

Introduction

- Langerhans cell histiocytosis(LCH) is a rare histiocytic disorder most commonly characterized by osteolytic bone lesions.
- Central nervous system(CNS) involvement observed at six percent and Diabetes insipidus (DI) one of the important signs of CNS involvement.

Case

- A forty-years-old woman presented with polyuria, polydipsia and headache. Water deprivation test was compatible with DI.
- Magnetic resonance imaging (MRI) shows 4x4 mm focal thickening at the distal infundibular stalk (Figure 1a-b). She was followed-up under desmopressin.
- At the sixth months of therapy, clinical situation negatively progressed with complaints of lethargy, diplopia, menstrual irregularity, headache and forgetfulness.
- MRI revealed progression of nodular thickness (18x15mm)through hypothalamus (Figure 2 a-b). Secondary hypogonadism was detected while other pituitary functions were normal .
- Multiple, irregularly shaped cystic lesions at he posterior and upper lobes of lungs and multiple lymph nodes with max 20 mm diameter was observed in computed tomography (Figure 4).
- There were no bone lesion in direct radiographies and there was no skin involvement (Figure 5a).
- Open thoracoscopic biopsy performed by surgeon and revealed LCH on histopathological examination with positive CD68, S100 and CD1a staining.
- After diagnosis, PET-scan performed and reveals increased osteoblastic activity at the 6th, 8th and 9th right ribs and in the central part of the left femoral shaft (Figure 5b). Bone marrow biopsy was reported as normocellular.
- External radiotherapy and six cycle of chemotherapy (vinblastine + prednisolon) was planned. Also she was on desmopressin, prednisolone and levothyroxine treatment.

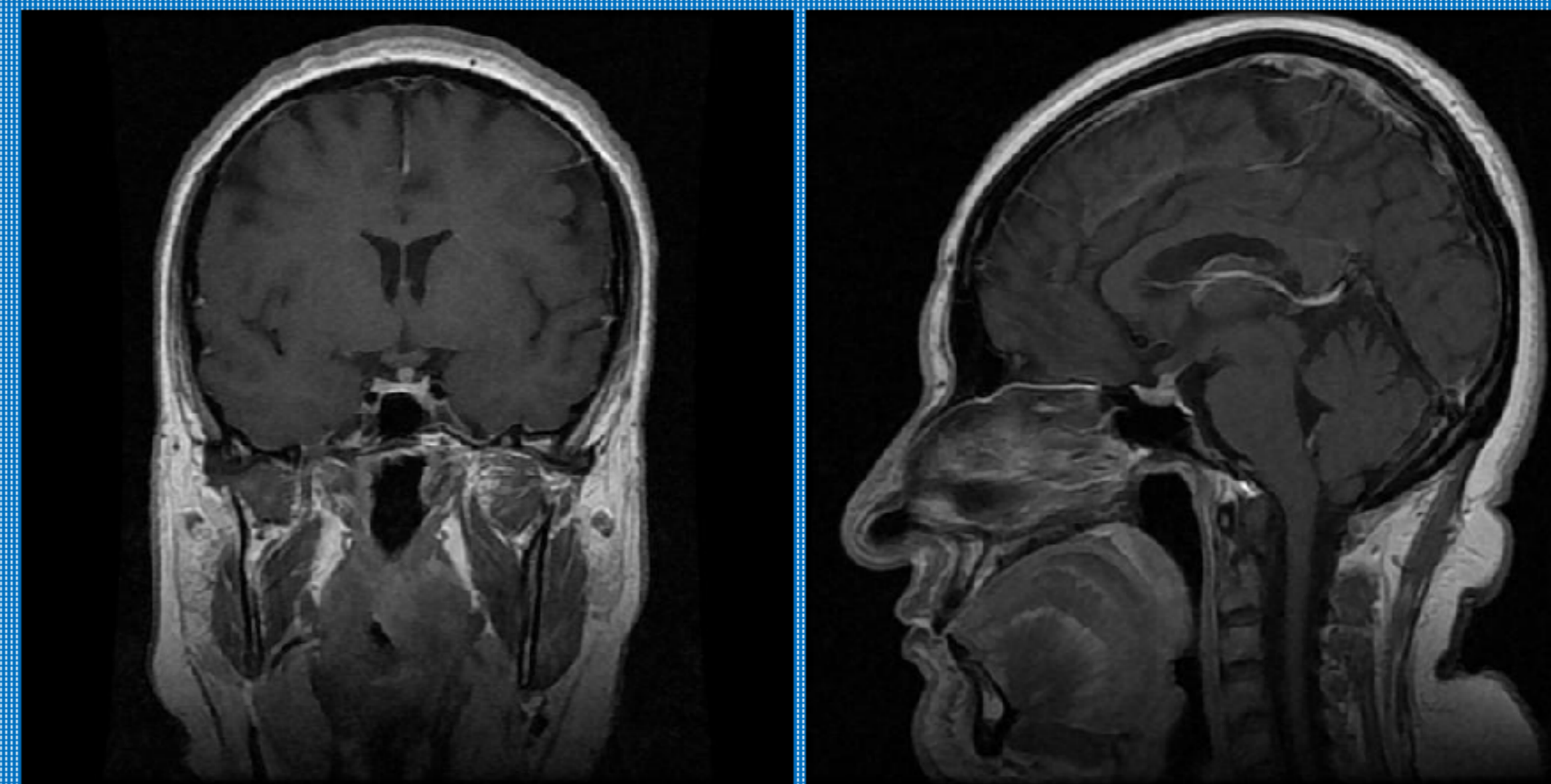


Figure 1 a-b

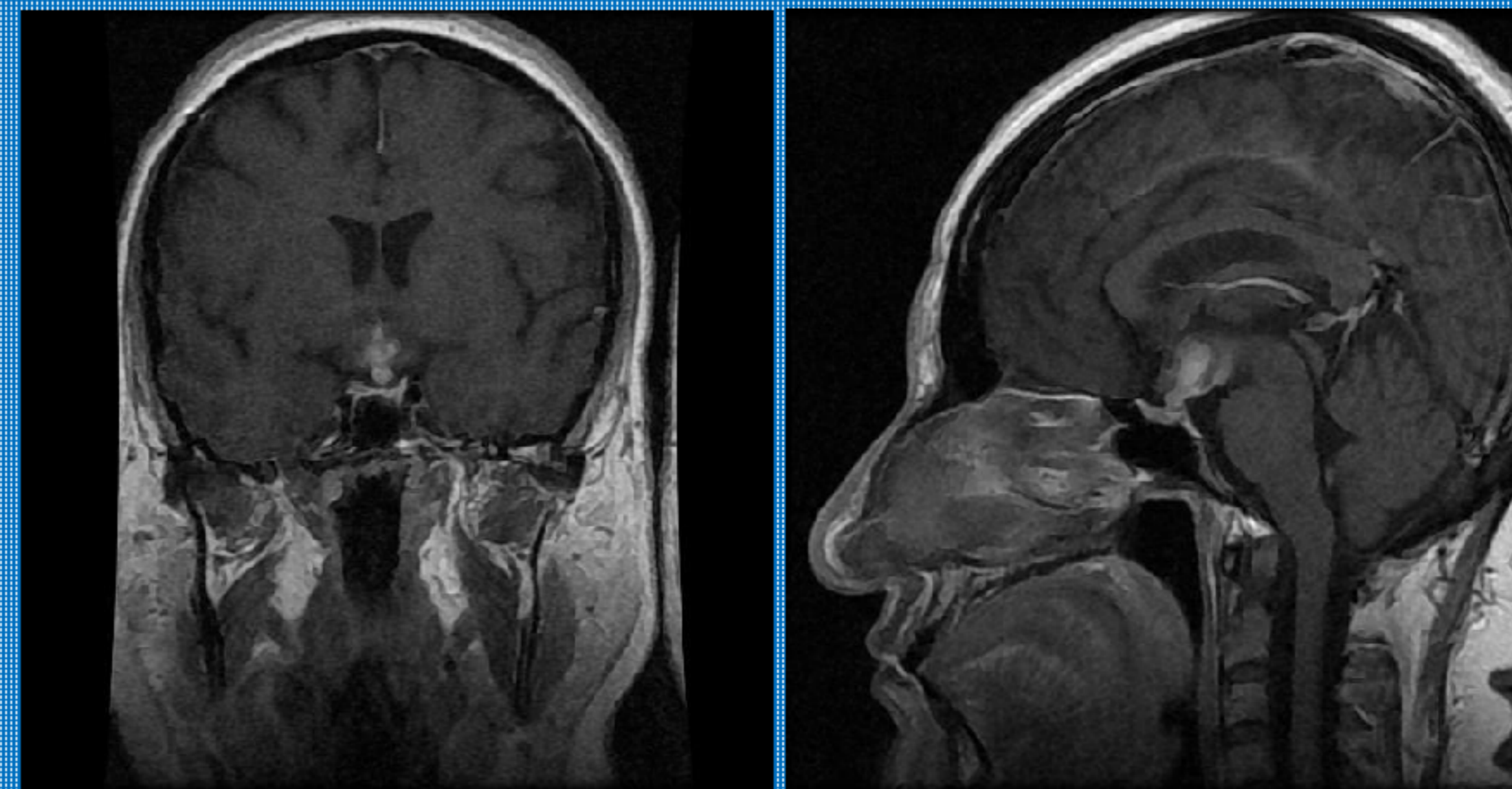


Figure 2 a-b

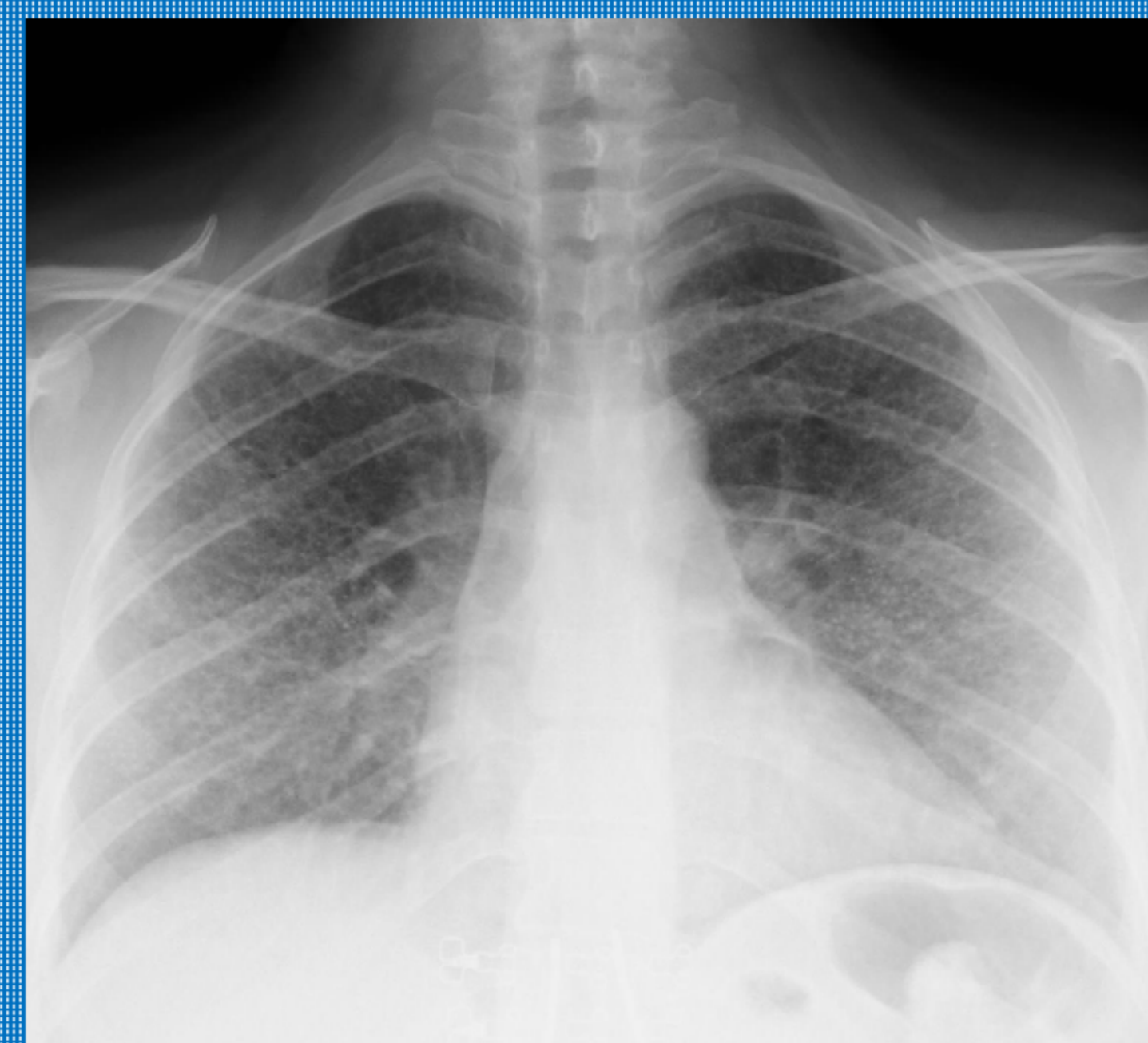


Figure 3

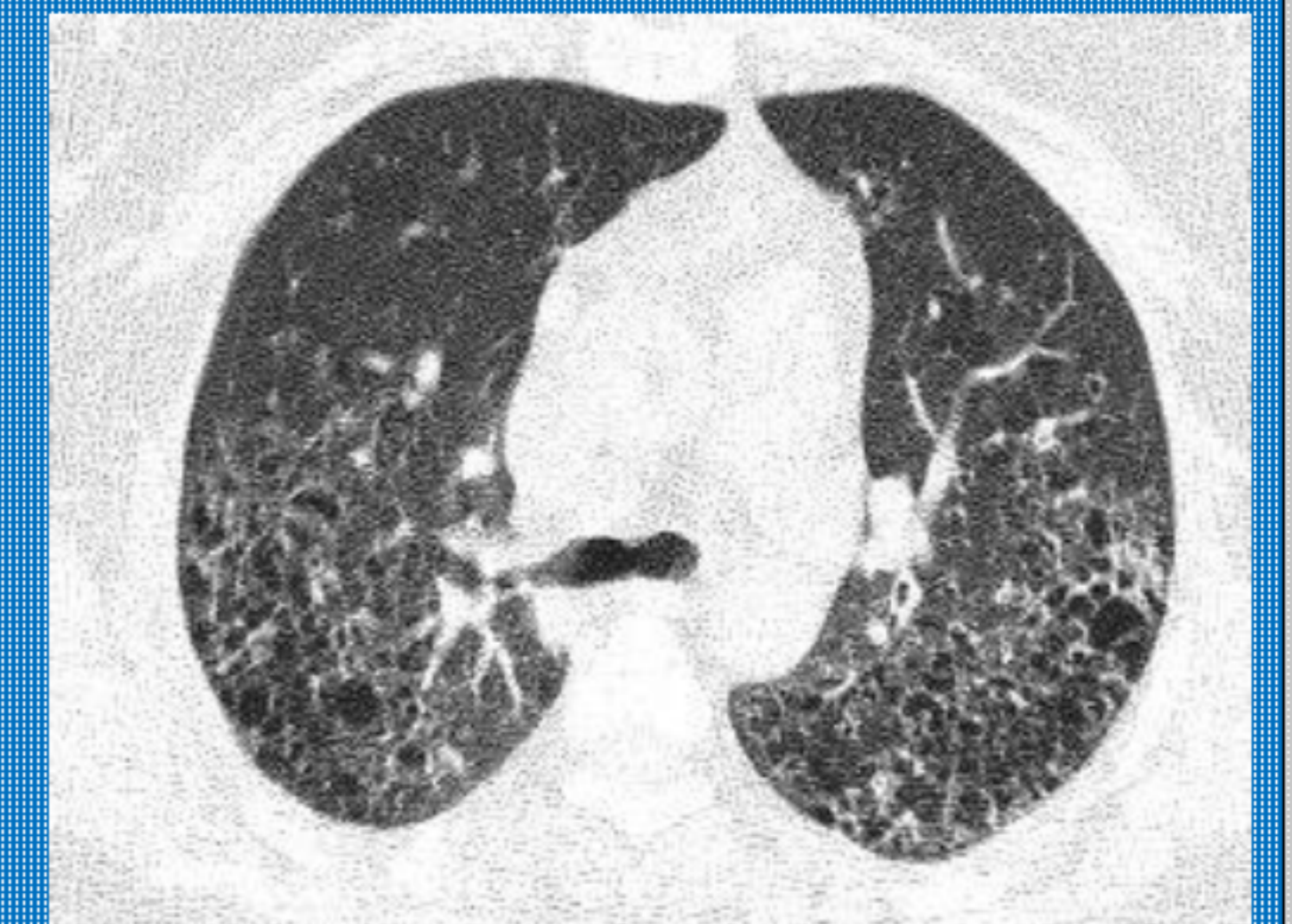
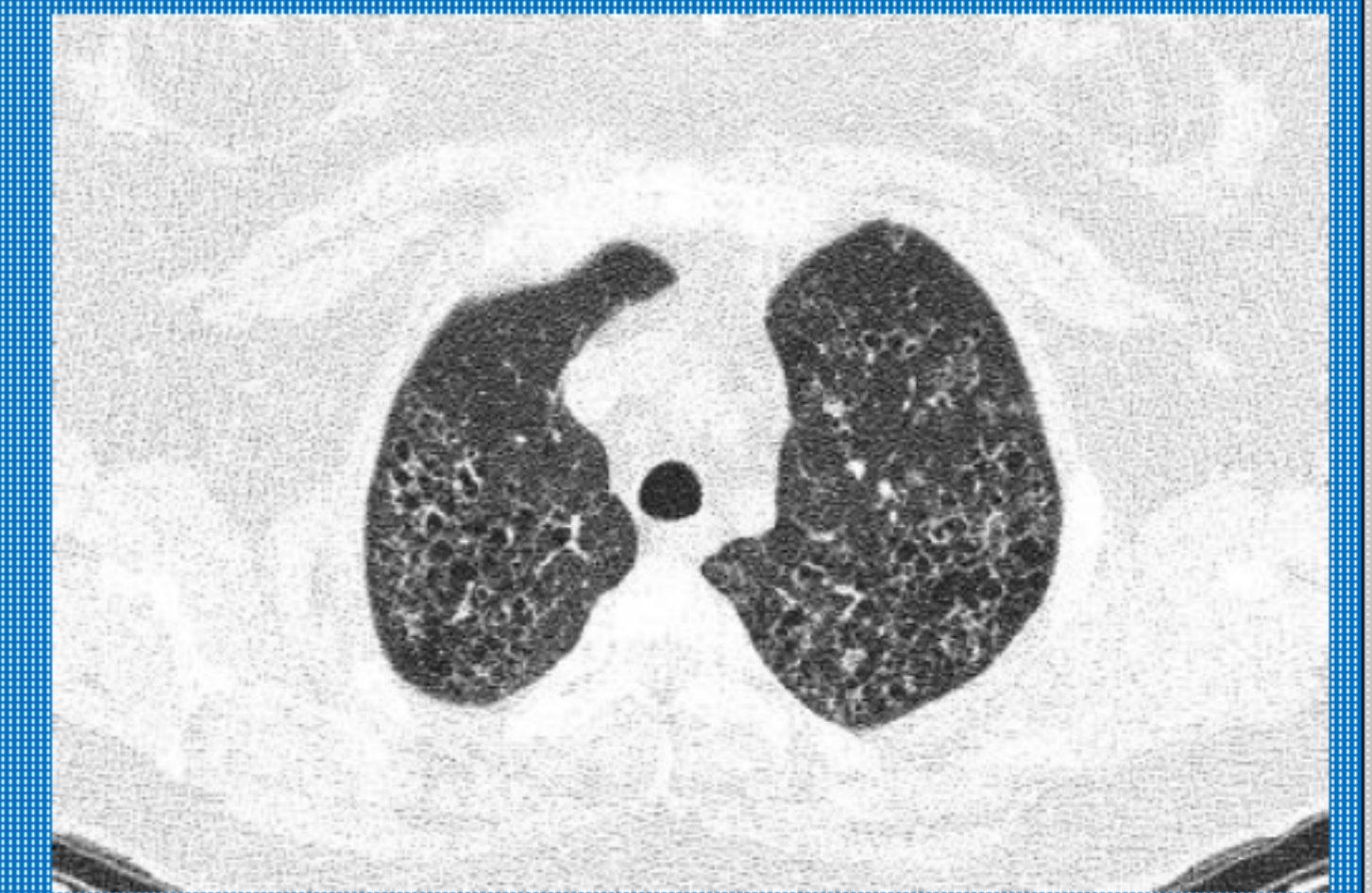


Figure 4 a-b

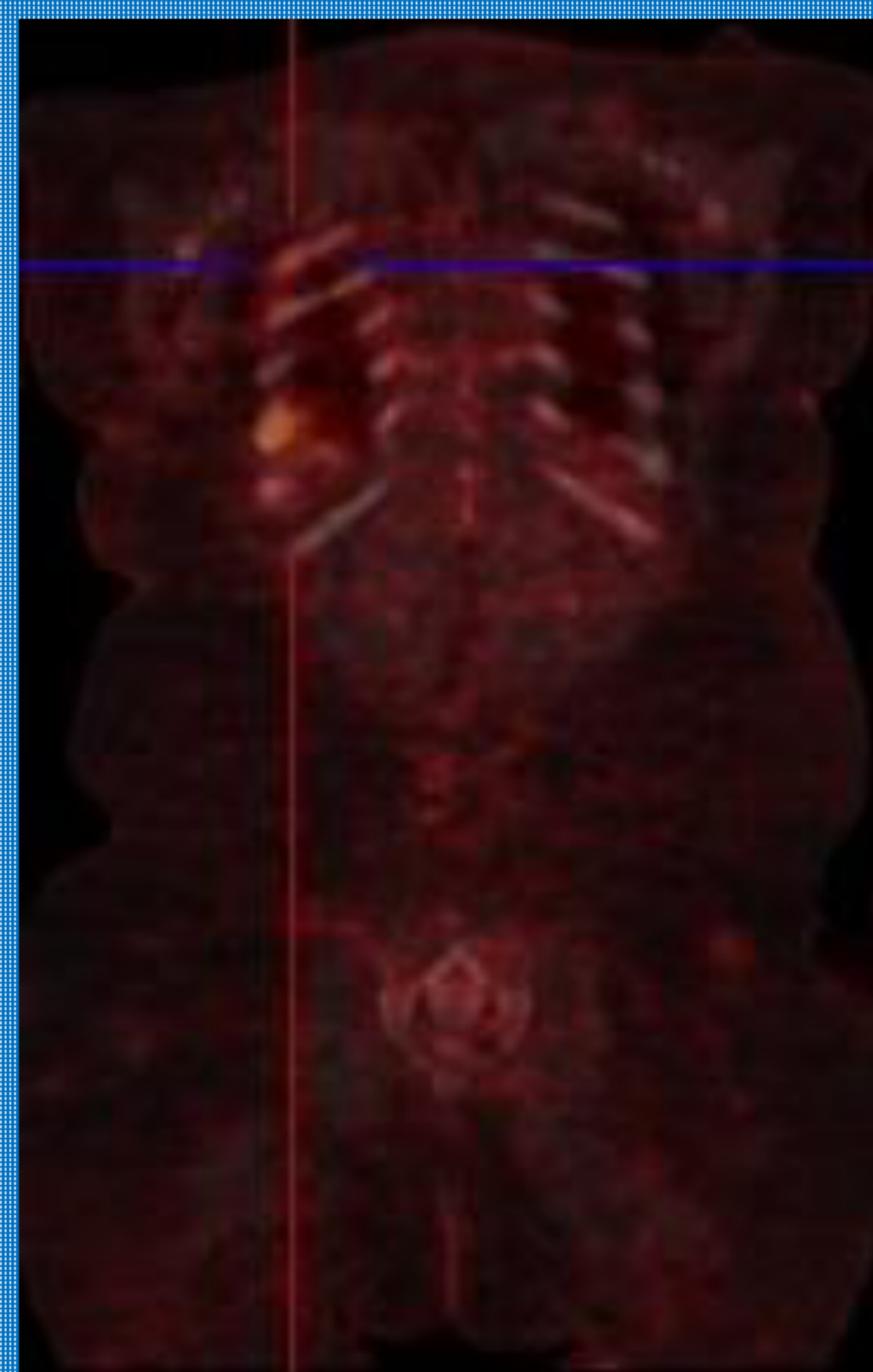


Figure 5 a-b

Conclusions

- We reported this case to refer to diabetes insipidus as a rare presentation of Langerhans cell histiocytosis which a rare and some times multisystemic progressive disease.
- Diabetes insipidus, is an important indicator for stalk lesions and may be marker of very rare conditions such as atypical tumor of the sellar region, sarcoidosis, lymphocytic hypophysitis or histiocytosis like in this case.
- It should be monitored dynamically because of the possibility of progressive nature of the disease.

