

Combined cytarabine and cladribine chemotherapy in an adult patient with refractory Langerhans cell histiocytosis

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Background:

- We report the case of a 25 year old male patient with refractory Langerhans cell histiocytosis (LCH)

Patient history:

- The male patient was diagnosed with LCH and the age of 13 with multifocal bone involvement (cranial, pelvic and femoral bones).
- First -line treatment according to the LCH-2-protocol (a 6-week course of oral steroids and weekly vinblastine, followed by threeweekly pulses with the same drugs).
- At the age of 21 there was a reactivation of LCH with lymph-node involvement. Therapy was initiated with vinblastine and methyl-prednisolone and consecutive maintenance therapy with 6-mercaptopurine and methotrexate until 06/2008.
- Five months after the end of maintenance therapy a new LCH reactivation was documented. Oral therapy with 6-mercaptopurine and methotrexate was restarted and continued for 2 years when the patient developed a methothrexate-induced hepatopathy.
- The patient presented to us for the first time 12 years after initial diagnosis with ulcerated lesions at the forehead (Fig. 1.).

- The CT-scan showed cutaneous and subcutaneous lesion at the skull, the pelvic and vertebral bones as well as disseminated lymph node involvement (Fig. 2, 3).
- Histology of skin lesions and bone marrow confirmed LCH activity.
- Combined chemotherapy with cytarabine (100mg/m², 2x/d, d1-d5) and cladribine (5mg/m², d2-d6) in 09/2011 according to a protocol which had demonstrated activity in childhood refractory LCH (Bernard et al., 2005) was started in a reduced dose (75%).
- The treatment course was repeated on day 29. During the treatment the patient showed severe nasal bleeding due to thrombocytopenia. After the second cycle treatment was continued with 2-CDA only.
- After the fourth cycle treatment was stopped due to severe marrow suppression.
- A rapid clinical response and complete radiological remission was observed already after the second chemotherapy course.

- Clinical and radiological follow up three months after the end of treatment (04/2012) showed complete remission. Meanwhile the patient could restart working daily.



Fig. 1: Ulcerated lesion at the forehead caused by LCH



Fig. 2: CT-scan of vertebral LCH lesion



Fig. 3: CT-scan of vertebral LCH lesion

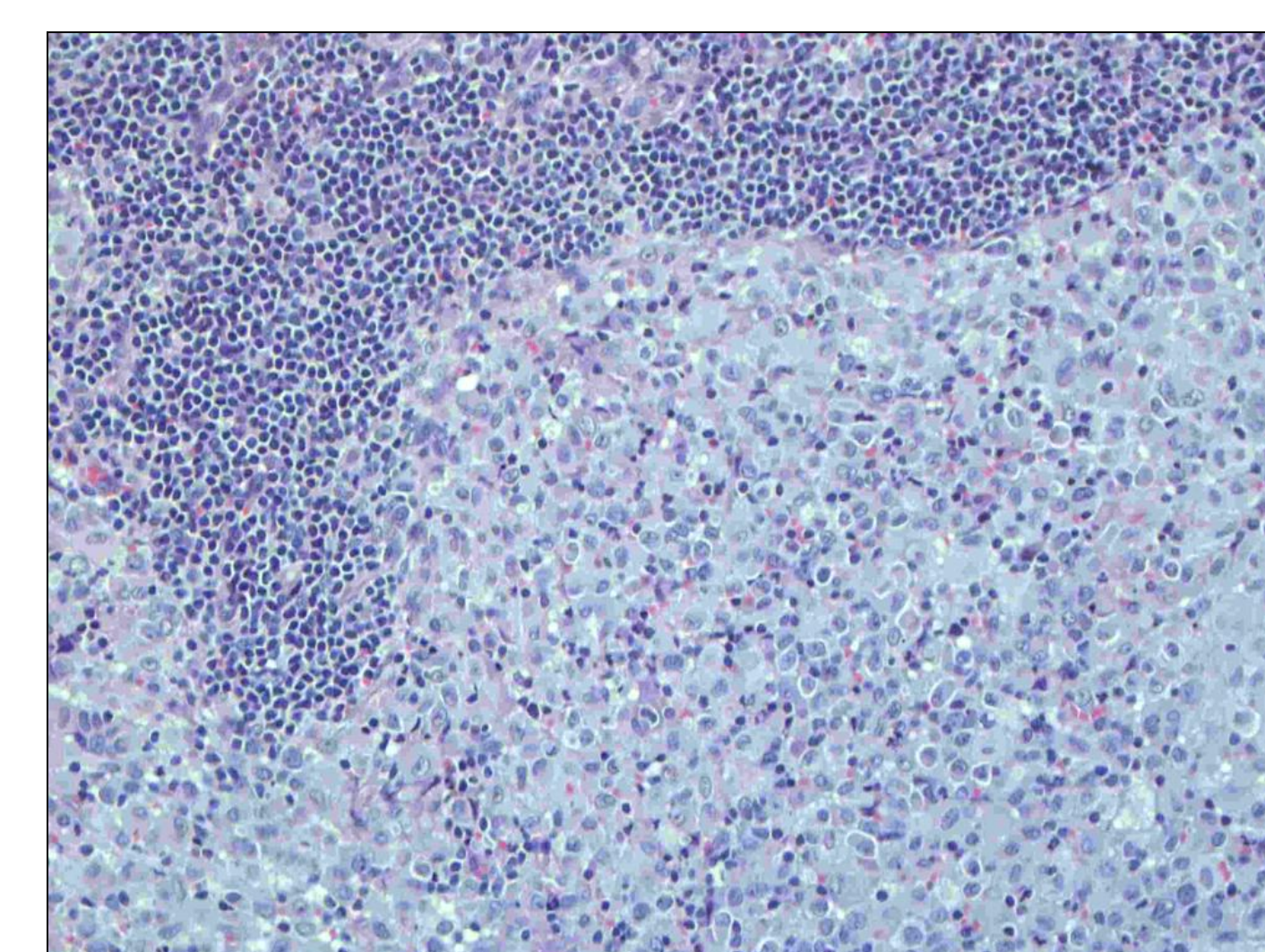


Fig. 4: Histology of a lymph node biopsy showing infiltration with LCH-cells

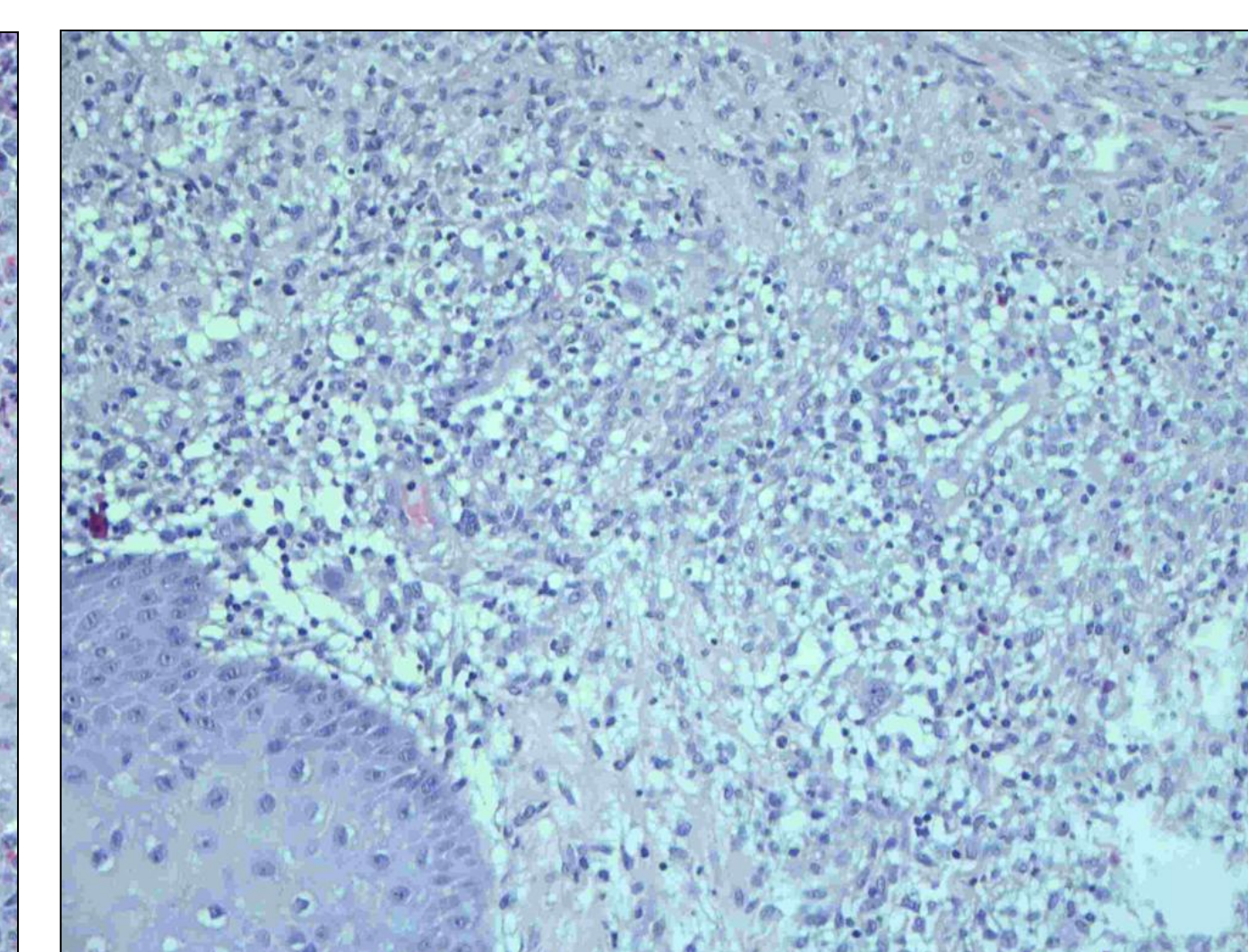


Fig. 4: Histology of a skin biopsy showing infiltration with LCH-cells