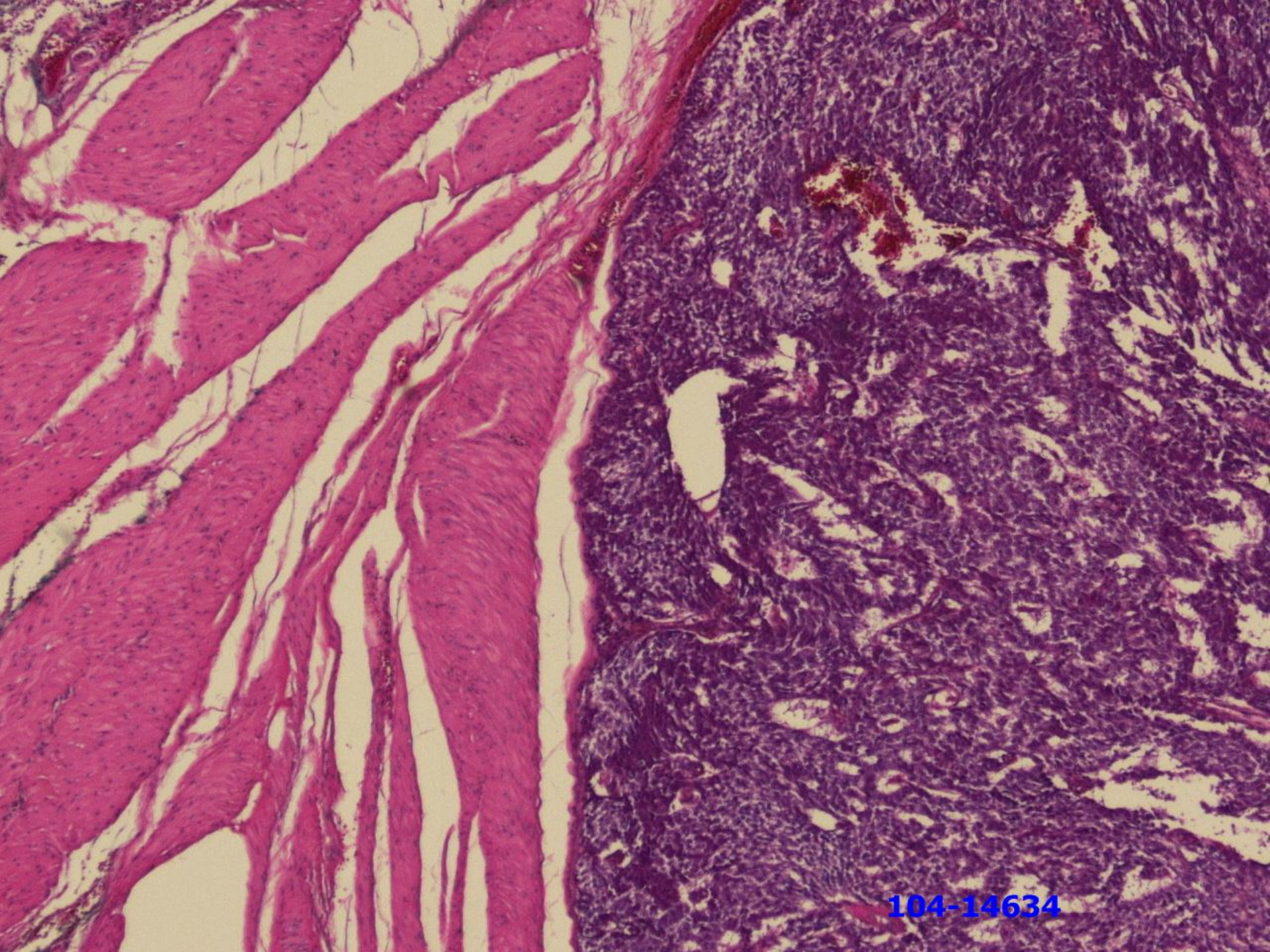


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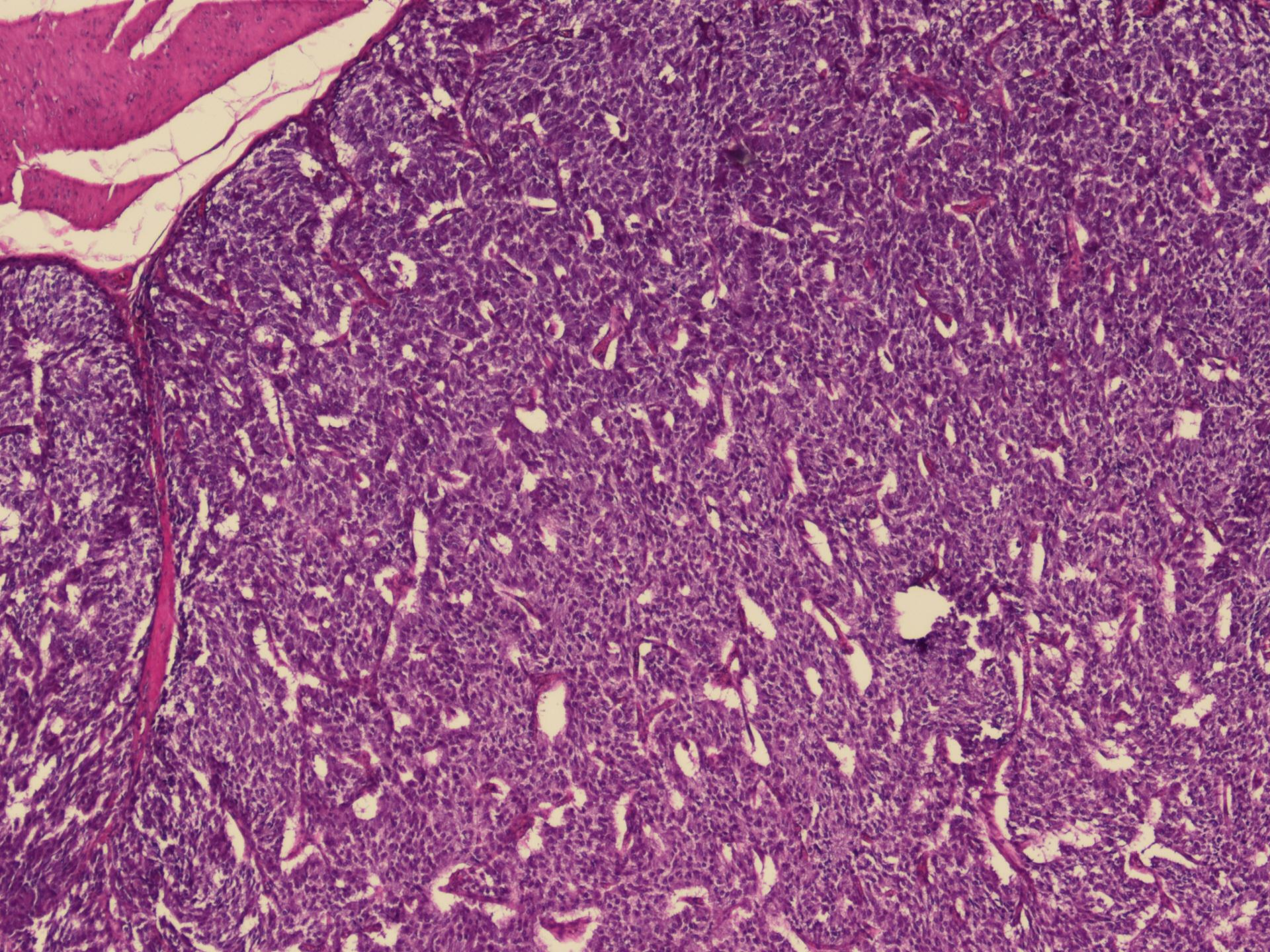
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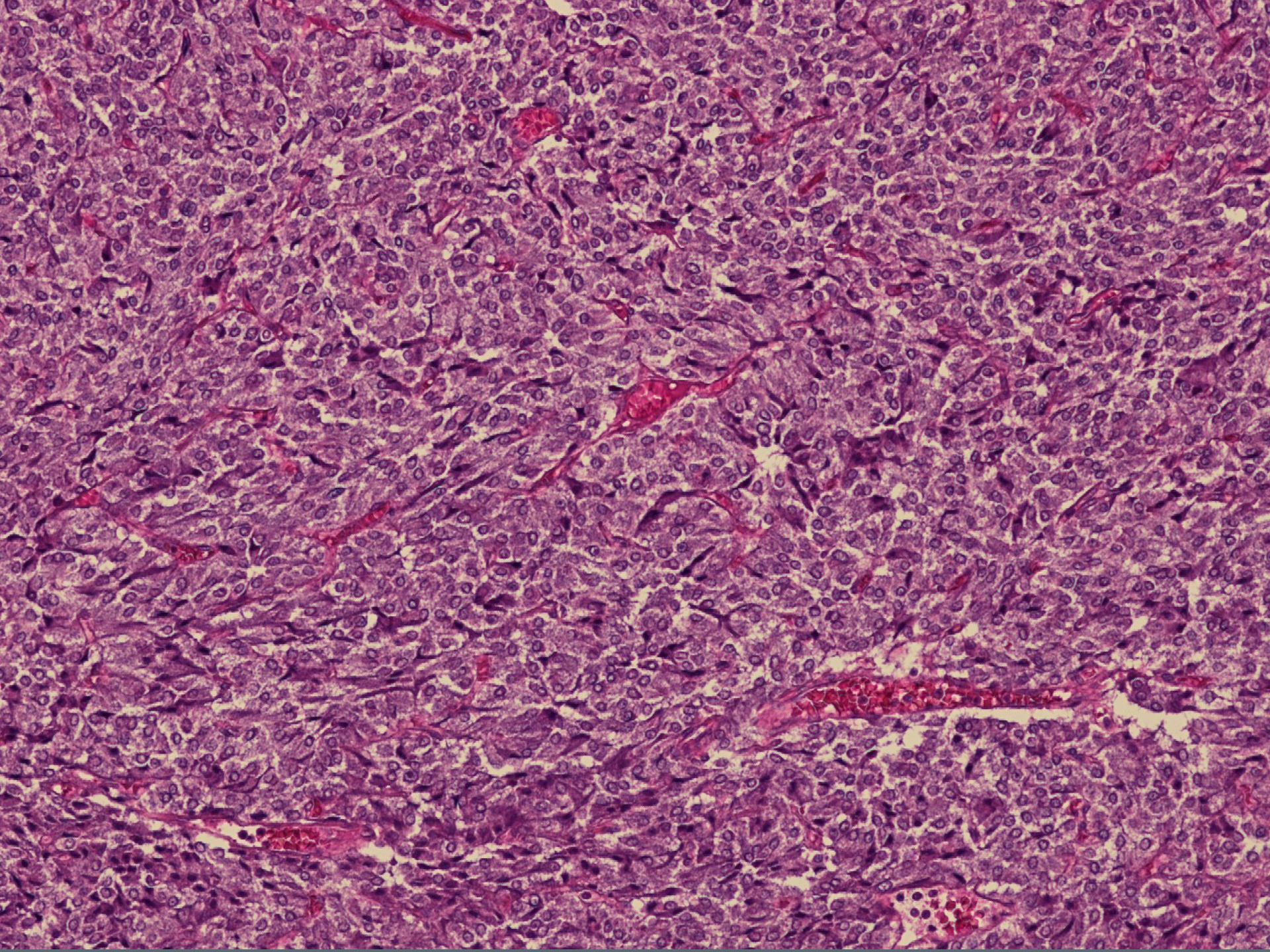
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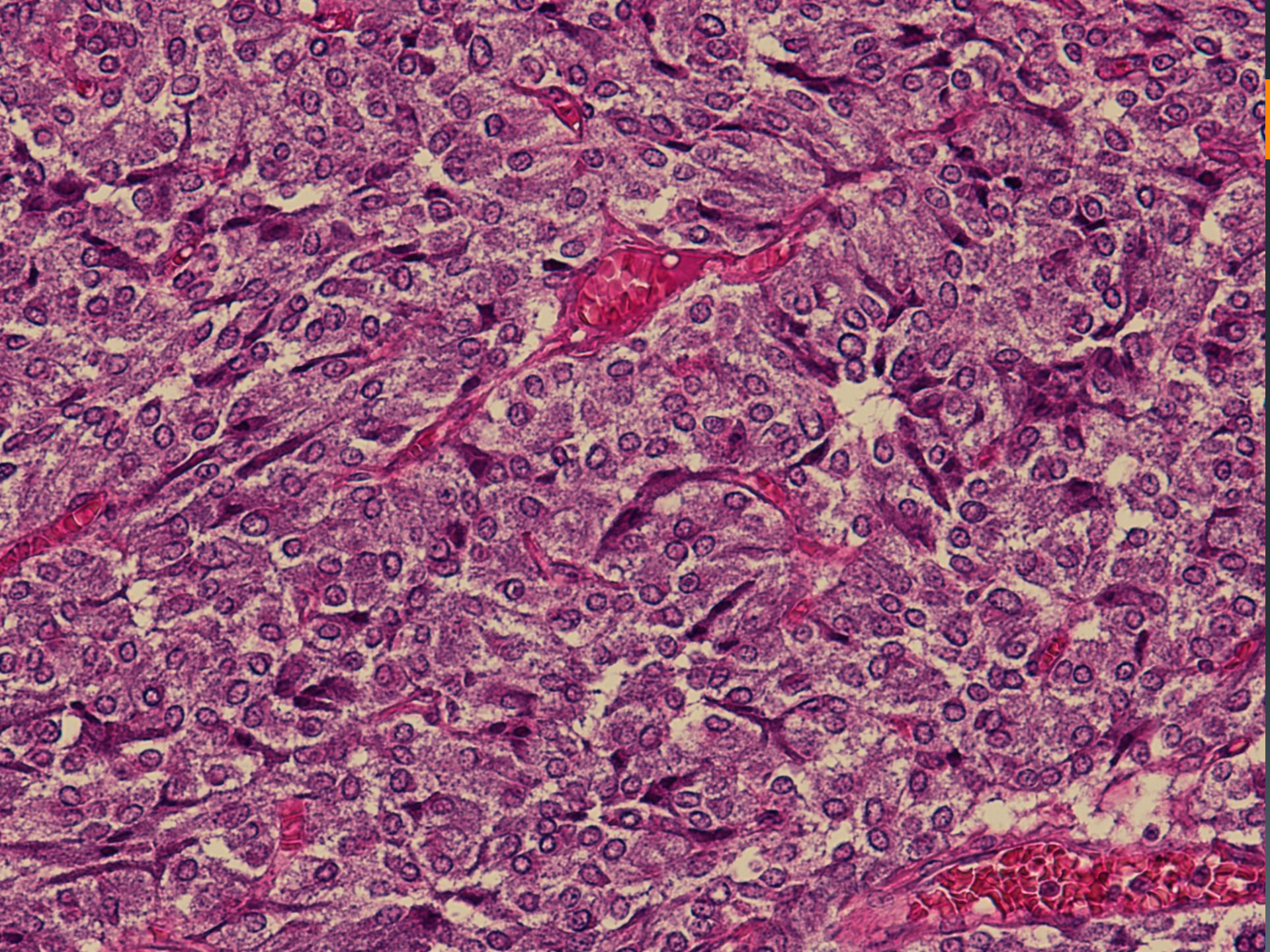
Male, 46 y/o, urinary bladder tumor

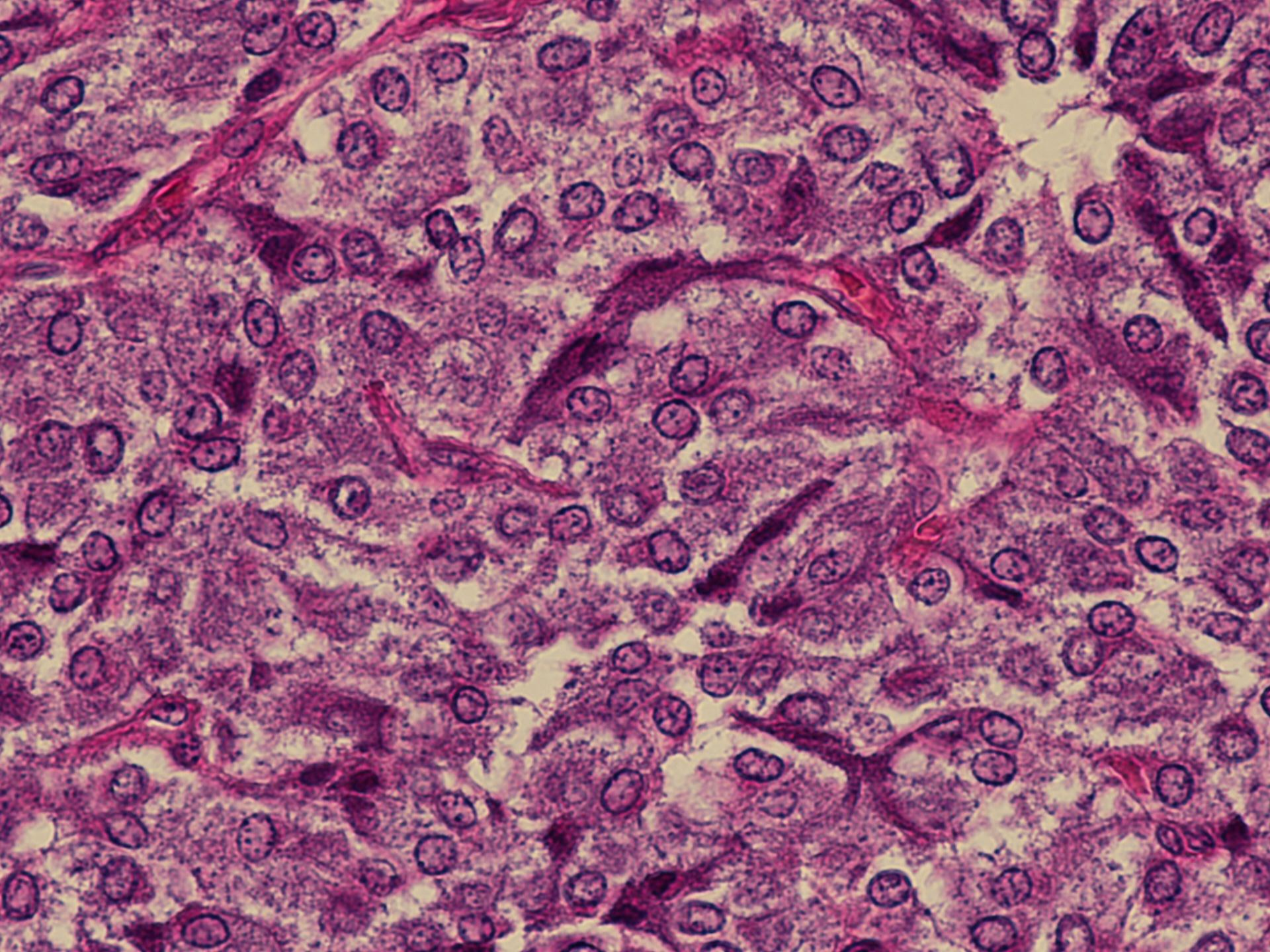


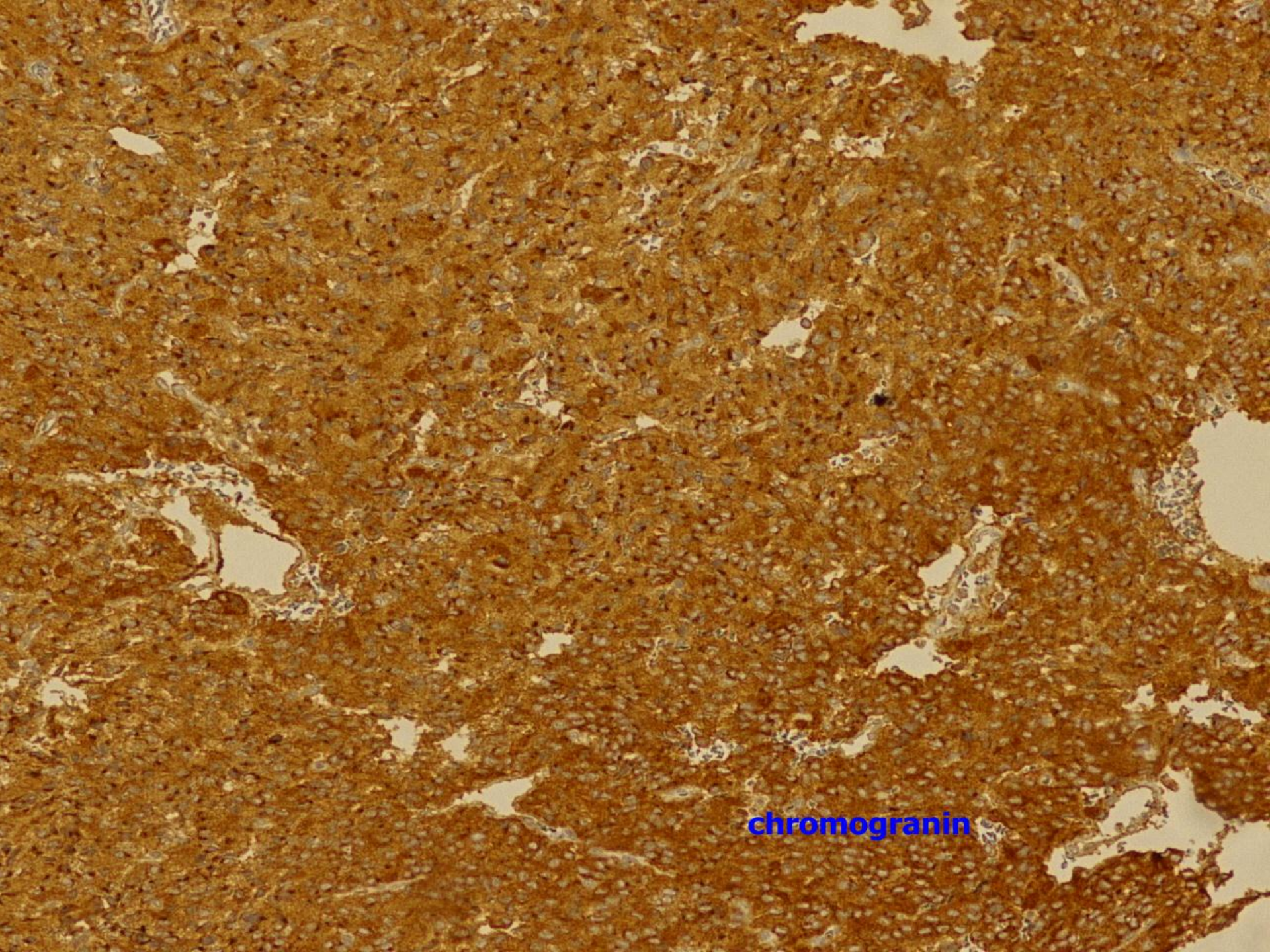
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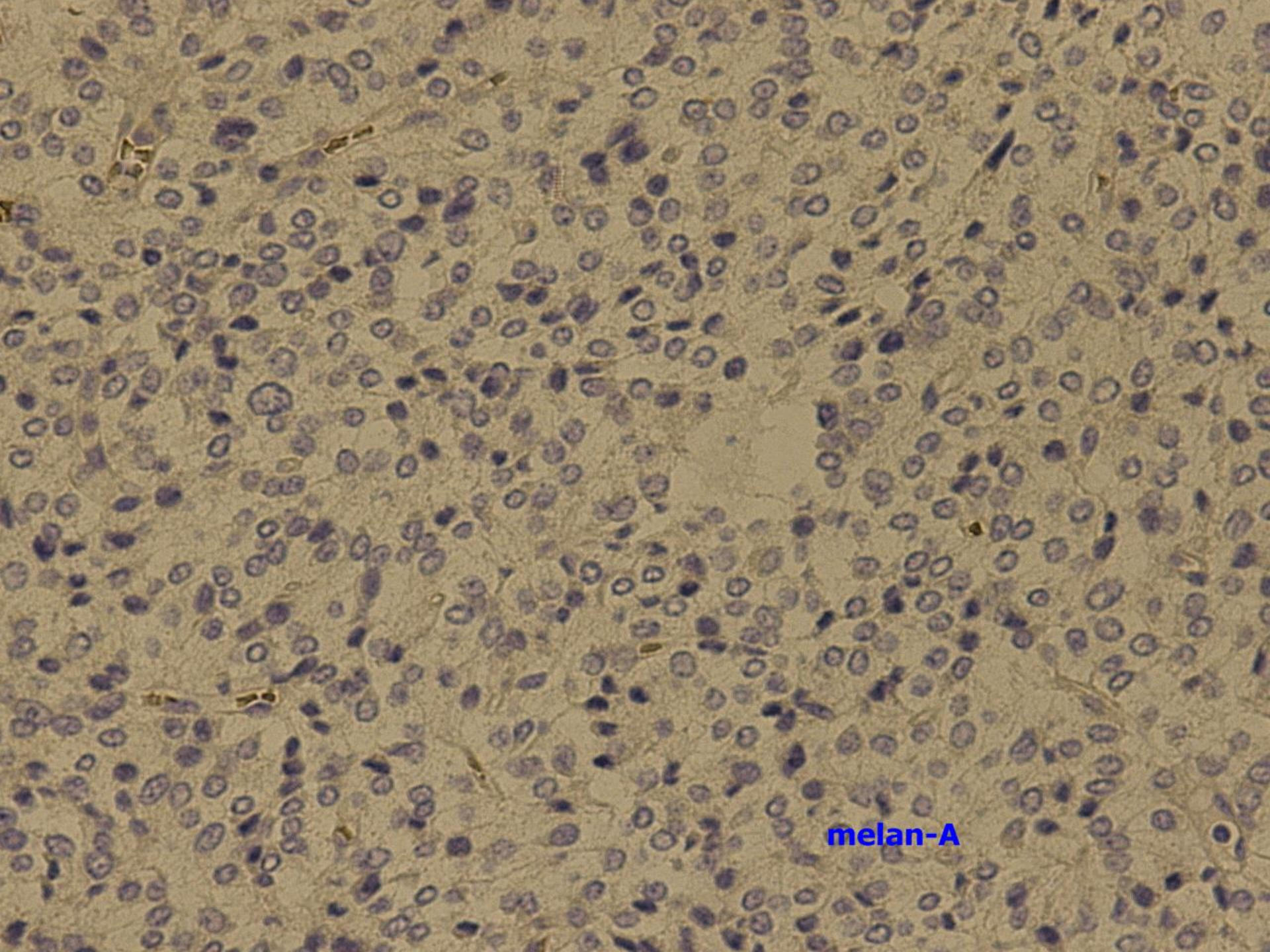








chromogranin



melan-A



Urinary Bladder

Paraganglioma



Definition

- Tumor derived from paraganglion cells in the bladder wall.
- Histologically identical to paragangliomas at other sites



Epidemiology

- Rare tumor, 200 cases by 1997
- Uncommon in bladder (0.06% to 0.1% of all bladder tumors)
- Age range 10-88 years (mean, 45 y/o)
- Median age 41 years vs. 70 years for urothelial carcinoma
- Male to female ratio 1 : 1.4

Clinical Features

- Triad: Sustained / paroxysmal hypertension
Intermittent gross hematuria
Micturition attack
- 2/3 pts: hypertension
- 50% have hematuria
- 50% have “micturition attack” during urination with full bladder, consisting of bursting headache, anxiety, tremulousness, pounding sensation, blurred vision, sweating, and even syncope



Gross

- **Any part** of the bladder; at **any level** of the bladder wall (37% in the dome, 20% in the trigone, 18% posterior wall, 13% anterior wall, the others in the bladder neck and lateral walls)
- **Muscularis propria** is the most common location
- Circumscribed or multinodular, usually < 4 cm
- Average diameter: 1.9 cm

Micro

- Nests of cells in **zellballen pattern** (diffuse patterns of large polygonal cells in clusters separated by a delicate fibrovascular stroma)
- Round cells with clear, eosinophilic, amphophilic granular cytoplasm, fairly uniform nuclei with smooth chromatin
- Frequently involves muscularis propria but without a desmoplastic reaction
- Mitoses are rare, and usually absent
- Necrosis, and vascular invasion are usually absent.



Immunohistochemistry

- **Positive stains:**
 - Neuroendocrine markers (Chromogranin, synaptophysin, neuron specific enolase (NSE))
 - Sustentacular cell: S100 (+)
- **Negative stains:**
 - Epithelial markers (CK7, CK20, or AE1/AE3)

Differential Diagnosis

- **Granular cell tumor:** S-100 (+), lack Zellballen pattern
- **Nested variant of Urothelial carcinoma:** lacks prominent vascular network of paraganglioma; keratin (+), neuroendocrine markers (-), usually associated with carcinoma in situ or noninvasive papillary carcinoma
- **Metastatic large cell neuroendocrine carcinoma:** necrosis, abundant mitotic activity, and cellular anaplasia. CK (+)
- **Malignant melanoma:** usually not a defined nesting pattern; is spindled or epithelioid, has prominent nucleoli; S100+, HMB45+, Melan A+



Criteria for Malignancy

- The criteria for diagnosing malignant paraganglioma are **metastasis** and /or “extensive local disease”
- **Long term F/U** is always indicated because metastases have been known to occur many years later
- In contrast to extraadrenal paragangliomas at other sites, of which approximately 10% exhibit malignant behavior, the frequency of malignancy in bladder paragangliomas is about **20%**



Predictive Factors

- NO reliable histologic criteria exist to distinguish malignant from benign neoplasms.
- The findings of nuclear pleomorphism, mitotic figures, and necrosis is not a reliable predictor of clinical outcome



Prognostic Factors

- A recent study: T1 or T2 did not show any recurrences or metastases
- High stage tumors (pT3) have poorer outcomes ([Cancer 2000;88:844](#))
- 4 features indicate increased potential for malignant behavior: younger age, hypertension, micturition attacks, invasive dispersion through the bladder wall



Treatment

- TUR (transurethral resection) /
wedge resection /
partial cystectomy
- Malignant paraganglioma: radical
cystectomy with removal of metastasis if
possible

