What is the Best Way to Approach a Zebra?

Cautiously, They May Buck
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Introduction
Erdheim-Chester Disease (ECD), a rare Non-Langerhans Cell Histiocytosis, is a benign proliferative disorder characterized by excessive production of histiocytes within multiple organ systems, often with life-threatening complications. The rarity of the condition and the paucity of research pose many challenges, some of which were best recognized following the death of this patient.

Background of Histiocytosis

Dendritic Cell-Related Histiocytic Disorders:
- Langerhans Cell Histiocytosis
- Non-Langerhans Cell Histiocytosis
  - Clinically and histogenetically defined
  - Caffey disease
  - Langerhans sarcomatoid histiocytosis
  - Cutaneous with a major systemic component
    - JXG family: Sinus histiocytosis with massive lymphadenopathy
    - Non-JXG family: Multicentric reticulohistiocytosis
  - Systemic:
    - JXG family: Erdheim-Chester disease
    - Non-JXG family: Blush histiocytosis with massive lymphadenopathy (R-D2)