Histiocytosis, a rare cause of hypopituitarism. Langerhans cell histiocytosis and Erdheim-Chester disease, two case reports of pituitary deficiency.

Hána V. jr., Kosák M., Hána V.
3rd Department of Internal Medicine, 1st Faculty of Medicine, Charles University and General Teaching Hospital in Prague

**Case 1**

36-year-old man diagnosed with diabetes insipidus, central hypogonadism, severe GH deficiency in 2006 on MRI in 2006, 2007, 2009 only absent neurohypophysis signal on T1, without other pathology

**Recurrent Bilateral external otitis** since 2011

**MRI** in 2014: hypothalamic lesion with a pituitary stalk enlargement, multiple white matter lesions in T2W sequences in mesencephalon, pons and cerebellum surrounding the IV. ventricle

2014 dg. **Folliculitis capitis** by local dermatologist

Histiocyte like cells infiltration of dermis, cells are CD 1a+, S100+, BRAF - in the scalp biopsy

**PET/CT** showed infiltration of hypothalamus, maxilla, a mandible with perifocal soft tissue involvement and regional lymphadenopathy

**Diagnosis:** Langerhans cell histiocytosis

**Treatment** initiated with cladribine (Litak)

**Conclusion:** In patients with central diabetes insipidus of unclear etiology precise physical examination and PET/CT scan can reveal a focus appropriate for a biopsy leading to the diagnosis.

**Case 2**

42-year-old man with isolated diabetes insipidus, diagnosed in 2014

**MRI** showed an infundibular infiltration of unknown origin

Several **skin papules** last 5 years, biopsy showed CD 68+, S100-, CD 1a - histiocyte infiltrate, not favoring a Langerhans cell histiocytosis, could be a multicentric histiocytosis

**PET/CT** showed osteolytic lesions in both scapulas, proximal humeri, clavicles, transversal procesus of Th7, left ilium and proximal parts of femur bones

Clavicle **biopsy** revealed foamy histiocytes, CD68+, CD 1a and S100-, BRAF +

**Diagnosis:** Erdheim-Chester disease (non-Langerhans cell histiocytosis)

**Treatment** iniciated with IFN α (Roferon)