

Inflammatory disease of the Meninges in Surgical Neuropathology

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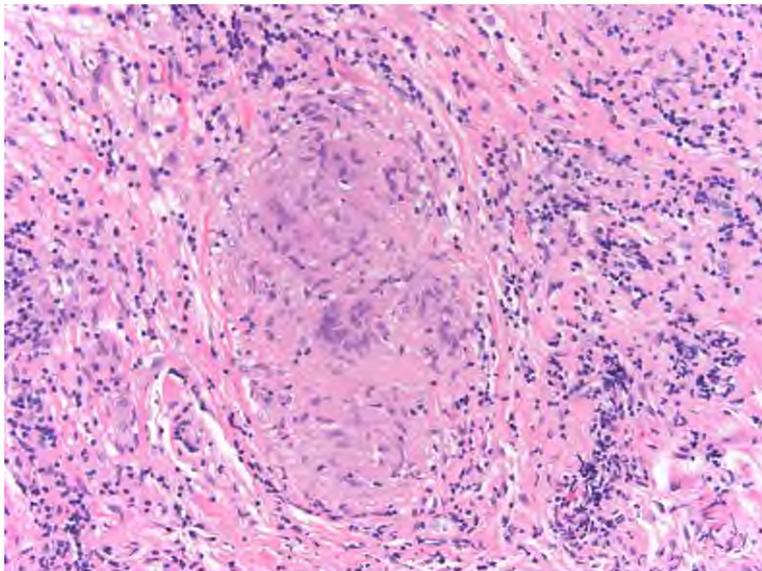
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Case-Based Questions (please see page 4 for answers)

1.	Findings diagnostic of IgG4 related disease include (circle all that apply):
a.	A dense lymphoplasmacytic infiltrate, which can be associated with increased numbers of eosinophils
b.	Well formed granulomas and a prominent neutrophilic infiltrate
c.	Fibrosis arranged at least focally in a storiform pattern
d.	Obliterative phlebitis

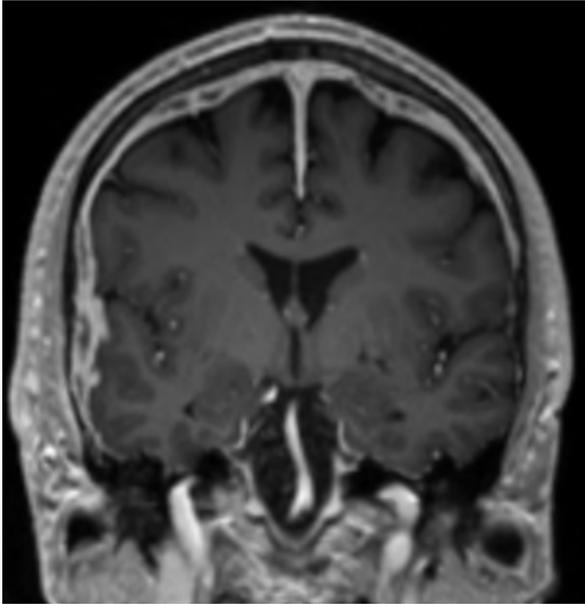
2. A 54-year-old woman with no prior medical history presents with right face pain. On imaging, she is found to have dural and leptomeningeal enhancement involving the tentorium, cerebellum and brain stem predominantly on the right. A microscopic photo of the biopsy is shown below:



The most likely diagnosis in this patient is:

a.	IgG4-related disease
b.	Granulomatosis with polyangiitis
c.	Sarcoidosis
d.	Rheumatoid arthritis

3. A 66-year-old man presents with confusion. The MRI from the patient is shown below:



A biopsy is obtained which shows areas of geographic necrosis, with vague granulomatous inflammation with giant cells and an inflammatory background rich in neutrophils.

Which of the following statements is true regarding this patient?

- | | |
|----|--|
| a. | This patient has a leptomeningitis and the most likely diagnosis is a fungal infection |
| b. | This patient has a pachimeningitis and the most likely diagnosis is granulomatosis with polyangiitis |
| c. | This patient has a leptomeningitis and the most likely diagnosis is IgG4-related disease |
| d. | This patient has a pachimeningitis and the most likely diagnosis is rheumatoid arthritis |

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Question 1: Correct answer and rationale: Answer: a, c, d

While a dense lymphoplasmacytic infiltrate, which can be associated with increased numbers of eosinophils, Fibrosis arranged at least focally in a storiform pattern and obliterative phlebitis are the three major histologic features associated with IgG4 related disease, well formed granulomas and a prominent neutrophilic infiltrate are features **inconsistent** with IgG4 related disease (Mod Pathol. 2012 Sep; 25(9):1181-92)

Question 2: Correct answer and rationale: **Answer: c**

The photo shows a well formed granuloma, without necrosis and with presence of giant cells typical of sarcoidosis and not seen in the other listed conditions.

Question 3: Correct answer and rationale: **Answer: b**

The MRI shows a pattern of enhancement involving the dura mater over the convexities and along the falx, consistent with a pachimeningitis rather than leptomeningitis. The histological findings with areas of geographic necrosis, with vague granulomatous inflammation with giant cells and an inflammatory background rich in neutrophils are most consistent with the diagnosis of granulomatosis with polyangiitis. Histopathological features typical of rheumatoid arthritis are instead presence of meningeal inflammation with a predominance of plasma cells, rheumatoid nodules and vasculitis.