Case #69

NAME Educational Activities Committee

Case provided by:

Dr. Jusmita Saifullan (PGY2, Duke University Hospital),
Dr. Paul Yell and Dr. Craig Nelson (North Carolina Office of the Chief Medical Examiner in Raleigh)


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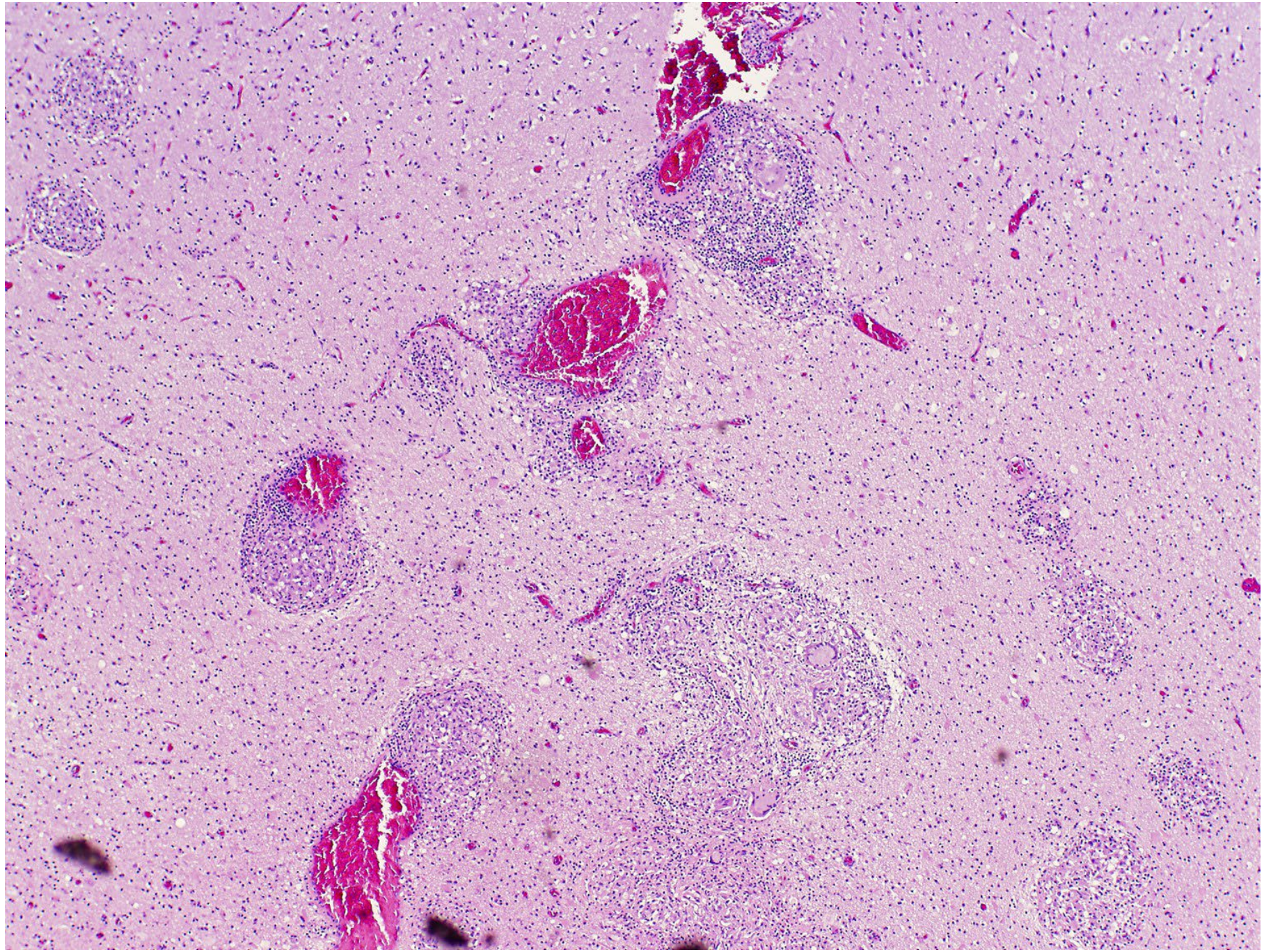


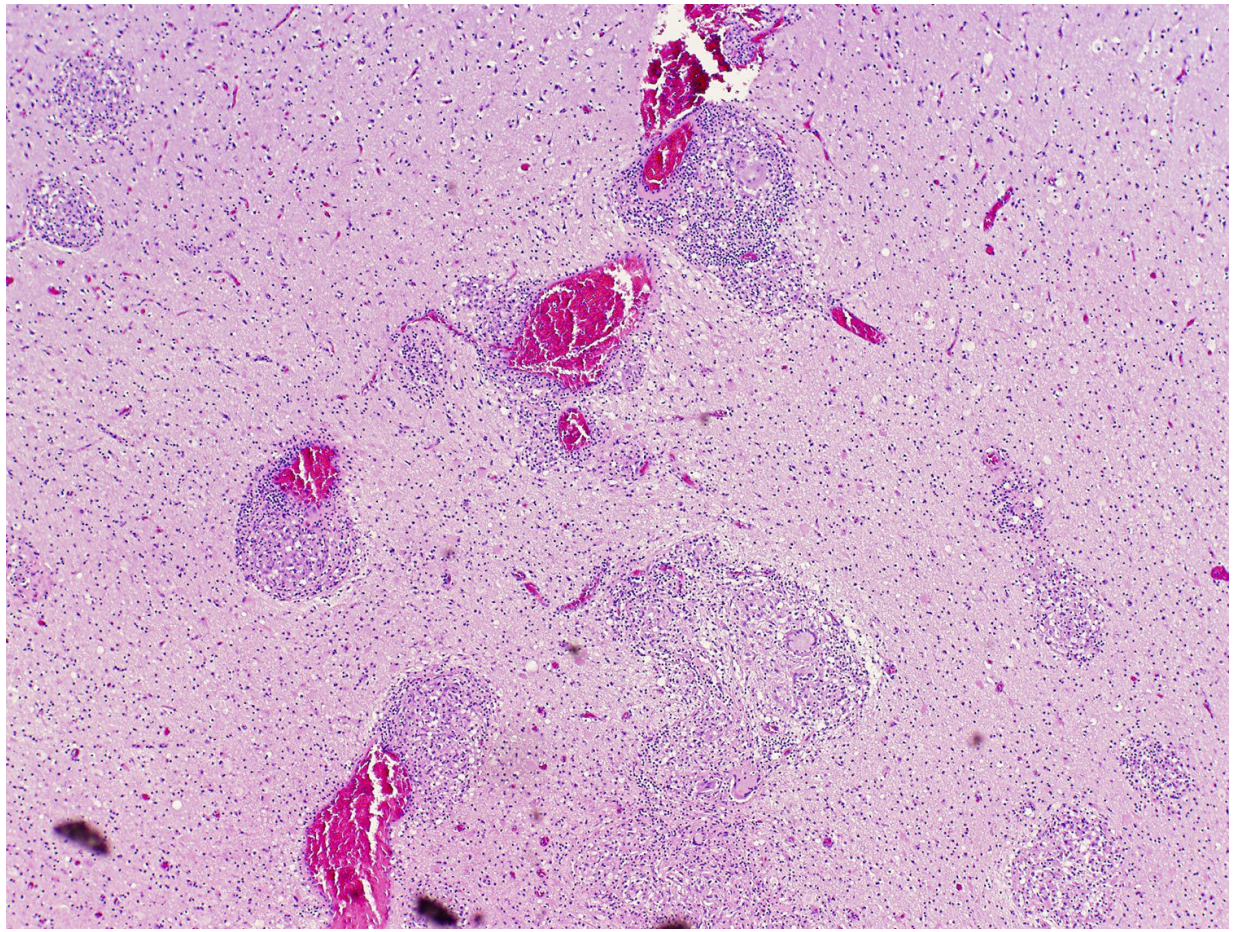
1. This is a case of a 40-year-old male that was found dead approximately an hour after he was last seen alive. Gross examination of the brain revealed fine granularity of the ventral frontal lobes. Sections of fixed brain showed friable, dusky parenchyma in this region. Representative gross and microscopic images of the lesions are shown. Histochemical stains including GMS and AFB were performed and were negative. Which of the following statements is true?

- ◯ The decedent’s medical record would reveal a history of non-CNS cancer.
- ◯ The cells of this lesion would stain positive for GFAP and Olig2.
- ◯ The decedent had likely been eating undercooked pork.
- ◯ Flow cytometry is necessary to make the final diagnosis.
- ◯ Histologic examination of the decedent's lungs may show non-caseating granulomas.

Answer...

E. Histologic examination of the decedent's lungs may show noncaseating granuloma – (CORRECT ANSWER, 71.98% of responses)

The histological sections of the brain lesions show many discrete, noncaseating granulomata composed of aggregates of epithelioid histiocytes and multinucleated giant cells, with surrounding lymphocytic infiltrates, consistent with neurosarcoidosis.



Neurosarcoidosis is a diagnosis of exclusion, with common mimics including tuberculosis and fungal infections. Therefore, when abundant granulomata are observed, infectious etiologies should always be considered. Special stains for fungal organisms (GMS) and acid-fast bacteria (AFB) can be performed to help rule out an infectious process, as was done for this case.

Sarcoidosis is a systemic inflammatory granulomatous condition that can affect multiple organ systems, especially the lungs, eyes and skin. Therefore, noncaseating granulomatous lesions in the decedent’s other organs, including the lungs, would be likely.

A. The decedent's medical record would reveal a history of non-CNS cancer

(3.38 % of responses)

This answer choice is alluding to a diagnosis of metastatic disease. Multiple brain lesions observed grossly should always raise the possibility of metastatic disease from a non-CNS primary. However, on histology the lesions are composed of giant cells and histiocytes (which can be highlighted by CD68 and CD163) and makes an inflammatory process more likely.

B. The cells of this lesion would stain positive for GFAP and Olig2

(5.48 % of responses)

This answer choice is suggesting a diagnosis of pilocytic astrocytoma, in which neoplastic cells generally stain positive for GFAP and Olig2. This entity is a solid-cystic glial neoplasm with a characteristic biphasic compact and loose pattern on histology. Rosenthal fibers, characteristic of this tumor, can also be found in neurosarcoidosis cases (while not present in our microscopic image) and could lead to the consideration of pilocytic astrocytoma in the differential. Nevertheless, well-formed granulomata or multiple brain lesions would not be characteristic for this entity.

C. The decedent had likely been eating undercooked pork (11.27 % of responses)

Various parasitic infections can typically present as multiple grossly identified lesions in the brain. The focus on undercooked pork here is hinting at a diagnosis of neurocysticercosis, where the brain is infected by the larval form of Taenia solium (which infects humans through consumption of contaminated pork). Unlike neurosarcoidosis, these lesions tend to be cystic with a characteristic single invaginated white scolex. Histologically, the scolex as well as hooklets can be seen (which can be highlighted by acid-fast staining).

D. Flow cytometry is necessary to make the final diagnosis (7.89 % of responses)

The presence of frequent lymphocytic infiltrates can raise the possibility of leukemia/lymphoma, which could be detected using flow cytometry. However, the many discrete granulomata seen here makes a lymphoma less likely. Additionally, the lymphocytes appear small and mature with no aggregate formation, further lowering a hematolymphoid process in the differential.

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