Case #57

NAME Educational Activities Committee

Case provided by:

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Image provided by Dr. Melanie Bois (Mayo Clinic)

1. A 35-year-old man with recent balance issues who developed “food poisoning” and was found dead in bed. The brain finding is most consistent with:

- ◯ Demyelinating disease
- ◯ Congenital anomaly
- ◯ Remote infarct
- ◯ Substance use disorder
- ◯ Neurodegenerative disease

Answer...

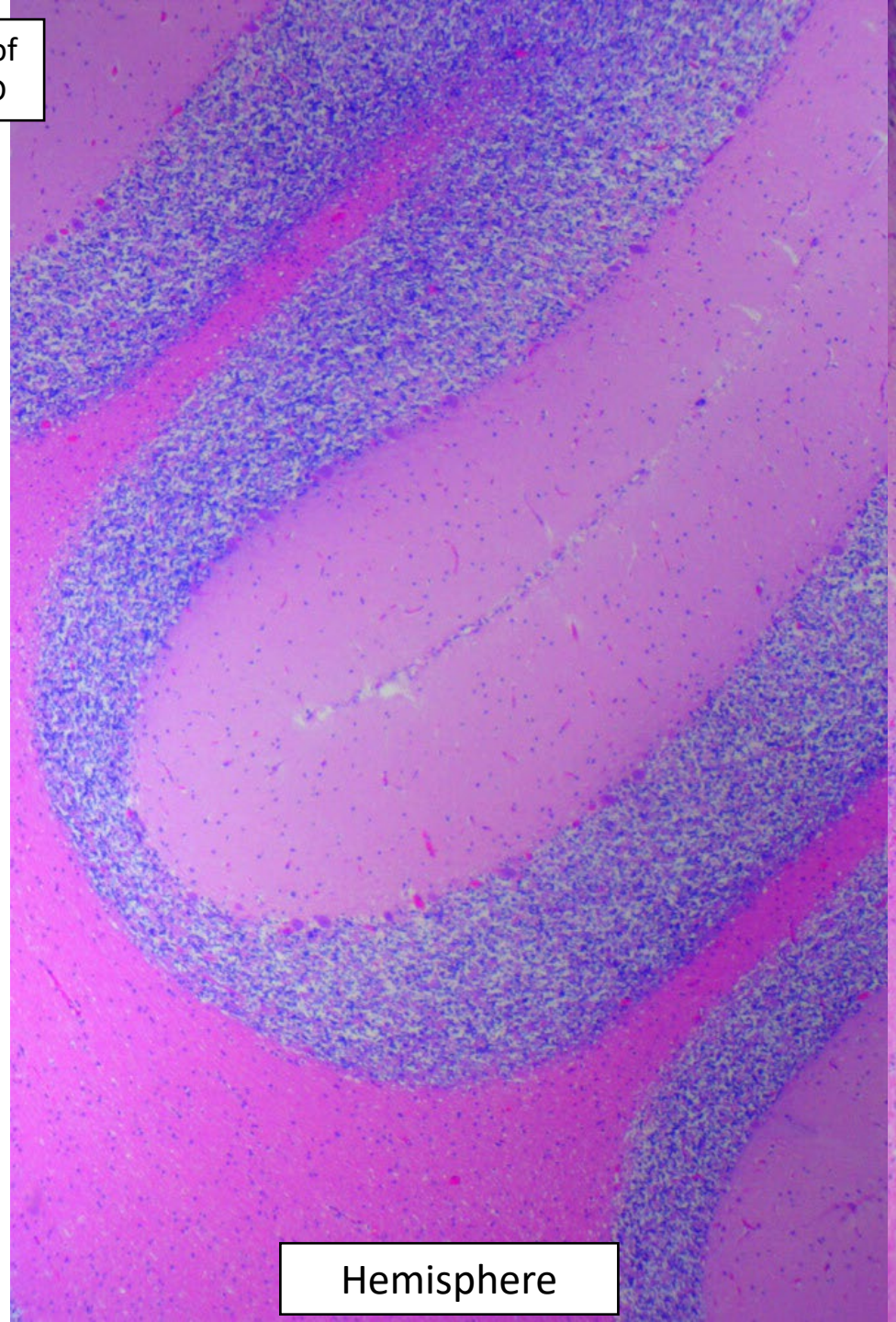
D. Substance use disorder – (CORRECT ANSWER, 55.17 % of responses)

The provided image shows atrophy of the superior cerebellar vermis. Loss of Purkinje cells were also identified on the histologic sections (see the following slides). This is a classic example of alcoholic cerebellar degeneration.

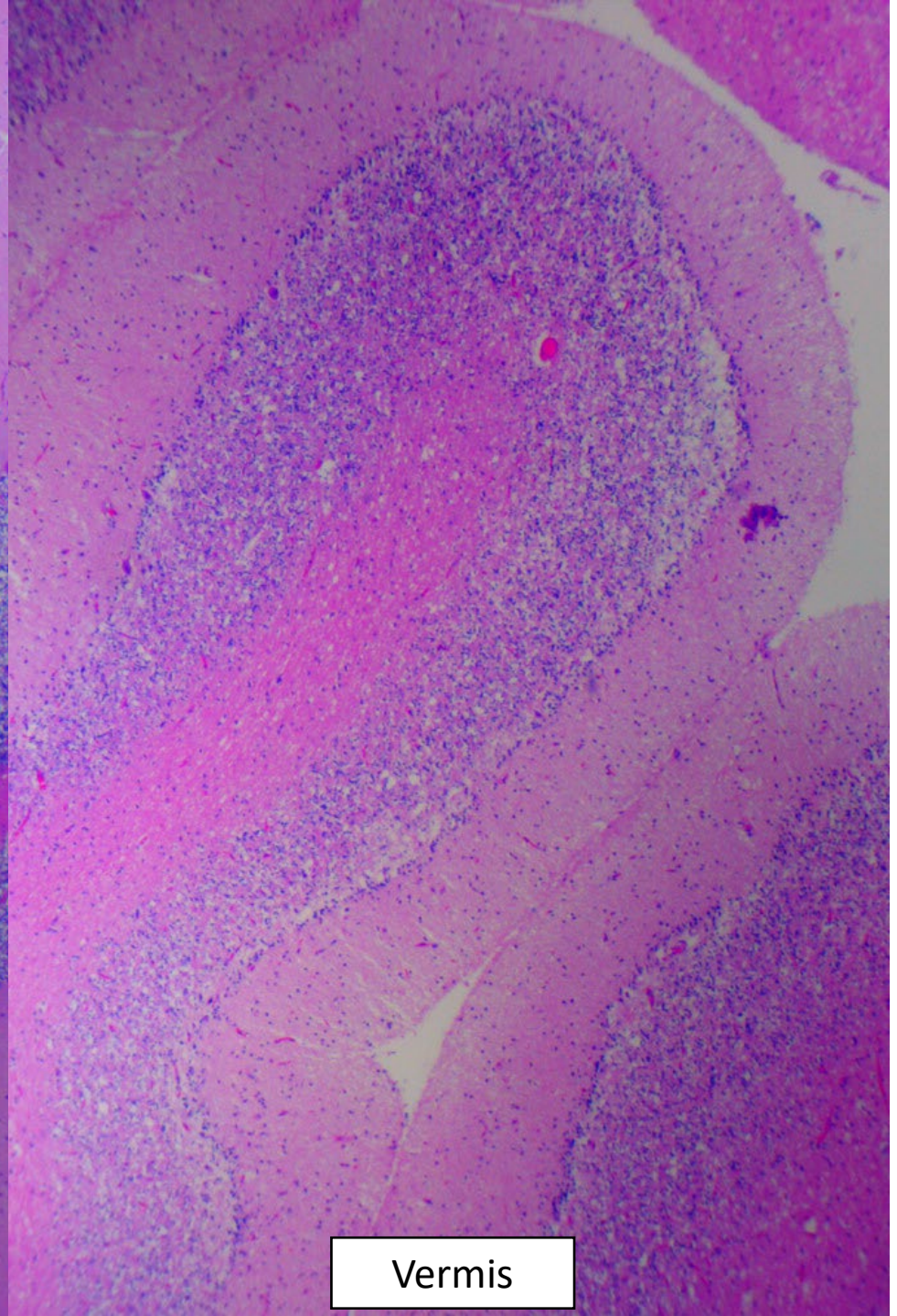
It is important to note that in these cases, it is the thiamine deficiency that alcoholics get from a lack of eating that results in Purkinje cell loss, as it is a co-factor for multiple enzymes, and not a direct effect of the alcohol. Therefore, vermal atrophy is a nutritional issue, not a direct toxic injury.

The selective involvement of superior vermis and hemispheres translates into disabling gait ataxia and leg dysmetria while coordinated movements of the arms and speech remain intact. The etiology behind the regional selectivity remains a mystery and the prevalence of alcoholic cerebellar degeneration varies greatly among the world's populations depending on local rates of chronic ethanolism.

Photos courtesy of Nicole Yarid, MD

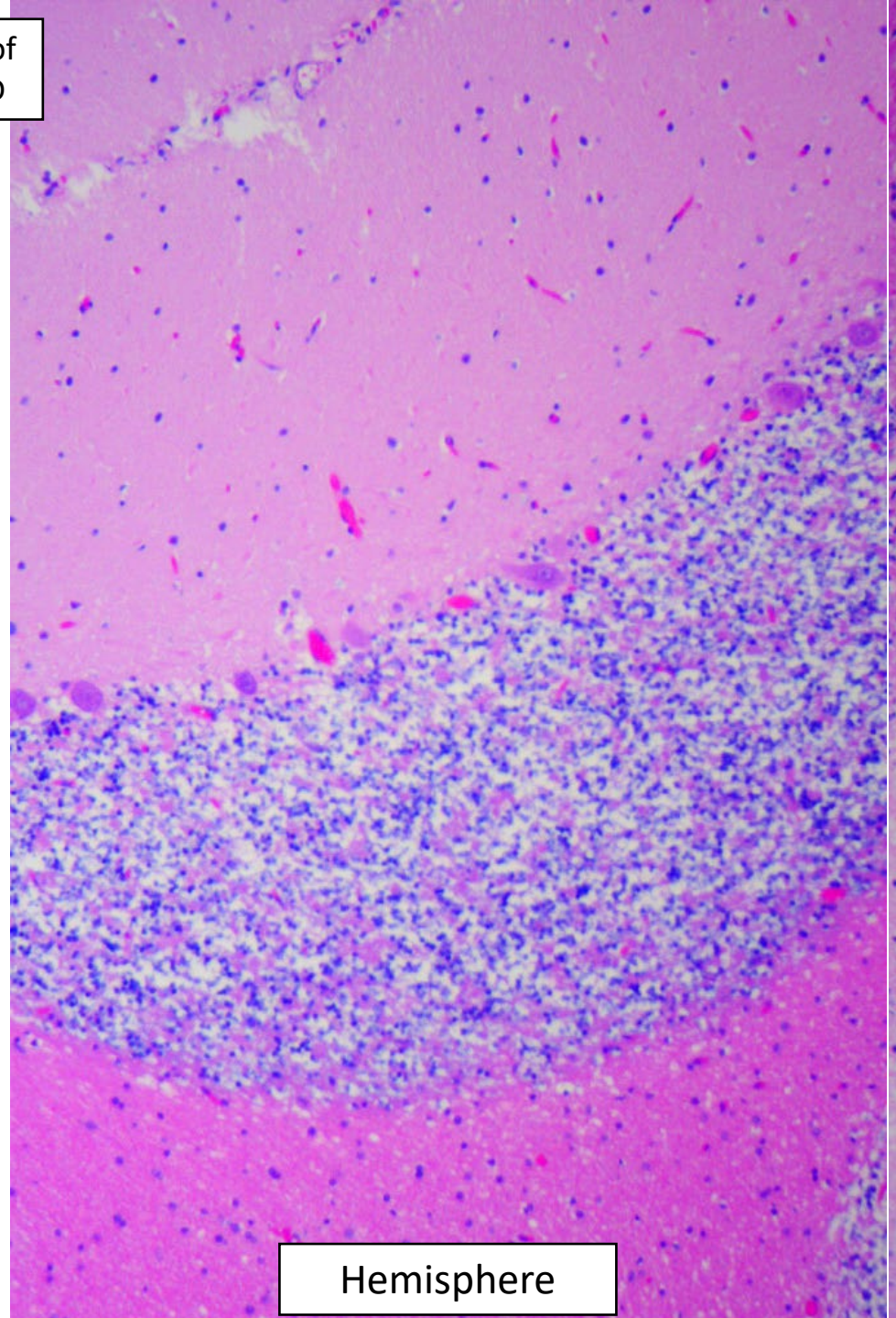


Hemisphere



Vermis

Photos courtesy of Nicole Yarid, MD



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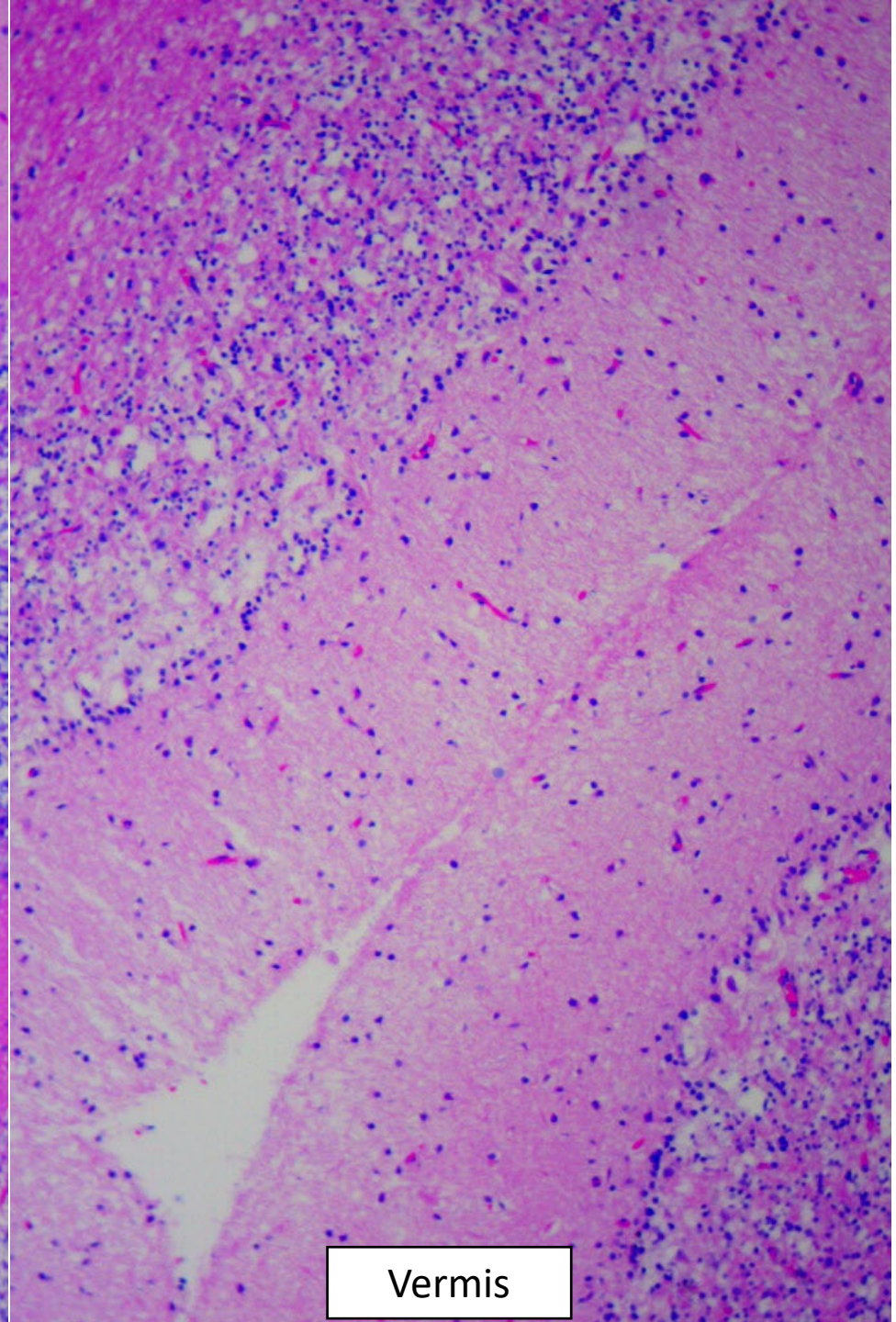
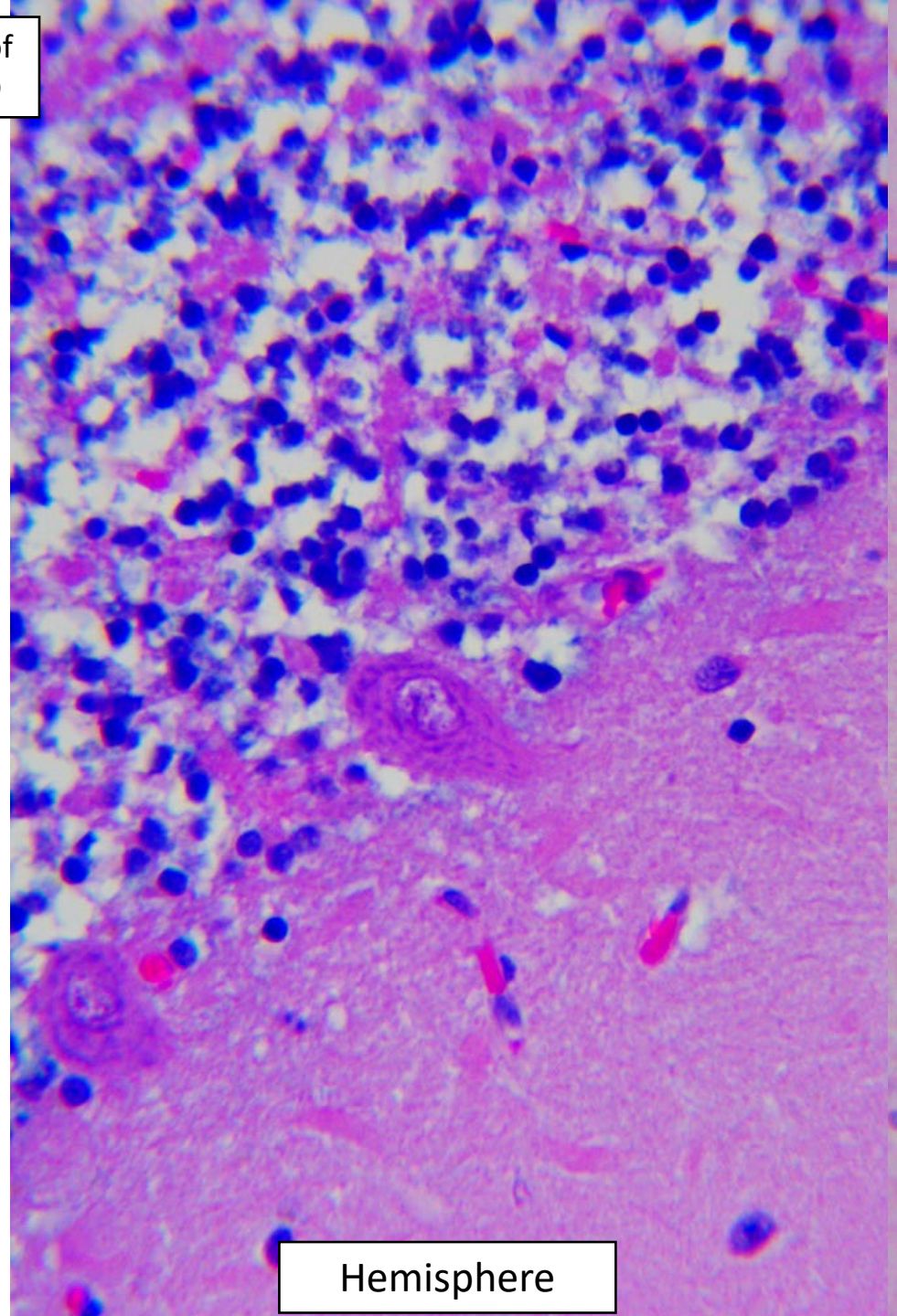


Vermis

Photos courtesy of Nicole Yarid, MD



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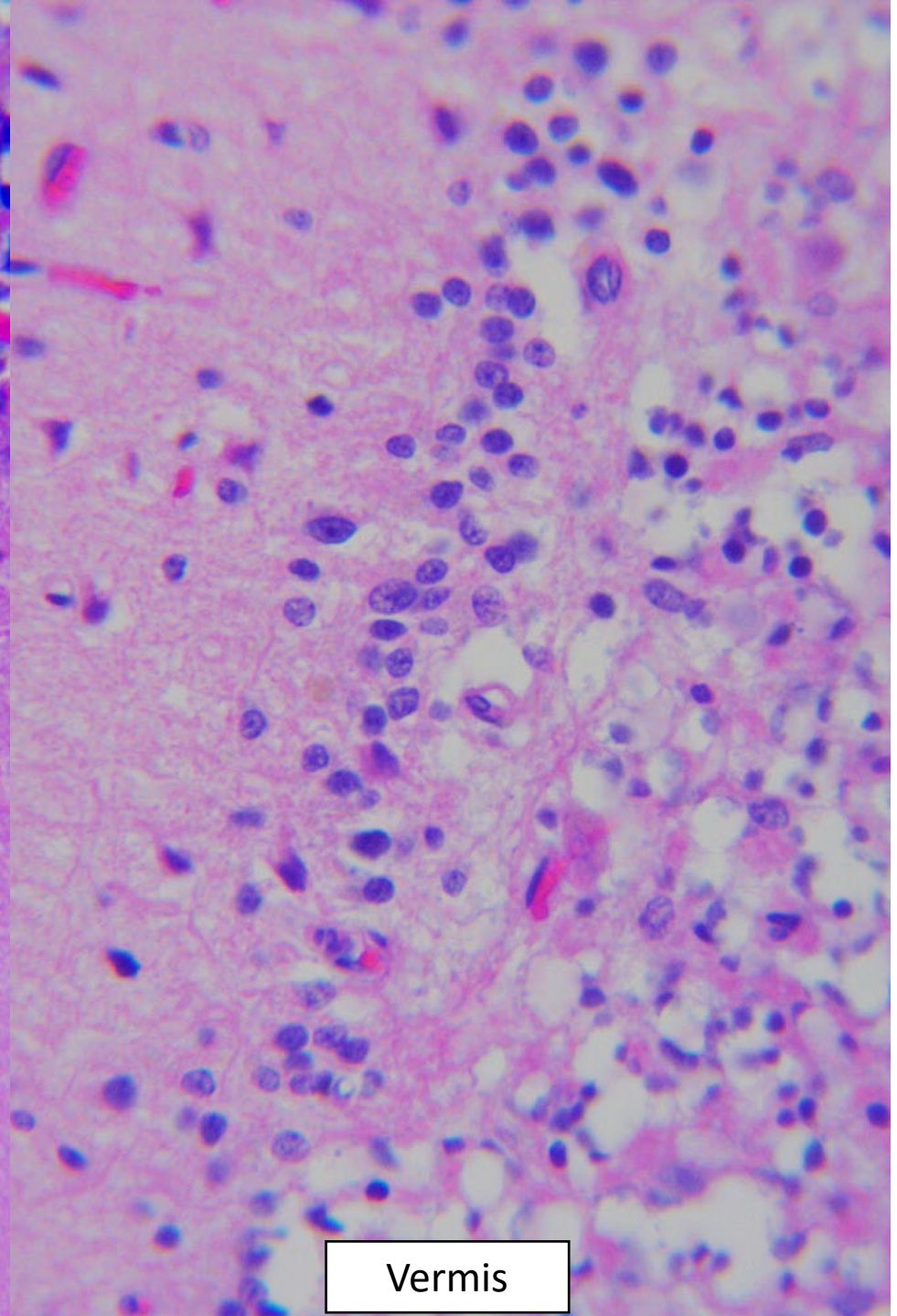

Vermis

A. Demyelinating disease (12.79 % of responses)

Plaques of multiple sclerosis (MS) are relatively uncommon in the cerebellum due to the paucity of white matter compared to the cerebrum. Posterior fossa lesions preferentially involve the floor of the fourth ventricle, the middle cerebellar peduncles, and the brainstem, a key feature that helps to identify MS plaques and differentiate them from focal areas of ischemic demyelination and infarction. Microscopy would also show a paucity of myelinated axons with microglial and astrocytes proliferation at the junction of the plaque and intact white matter.

B. Congenital anomaly (7.33 % of responses)

Congenital anomalies, such as hereditary ataxias, associated with cerebellar atrophy are a heterogeneous group of disorders. Even within the narrower category of spinocerebellar ataxias (SCAs) neuropathology is diverse. While the adult-onset balance issues and cerebellar vermis atrophy are compatible with subtypes of SCA, the lack of cytoplasmic neuronal inclusions suggests an alternate etiology. In contrast, the cerebellar lesion in Friedreich ataxia is largely restricted to the dentate nucleus.

C. Remote infarct (9.20 % of responses)

Due to the smaller caliber of arteries supplying the cerebellum, embolism in this location is less common than in the cerebral hemisphere. Remote infarctions grossly appear as cystic necrosis of the tissue in the territory of the involved artery. While strokes in this age group do occur, they are uncommon, and the lack of cystic change macroscopically and macrophages microscopically makes a remote infarct unlikely.

E. Neurodegenerative disease (15.52 % of responses)

Consistent patterns of cerebellar atrophy can be found in patients with Alzheimer's disease, amyotrophic lateral sclerosis (AML), frontotemporal dementia (FTD), multisystem atrophy (MSA), and progressive supranuclear palsy (PSP). Evidence suggests that these cerebellar changes are highly disease-specific and correspond to the cortical or subcortical changes characteristically reported in each disease. Vermis atrophy alone is not a reported pattern of cerebellar degeneration in these diseases and the patient is not within the usual age-of-onset range for most of these entities.

REFERENCES

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