

18 Langerhans cell histiocytosis**Micro**

- These are lesional tissue fragments.
- The lesion is cellular and composed of histiocytes, Langerhans cells and eosinophils.
- There is no atypia or mitotic activity.
- There is no evidence of malignancy.
- No native or background tissue is seen.

Diagnosis

- Langerhans cell histiocytosis

Further work/comment

- Confirmatory immune markers are CD1a and S100. And Langerin
- EM shows Birbeck granules.
- The lesion could be solitary, one of multiple lesions or multiple organ involvement.
- The disease, formally called histiocytosis X, has a broad spectrum of clinical behaviors; some cases are self-limited, whereas others involving multiple organs cause significant mortality.
- Investigations to look for more widespread disease are advised.
- The disease is associated with BRAF mutation and responds to RAF pathway inhibitors.