SARCOIDOSIS

Besnier, Boeck, Schaumann disease

Multisystemic , idiopathic disease , characterized by non-caseating granulomas , spread in different tissues and organs

Diagnosis by exclusion (tuberculosis, fungal infections, berylliosis etc.)

PATHOGENESIS

- Cell mediated immune response to unknown antigens
- Altered immune response to non-identified agents
- Anergy (immune unresponsiveness) to common cutaneous tests
- At peripheric sites : ↓ of T lymphocytes with reduction of CD4/CD8 ratio
- In patients with Sarcoid lungs lesions: ↑ of T lymphocytes and increased (↑) CD4/CD8 ratio in bronchoalveolar lavage fluid.
- In serum : ↑ of Polyclonal IG (no ↑ of B lymphocytes)

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Involved sites

- Lymph nodes (neck, hylum of lungs)
- Lungs
- Joints (lower limbs arthritis)
- Skin (face, sup. trunk = erythema nodosum)
- Salivary and lacrimal glands
- Eyes (iridociclytis, choroiditis, retinitis)
- Oral mucosa
- Spleen, liver etc.
- Bones (multiple, cystoid osteitis)

Løfgren syndrome

Mikulicz syndrome

Heerfordt syndrome

Jungling syndrome

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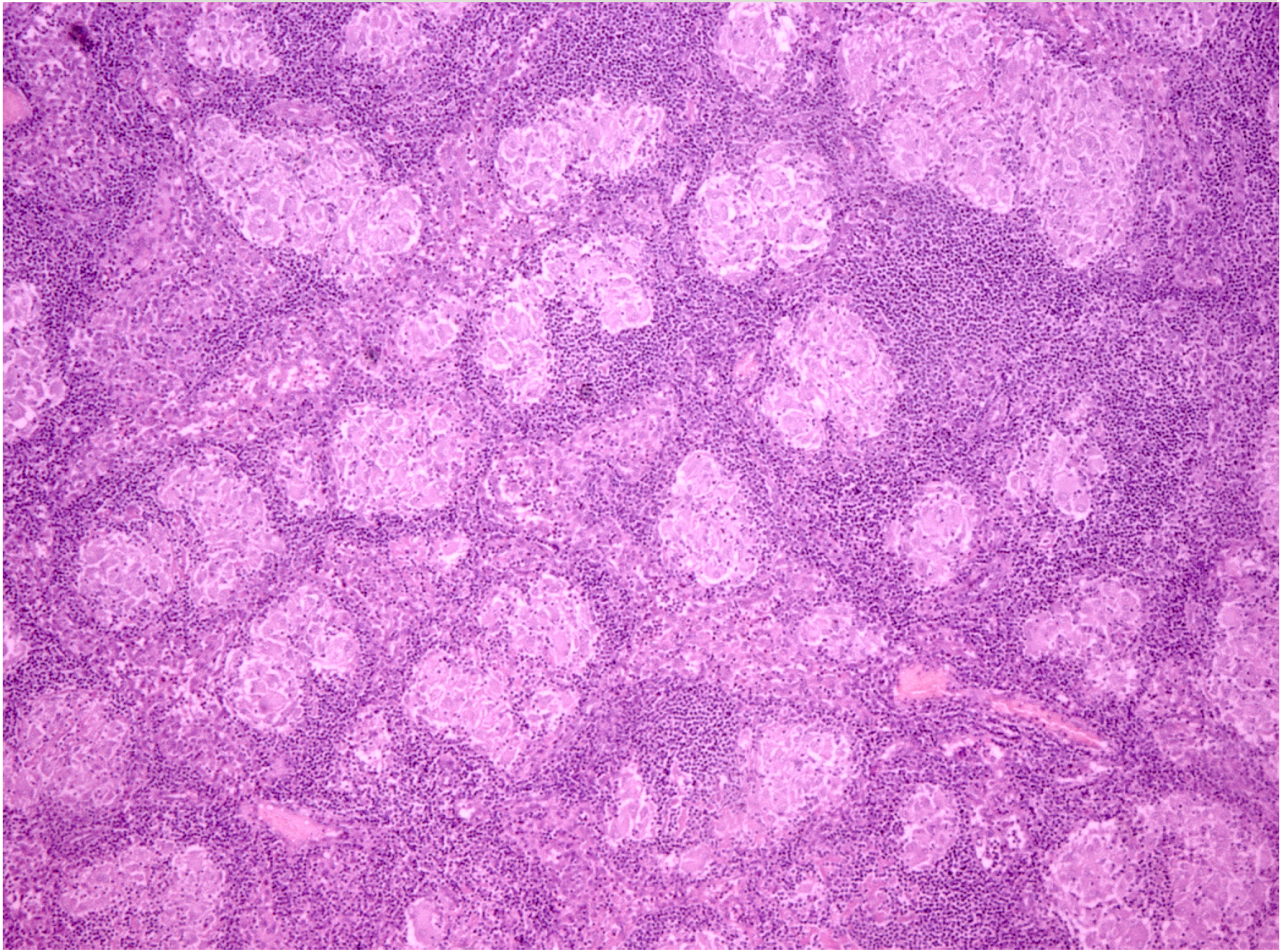
MACROSCOPIC FEATURES:

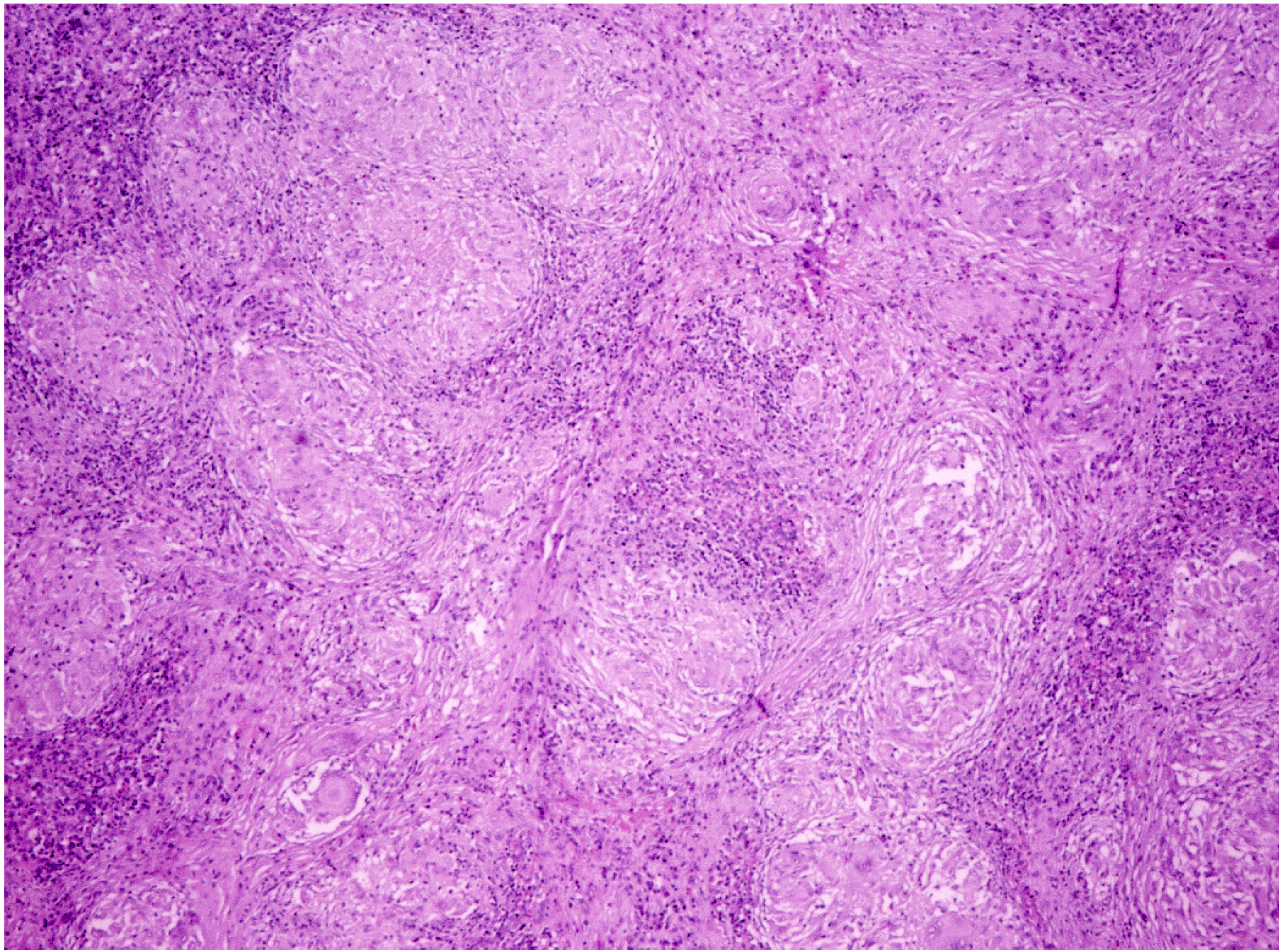
Deep sites: nodular lesions, sharply demarcated, non-confluent with well defined borders

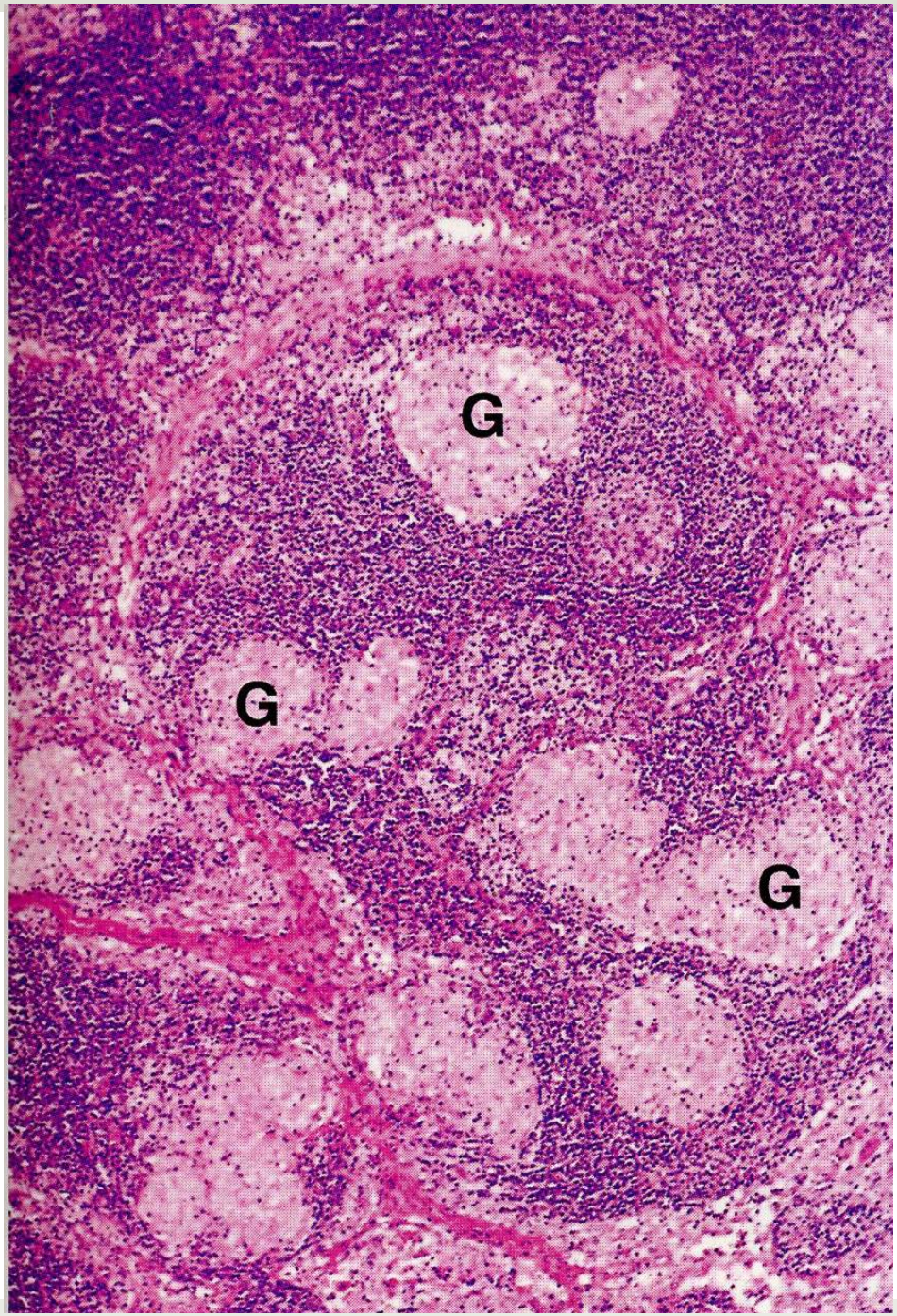
Skin and mucosal sites : subcutaneous/mucosal nodules or purplish hard plaques

MORPHOLOGY OF SARCOID GRANULOMA

- Nodules of tightly packed epithelioid cells
- Multinucleated giant cells, similar to Langhans' cells or foreign-body type giant cells
- In the periphery, T-helper CD4+ lymphocytes
- Lack of central necrosis, sometimes central area of hyaline sclerosis
- Possible evolution into Scleroderma (hardened or scarred tissue)
- Inclusion bodies (Schaumann bodies): concentrically laminated inclusions made of calcium and proteins
- Asteroid bodies



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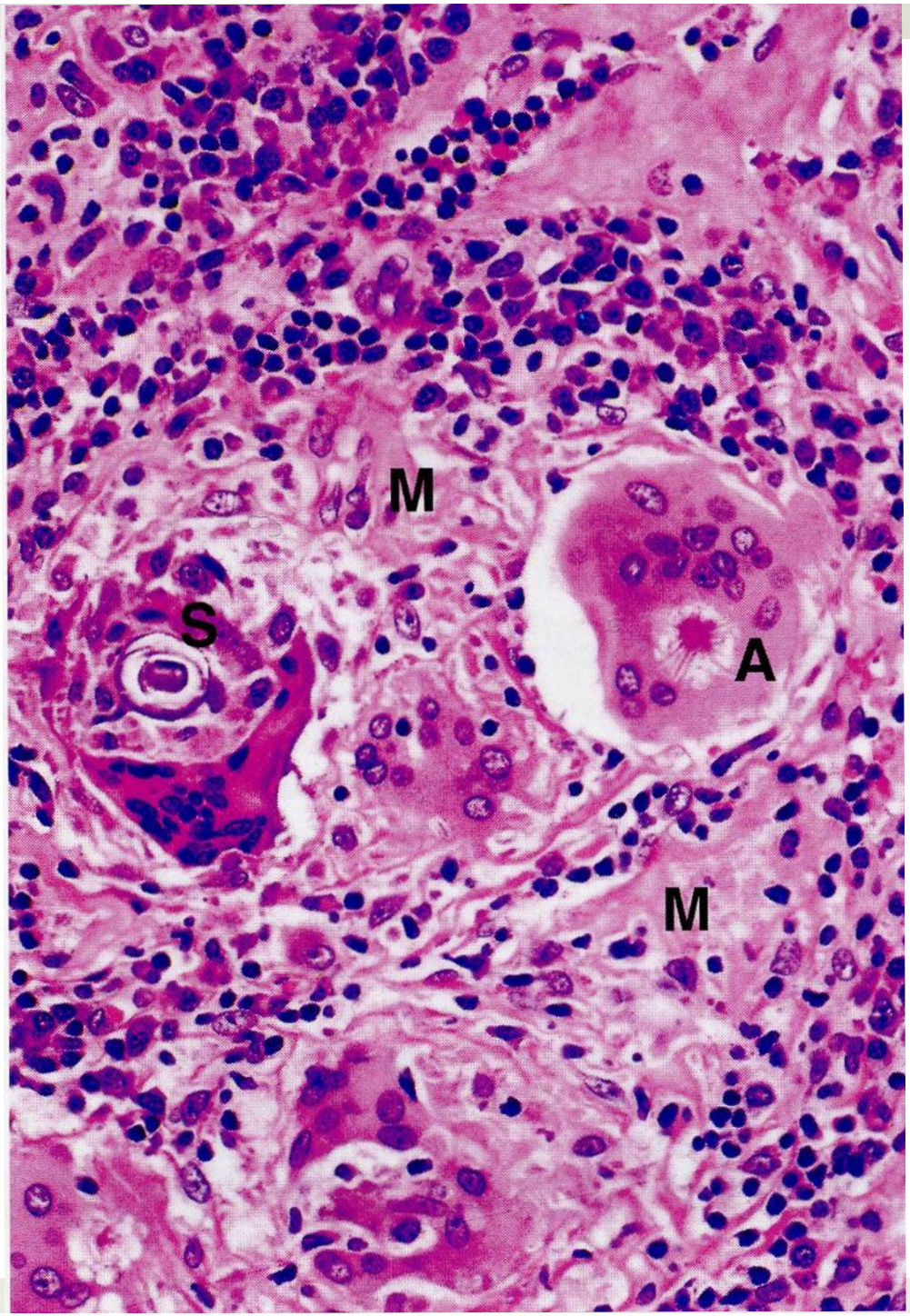
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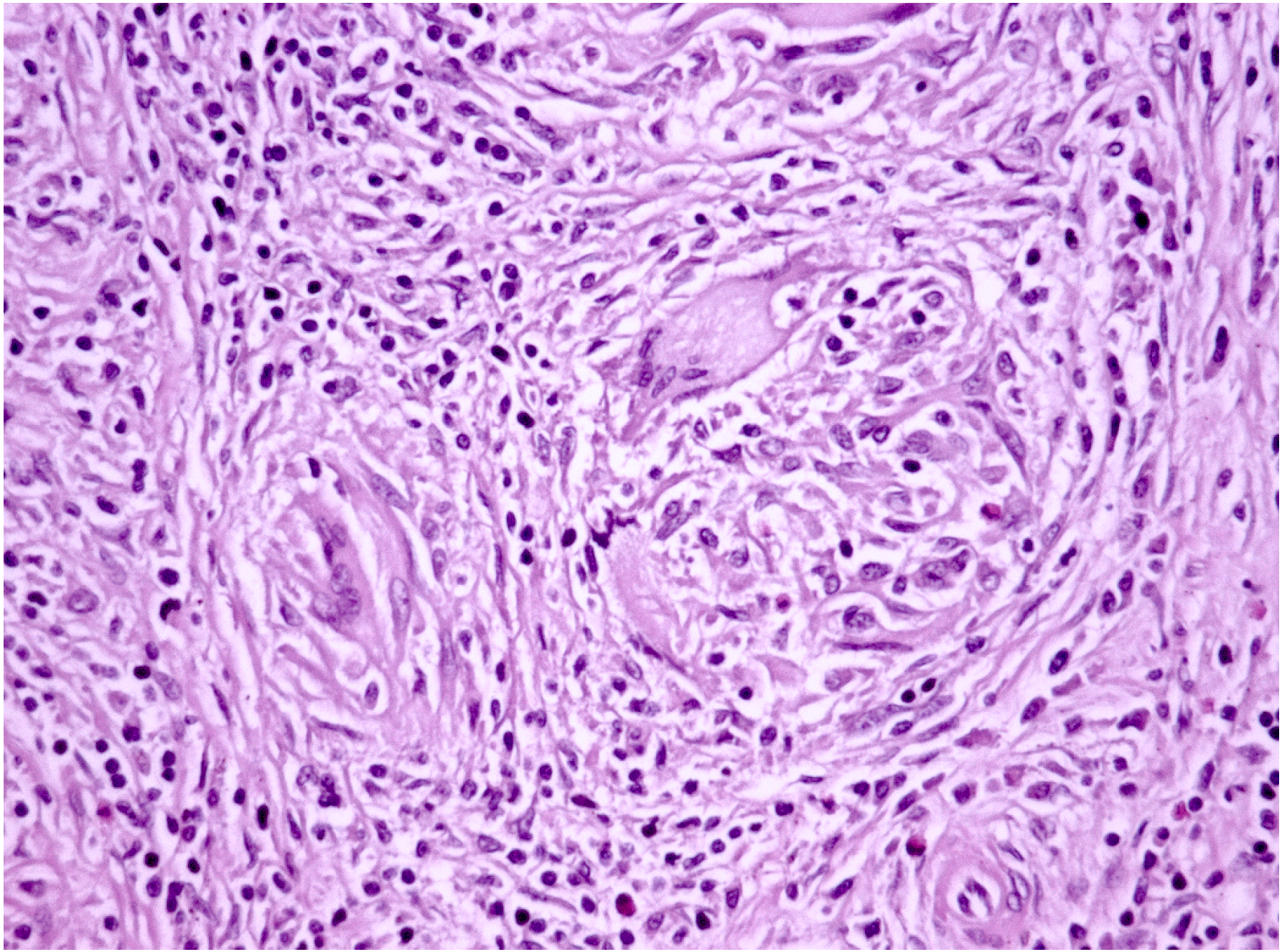

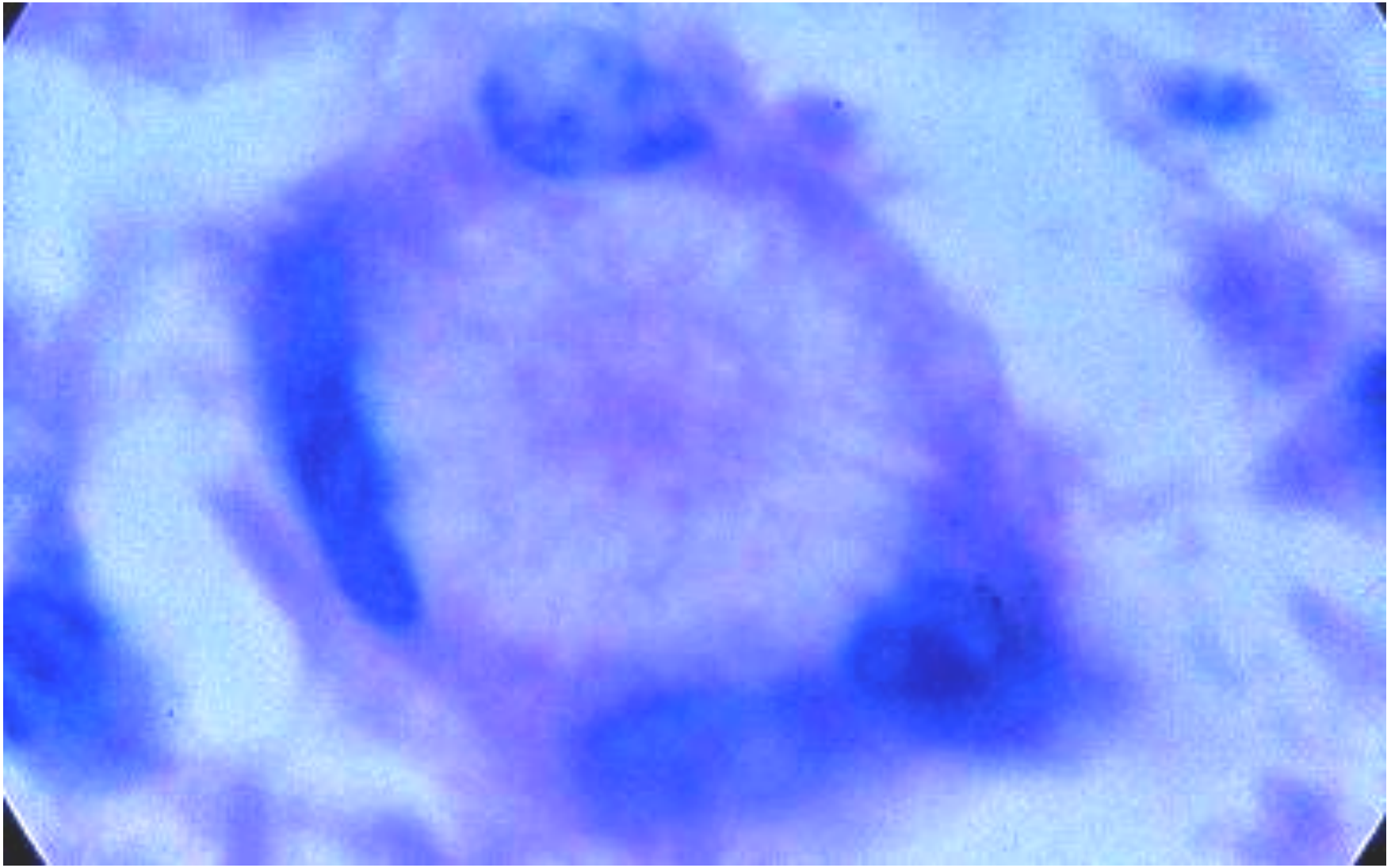

EVOLUTION AND COMPLICATIONS

- 65% = Healing with few sequelae
- 20% = Chronic respiratory insufficience, visus decrease
- 10% = progressive course (pulmonary fibrosis,
- Pulmonary Heart disease)