PILOCYTIC ASTROCYTOMA

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A. Definition:

- Synonyms: Juvenile Pilocytic Astrocytoma, Piloid "cystic" or "childhood" Astrocytoma
- Slow growing, benign (WHO grade 1) Astrocytic tumor
- Well circumscribed midline CNS lesion in children
- Grossly cystic with mural nodule, biphasic histology

B. Clinical issues:

I. Epidemiology:

1. Incidence:

- 6% of intracranial tumor < 1 per 100.000/ years
- Most common pediatric primary brain tumor (30%)
- 2nd after embryonal tumor in infants

2. Age:

- Peak incidence: 8 – 13 years old; > 70% in < 20 years old
- Cerebral / spinal lesions tend to manifest at older age

3. Site:

- Mostly axial (ventricle, midline through out neuraxis)
- 12 – 18 % cerebellum (cerebellar astrocytoma)

- Secondary invasion of brainstem in up to 30%
- 8 – 20% in cerebral hemispheres
  - Especially in medial temporal lobe
- 3 – 5% in optic nerve (“optic nerve glioma” chiasm)
  - Often extends into hypothalamus 3 rd ventricle
- 3 -6% in brainstem (“brainstem glioma”)

- - Often dorsal / exophytic; in medulla / midbrain
- - Spinal cord, thalamus, basal ganglia infundibulum

4. Presentation:

- Long-time symptom prior to presentation/ diagnosis
- Neurological symptoms depend on tumor sites
- - Cerebellum: ↑ ICP, CSF obstruction, hydrocephalus, papilledema, headache, ataxia, nausea vomiting
- - Optic nerve: visual acuity / field defects, proptosis

- - Hypothalamus: endocrine, electrolyte imbalance, diabetes insipidus, precocious puberty

- - Brainstem: cranial nerve deficits, CSF obstruction
- - Supratentorial: mass effect, ↑ ICP, seizure/epilepsy
- - Thalamus/basal ganglia: ↑ ICP, weakness/paresis

C. Image findings:

I. MRI findings:

- Hypo to iso intense on T1, hyper intense on T2

II. CT findings:

- Decrease from iso to hypodense, contrast – enhancing mass
- Lack of peritumoral edema, uncommon calcifications

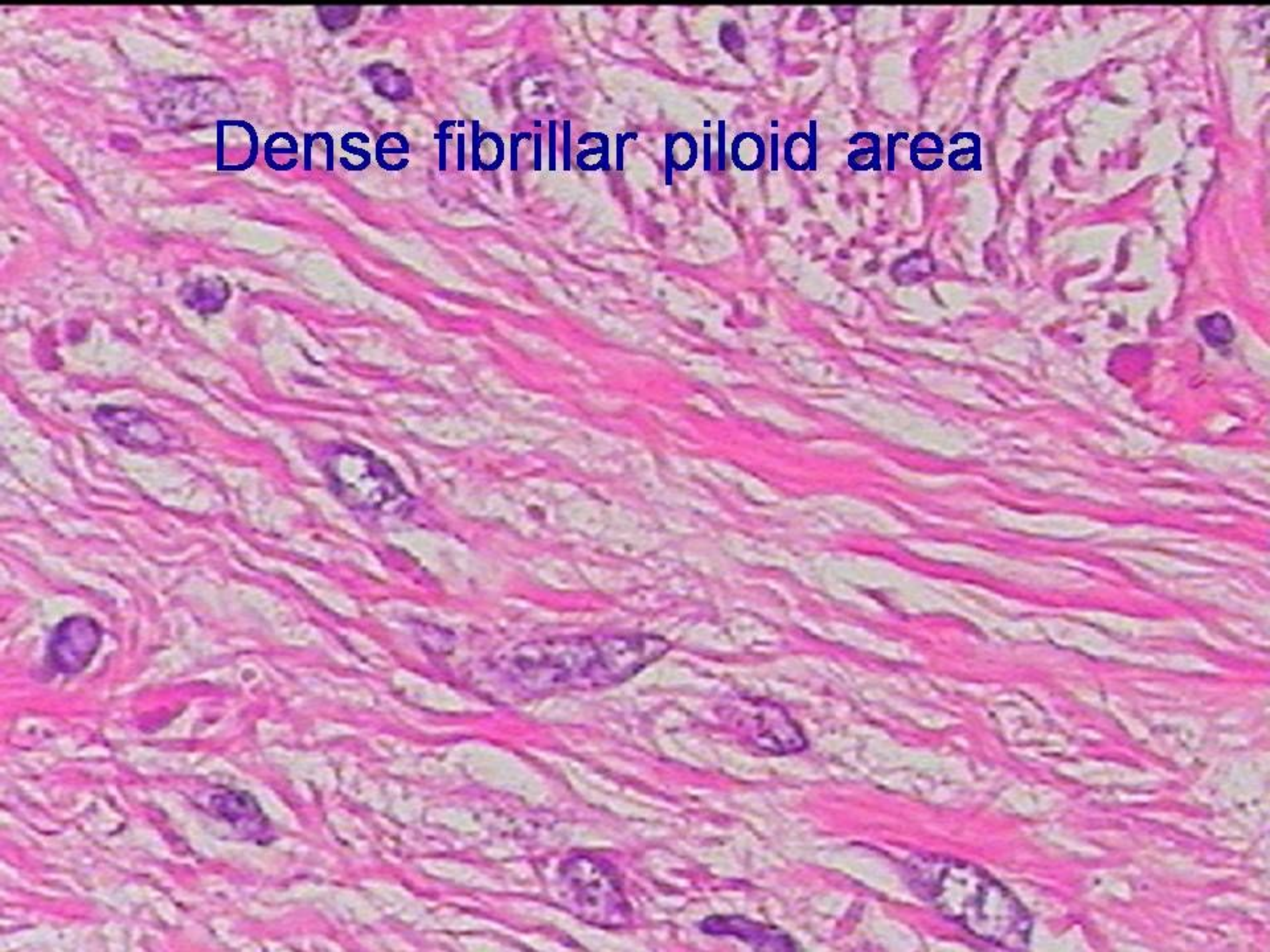
- Round or oval cystic lesions with mural nodules (especially cerebellar, supratentorial lesions)
- Fusiform enlargement ("pencil shaped glioma")
- Longitudinal growth within spinal cord, optic nerve

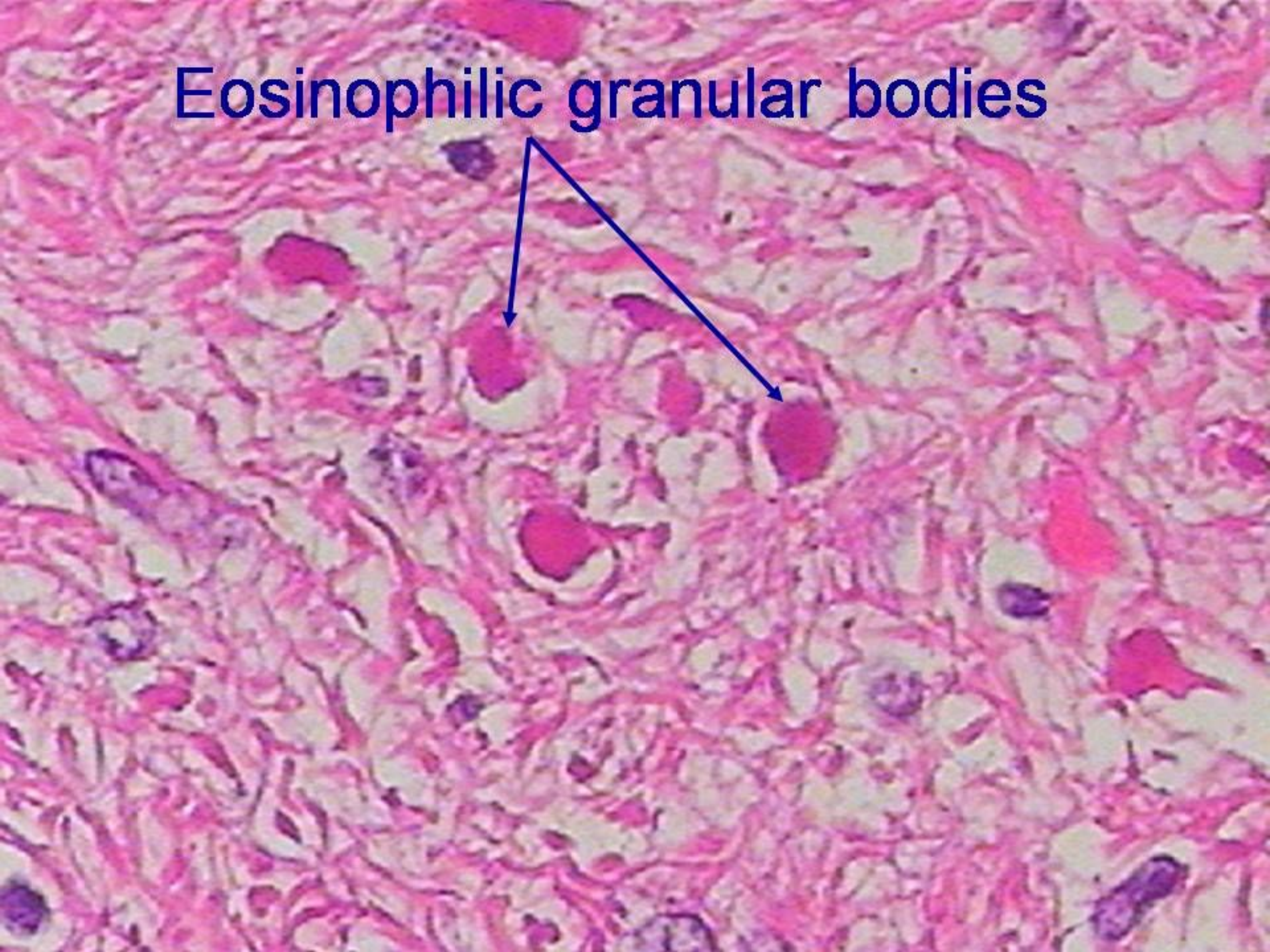
D. Pathology:

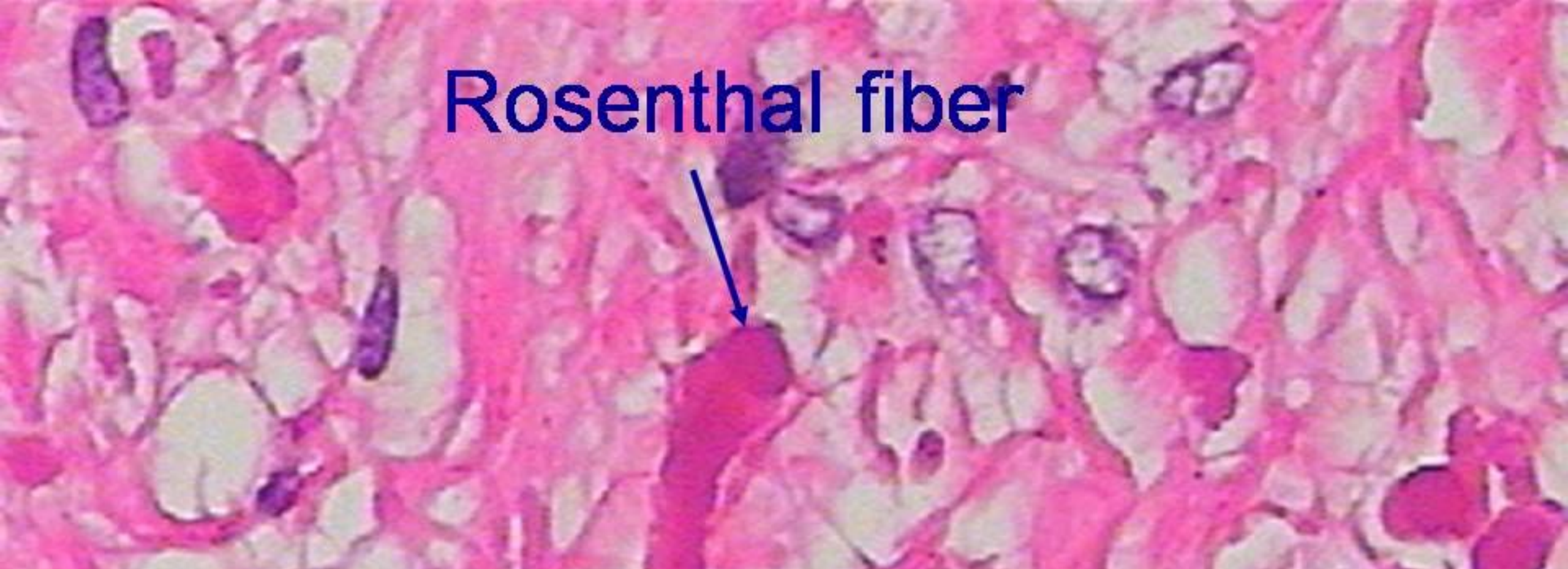
I. Macroscopic features:

- Well delineated expansive rather than infiltrative
- Gray pink, soft with cysts, mucoid degeneration

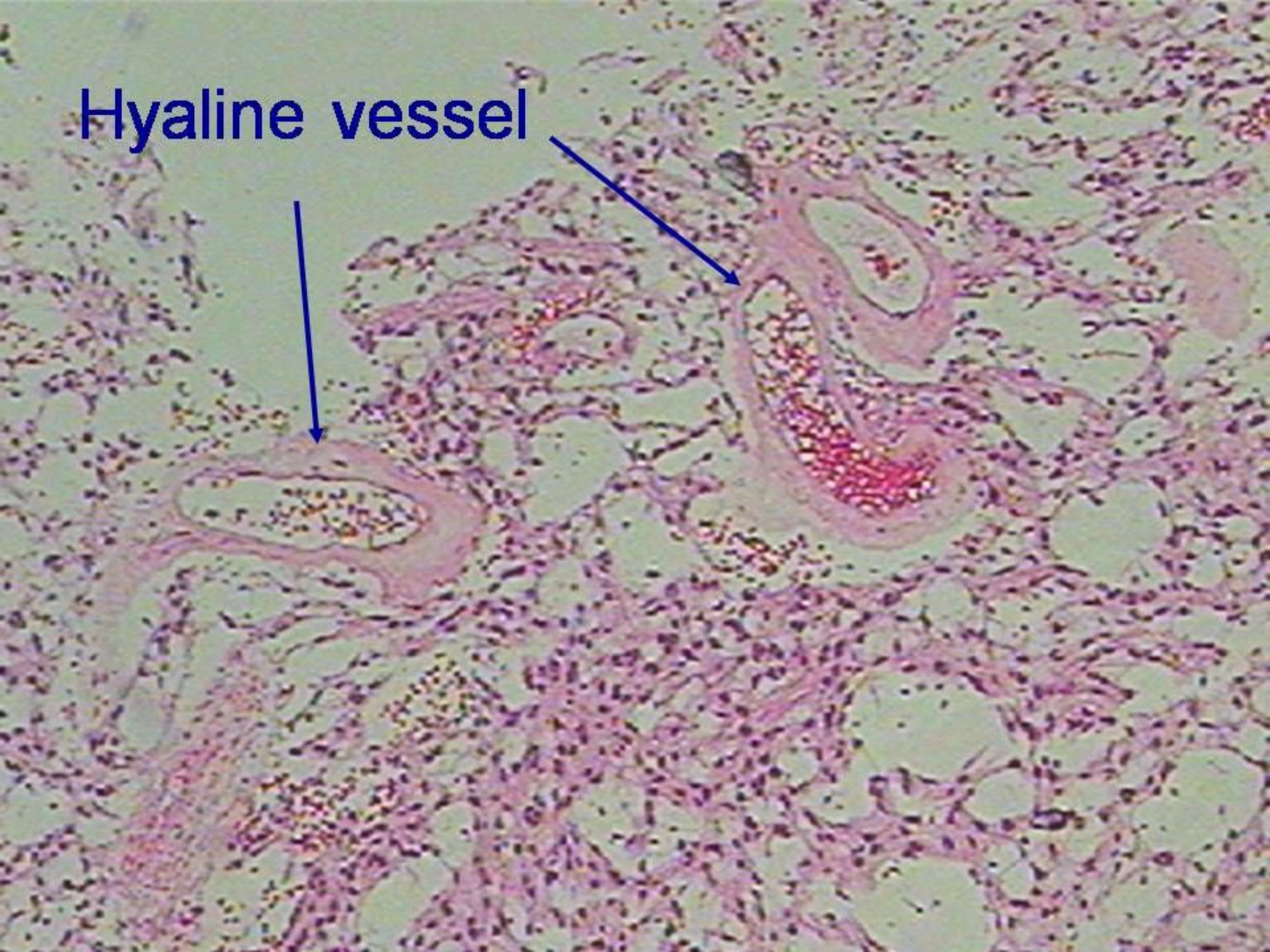
II. Microscopic features


Dense fibrillar piloid area


Eosinophilic granular bodies


Rosenthal fiber

Microcystic area


Hyaline vessel

E. Treatment:

I. Surgical approaches:

- Complete surgical resection is the goal even though it may be difficult due to location of lesion

II. Adjuvant therapy:

- No evidence for efficiency of radiation on survival

- Chemo therapy for progressing, unresectable lesion

F. Prognosis:

- Long period of recurrence (> 20 years):

  - Depends on completeness of surgical resection

- >80% survival 5-10 years, regardless of location
- Reaches to 100% after complete surgical resection

Thanks for your attention