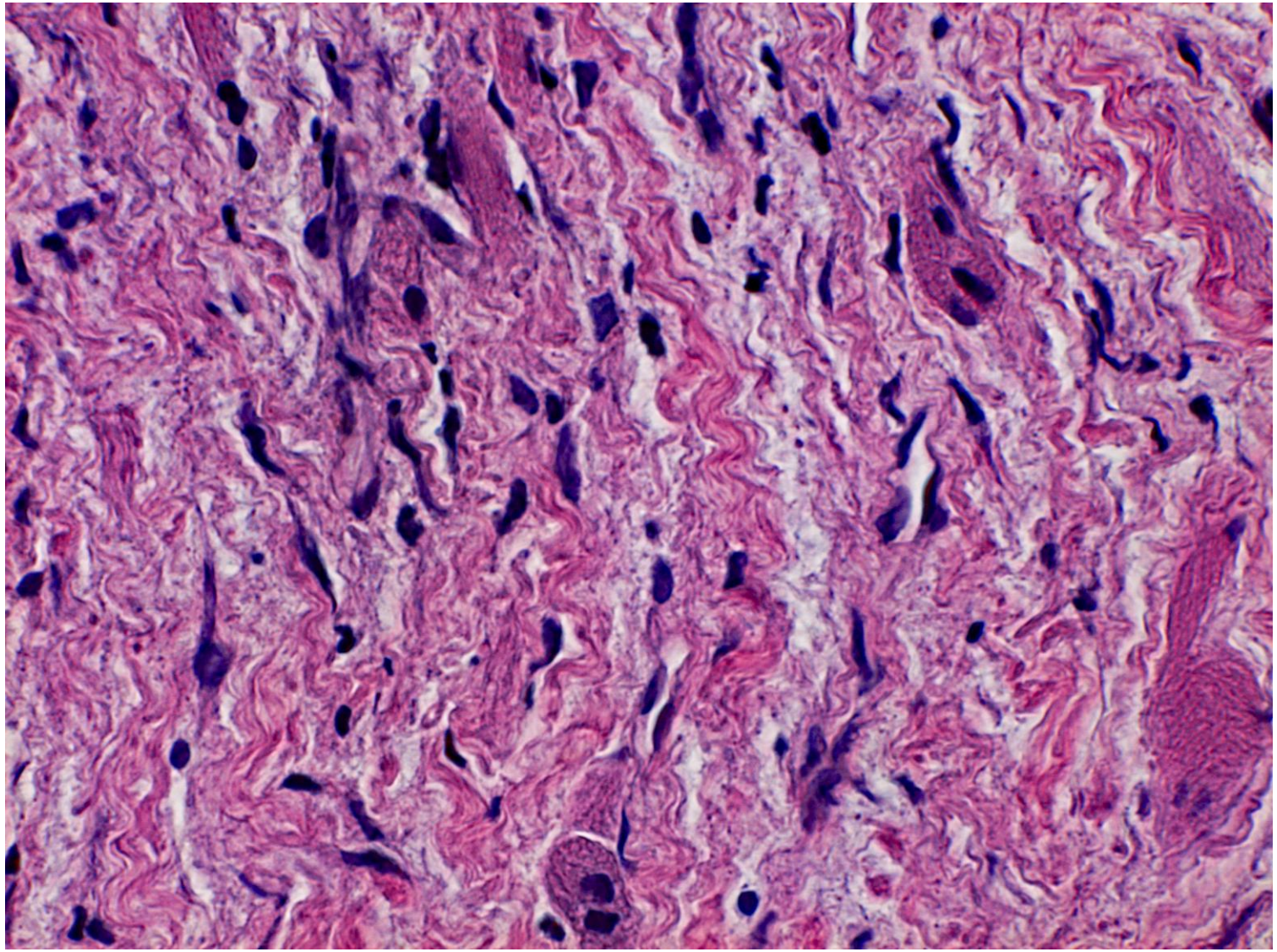
Orbital (ocular) myositis seen in a 59 y-o female patient. The patient presented with an orbital mass with diplopia. Biopsy reveals fibrosing chronic inflammation with lymphoid clustering in the striated muscle tissue. Regenerative muscle cells are intermingled with the inflammatory cells (H&E-3).



Orbital (ocular) myositis seen in a 59 y-o female patient. The patient presented with an orbital mass with diplopia. Biopsy reveals fibrosing chronic inflammation with lymphoid clustering in the striated muscle tissue. Regenerative muscle cells are intermingled with the inflammatory cells (H&E-4).



Orbital (ocular) myositis seen in a 59 y-o female patient. The patient presented with an orbital mass with diplopia. Biopsy reveals fibrosing chronic inflammation with lymphoid clustering in the striated muscle tissue. Regenerative muscle cells are intermingled with small lymphocytes and plasma cells (H&E-5).



Orbital (ocular) myositis seen in a 59 y-o female patient. The patient presented with an orbital mass with diplopia. Biopsy reveals fibroinflammatory change of the striated muscle tissue. Regenerative muscle cells are embedded in the fibrotic stroma (H&E-6).