CASE OF THE MONTH OCTOBER

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History

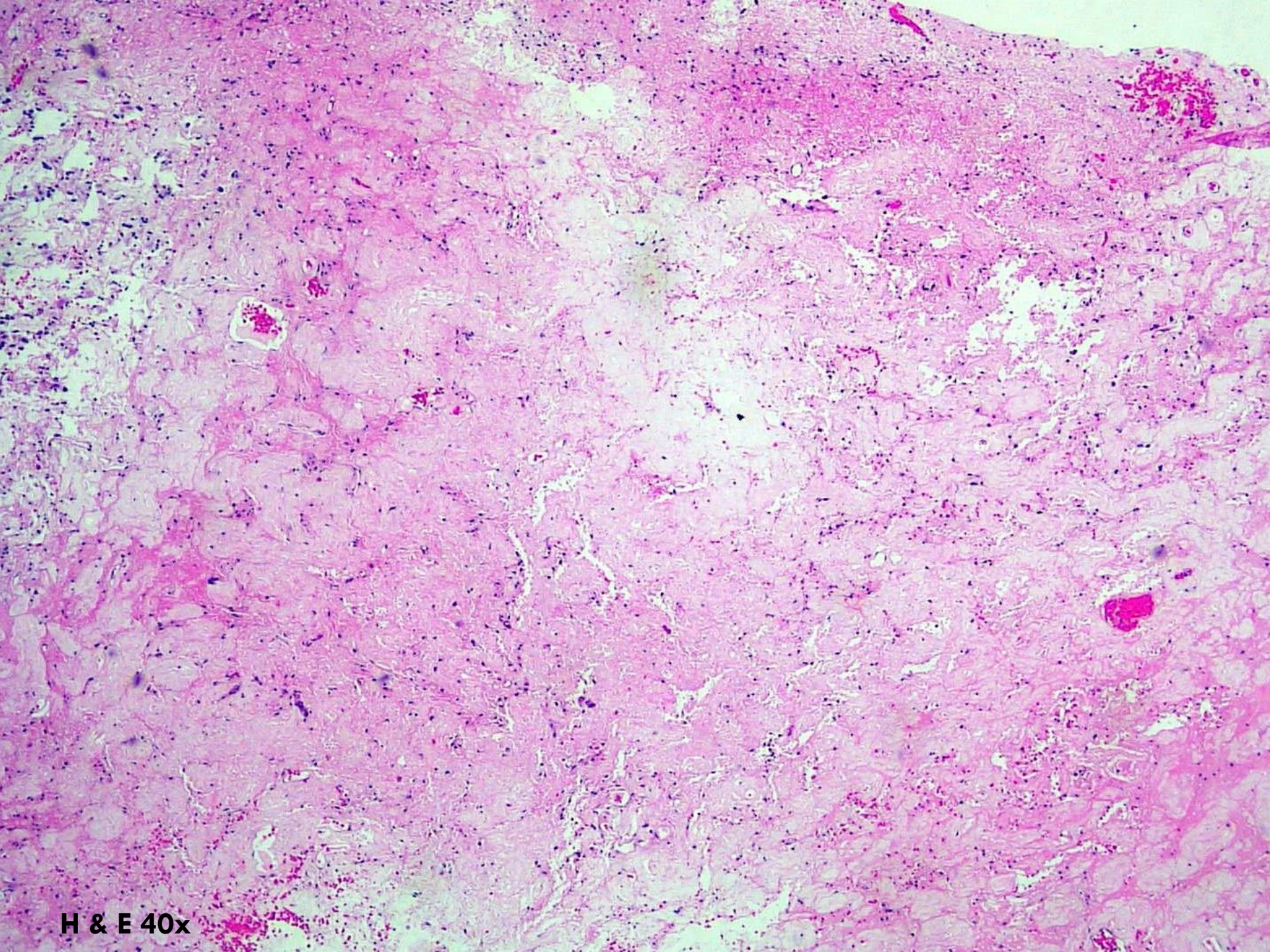
- 55 years male
- Known case of DM
- Headache x 6-7 months
- Giddiness
- Altered consciousness

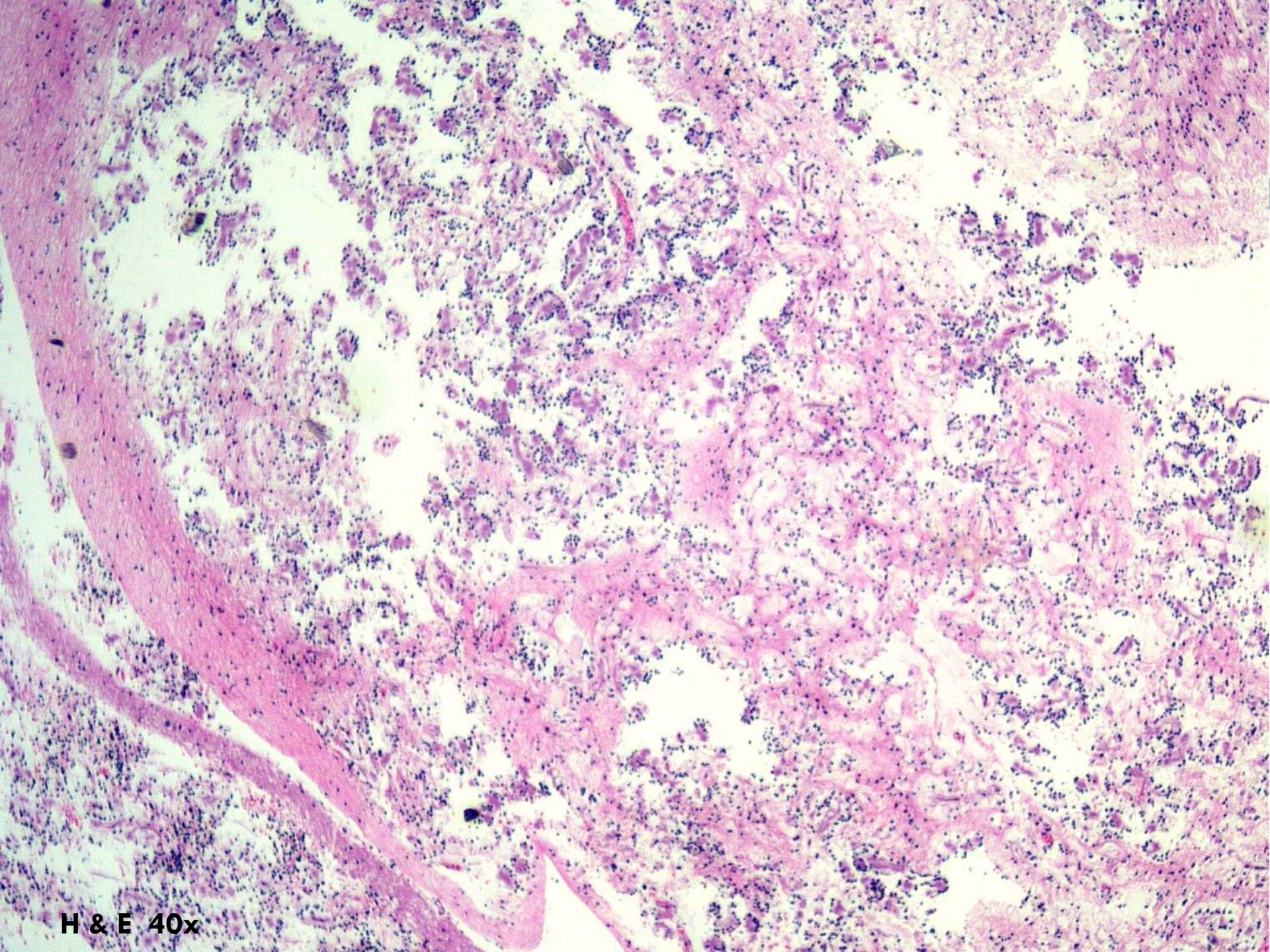
Investigations

- MRI
  - Posterior fossa lesion with hydrocephalous
  - Calcified non enhancing lesion, measuring 2.6 x 2.5 cm in midline cerebellum and fourth ventricle
- Other investigations were with in normal limits

Management

- Suboccipital craniotomy and decompression of tumour with external ventricular drain placement
- Intra-operative findings were suggestive of solid cystic tumor arising from fourth ventricle which was whitish in appearance
- Tumour was removed in piecemeal


H & E 40x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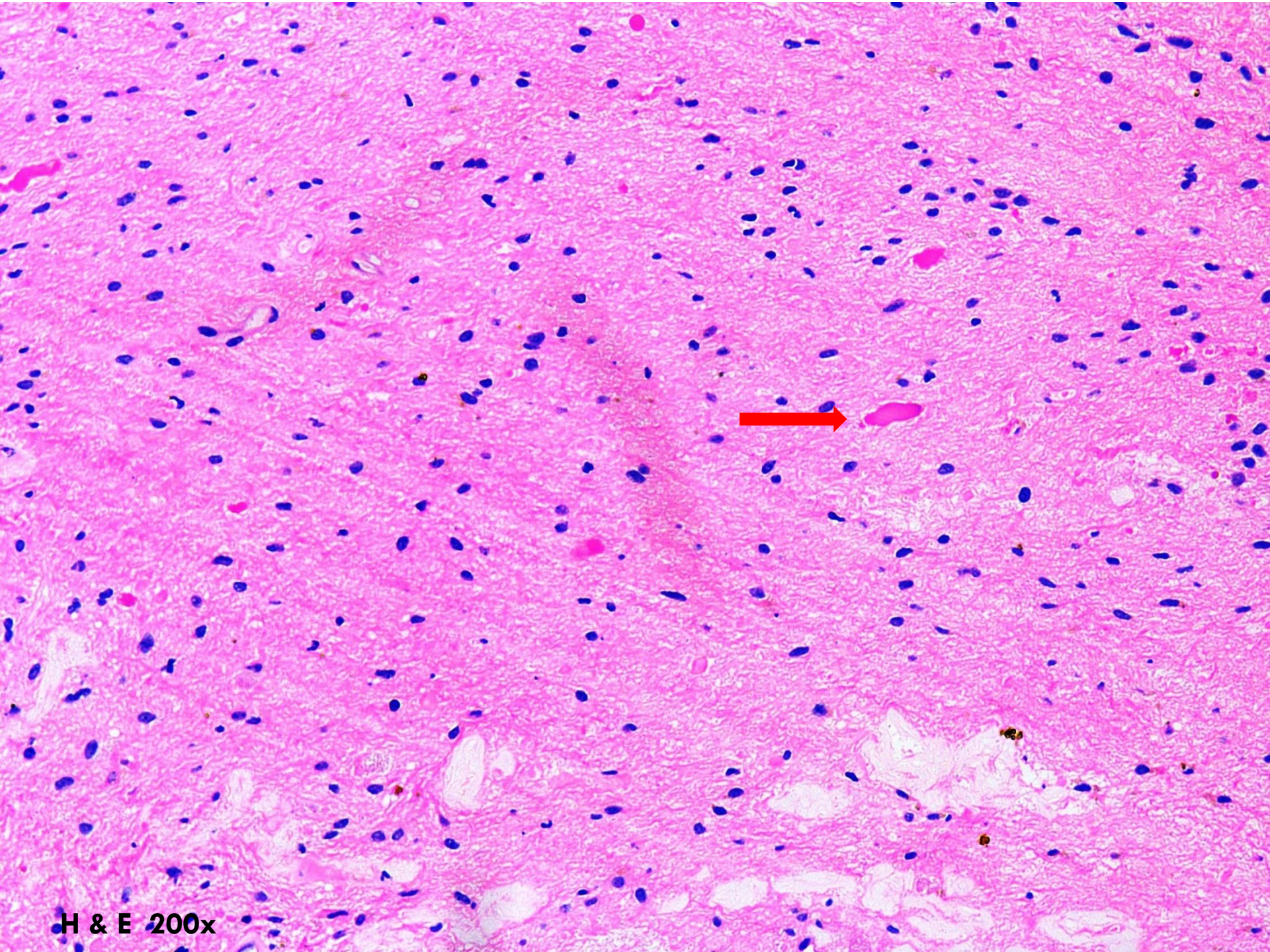
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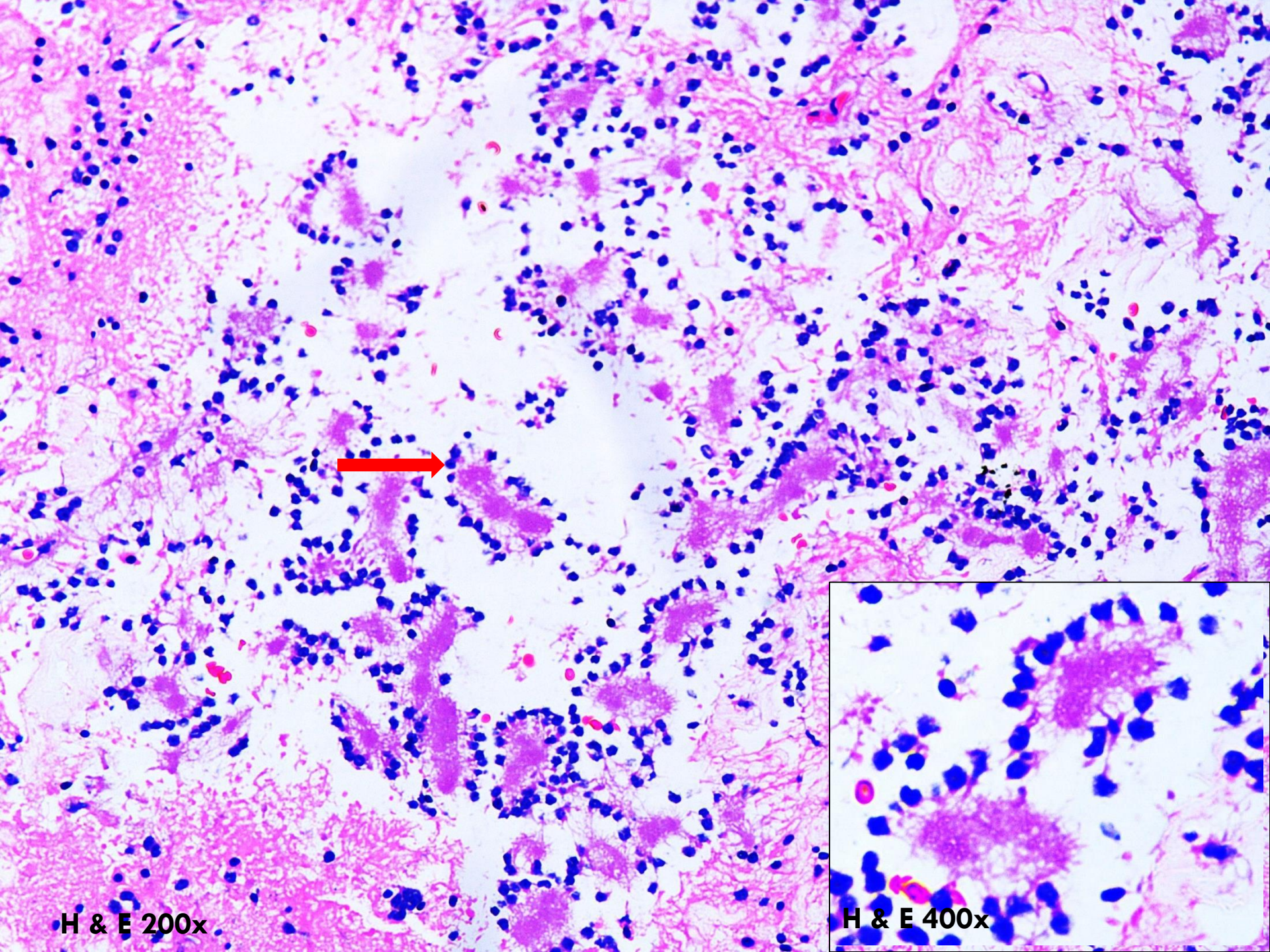
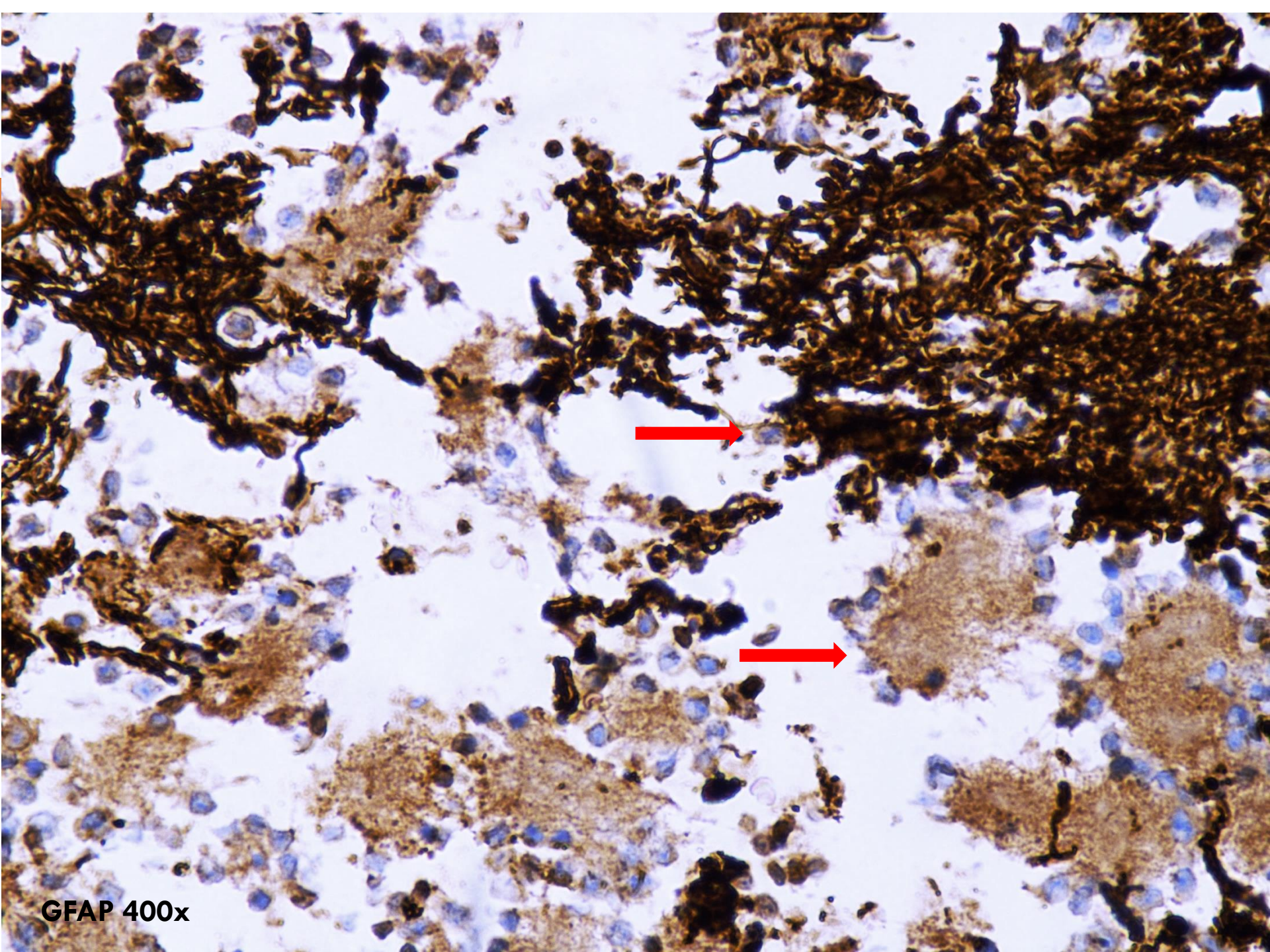

H & E 200x
H & E 400x

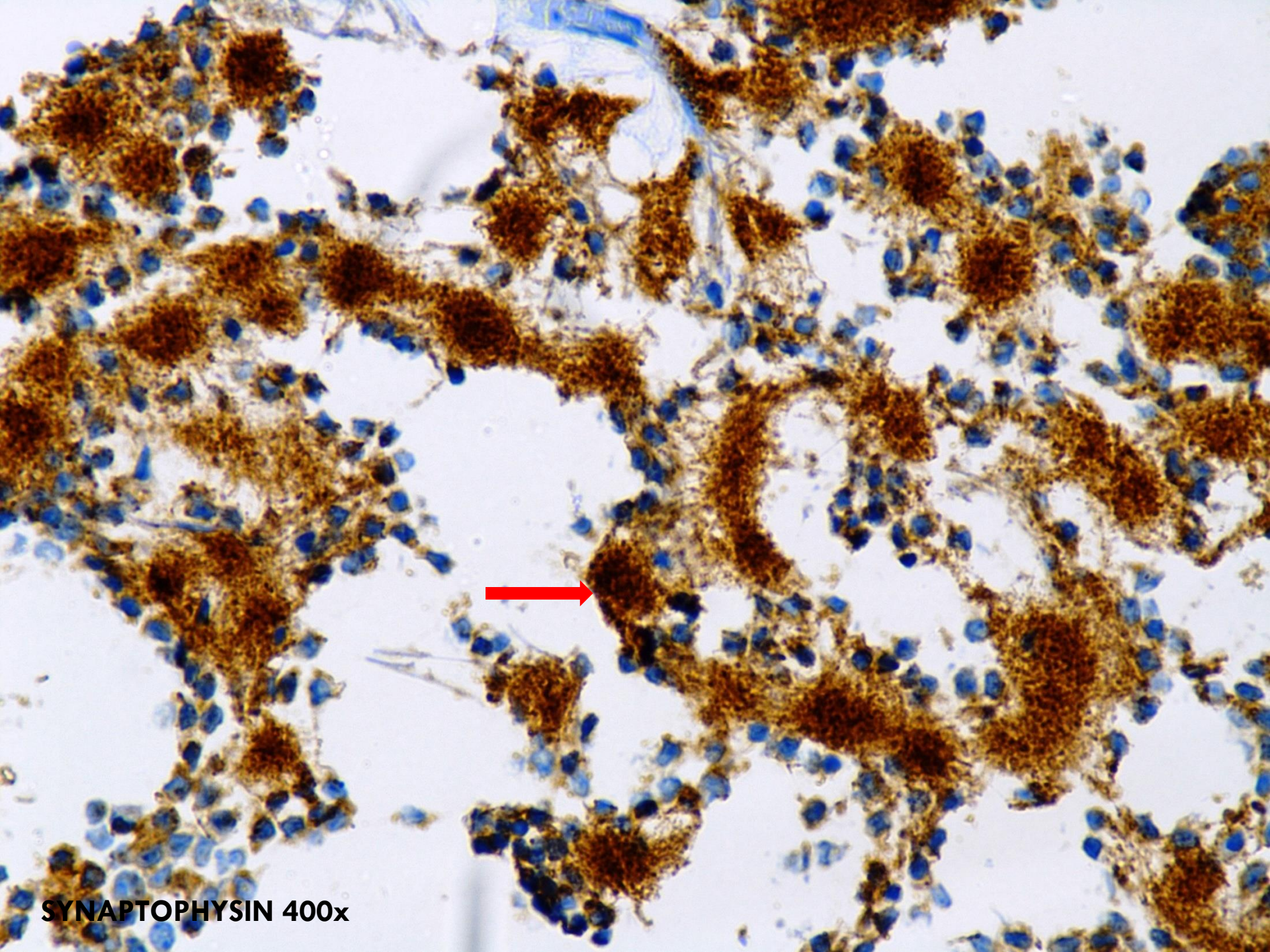
Morphological features

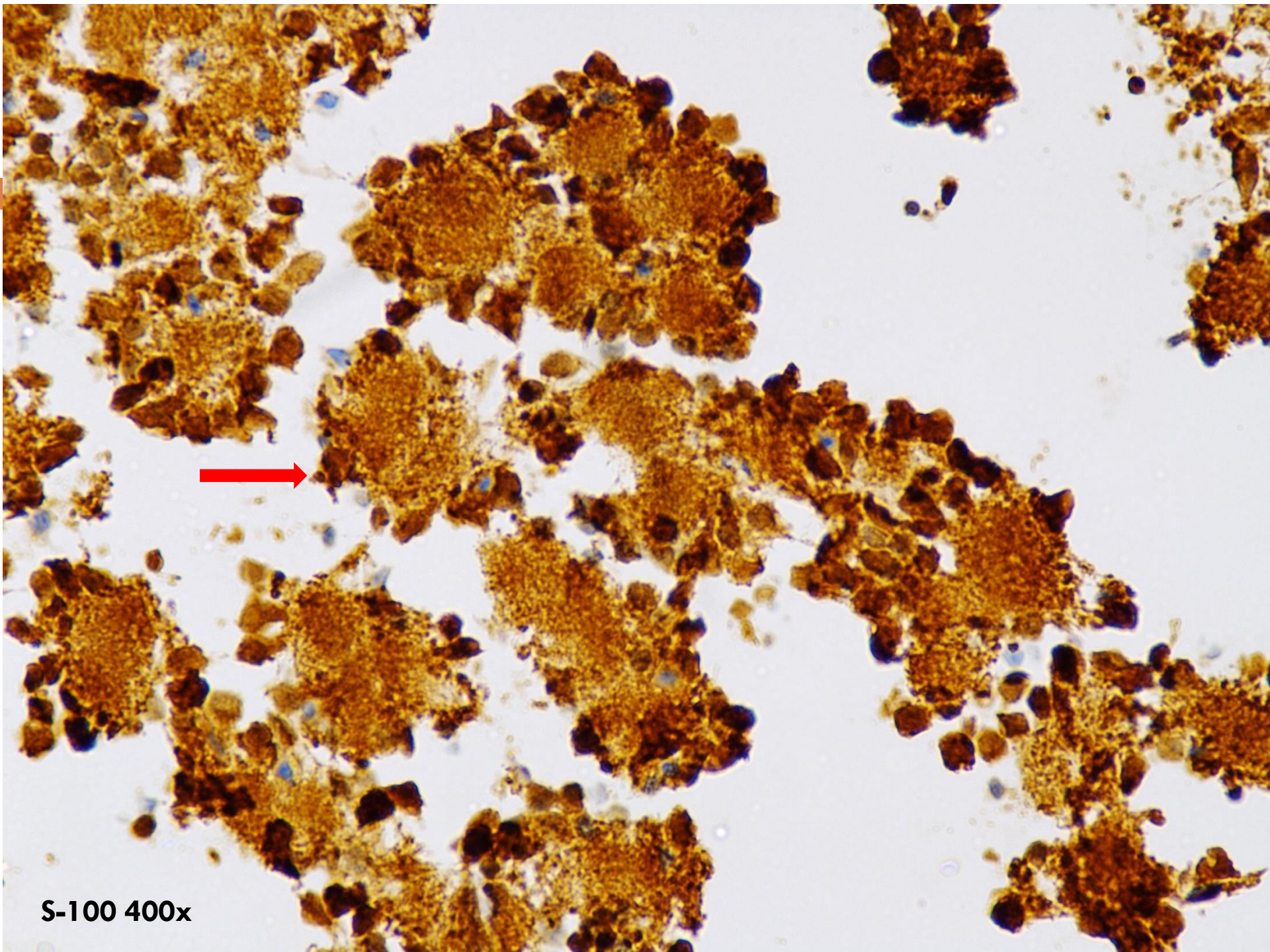
- Biphasic tumour
  - neurocytic
  - glial component
- The neurocytic component
  - consists of ring shaped neurocytic rosette around eosinophilic neuropil cores.
  - tumor cells have spherical nuclei scant cytoplasm
  - background is myxoid
- The glial component
  - shows spindle to stellate shaped nuclei with dense chromatin in a fibrillary background
  - rosenthal fibers, hemosiderin deposits are seen focally
- No necrosis or calcification seen.

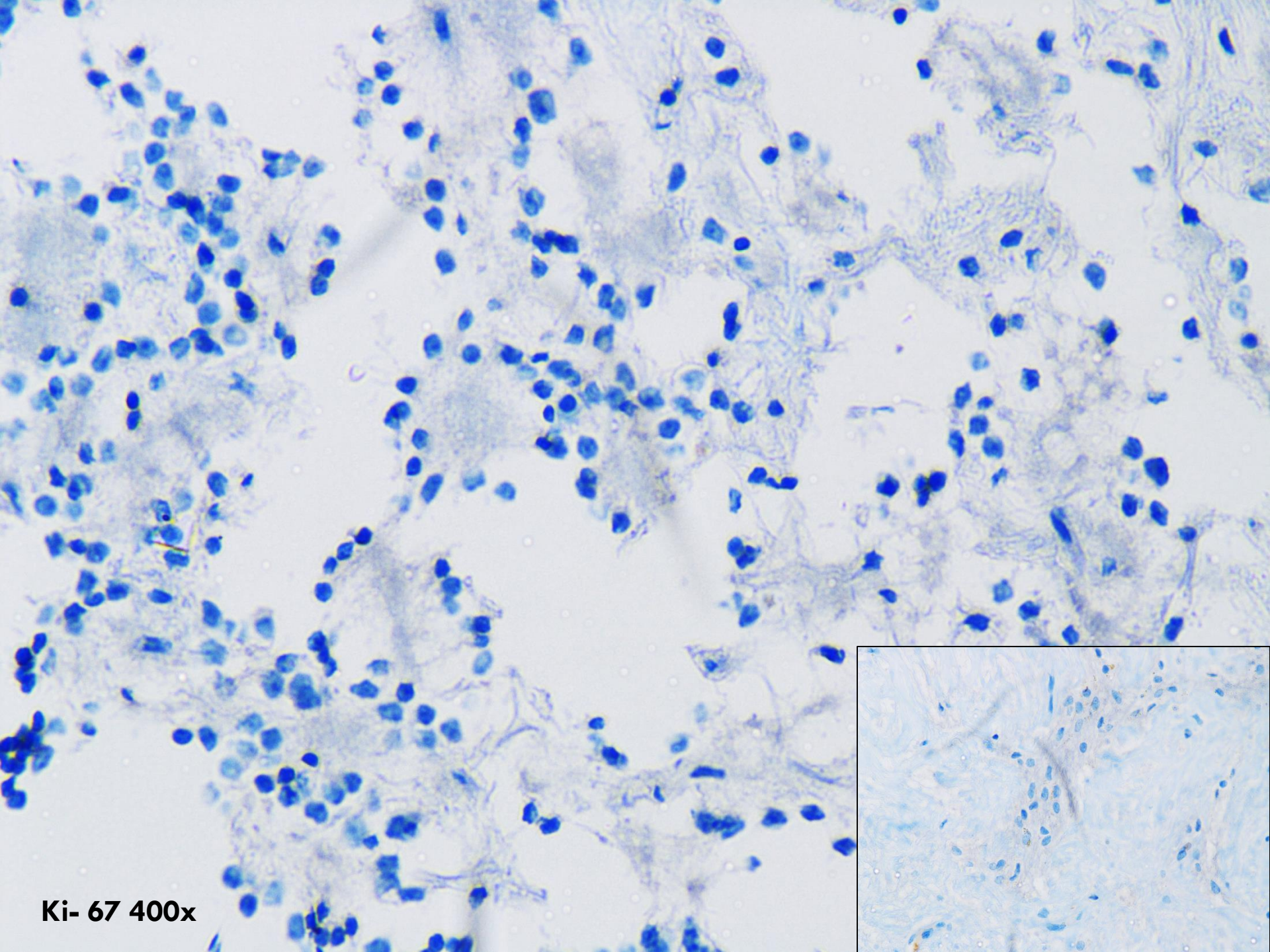
Differential diagnosis

- Based on glial component morphology
  - Pilocytic astrocytoma
  - Dysembryoplastic neuroepithelial tumour
- Based on neurocytic rosettes
  - Glioneuronal tumor with neuropil like islands
  - Rosette forming glioneuronal tumour (RGNT)
  - Ependymoma


GFAP 400x


SYNAPTOPHYSIN 400x


S-100 400x


Ki- 67 400x

Summary of case

- Tumour in cerebellum and fourth ventricle
- Histologically : biphasic tumour neurocytic and glial component
- On IHC synaptophysin positive in neuropil
- S 100 positive in neurocytic cells
- GFAP positive in glial component

Final diagnosis

Rosette Forming Glioneuronal Tumor

WHO grade 1

Discussion

- 2007 WHO classification of CNS tumors
  - "rosette-forming glioneuronal tumors of the fourth ventricle"
- Presence of this entity in various anatomical locations
  - cerebellar hemisphere and/or vermis , pineal region, chiasma, lateral and third ventricle, hypothalamus, and spinal cord
- 2016 WHO classification
  - renamed to "rosette-forming glioneuronal tumors" histologically classified as grade I

- Rare entity
  - only 150 cases of RGNTs have been described
- Young adults with female predominance are mostly affected
- Generally well demarcated
  - sometimes minor to moderate infiltration
- Histopathologic examination
  - Biphasic neurocytic and glial architectures
  - Oligodendroglial-like cells
  - Cellular atypia, mitotic figures, necrosis, and calcification are rarely visible

- Genetic testing of RGNTs may reveal mutations in PIK3CA and FGFR1 genes
- Although RGNTs are WHO grade I tumors and are considered benign
- Some reports have presented cases with intra-ventricular dissemination and rapid progression
- Management is usually through surgery with gross total resection (GTR) providing better prognosis

THANK YOU