7 yo female presents with slowly progressive dysphagia, headache, ataxia, and dysarthria. MRI revealed a 2.6 cm enhancing lesion within the left posterior fossa within the parenchyma of the left medulla, possibly originating from the dural surface.

Contributed by Dr. Nishant Tiwari
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Factor XIIIa

Contributed by Dr. Nishant Tiwari
Special stains (GMS, AFB, Gram) and blood cultures were negative.

What are your top differential diagnoses?
Final diagnosis: **ALK-positive histiocytosis**

- More common histiocytic disorders in children include:
  - Erdheim-Chester disease
  - Juvenile xanthogranuloma
  - Rosai-Dorfman disease
  - Langerhans cell histiocytosis

- This lesion consisted of sheets of bland histiocytoid cells with irregularly folded nuclei & fine chromatin

- ALK IHC was diffusely positive in the lesional cells
ALK-positive histiocytosis

• One of the newest subtypes of histiocytic disorders (described ~10 yr ago)
• Predilection for neonates/infants
• Often with multi-organ involvement, but also may present exclusively as intracranial/CNS disease
• Morphology may overlap with Rosai-Dorfman disease, Langerhans cell histiocytosis, Erdheim-Chester disease, or juvenile xanthogranuloma
  • Touton-like giant cells reported in several cases
• KIF5B-ALK is the most commonly-reported fusion

References: PMID: 30573850, PMID: 31119374