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Langerhans cell sarcoma: case report and literature review

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Introduction: Langerhans cell sarcoma (LCS) is an extremely rare neoplasm of Langerhans cells with malignant cytological features, which is highly aggressive with >50% mortality from progressive disease. According to the most recent WHO Classification of Tumours it belongs to the category of histiocytic and dendritic cell neoplasms. Herein we report a case of a 62-year-old patient with an osteo-destructive lesion of the left scapular.

Case Presentation: We present the case of a 62-year-old male patient, who was advised to our department with a three-month history of pain in the left shoulder. Plain radiographs and magnetic resonance imaging of the scapular showed a destructed bone with multiple osteolysis. For diagnostic reasons an incisional biopsy was performed. Based on histopathological and immunohistochemical investigations a Langerhans cell sarcoma was diagnosed. Further staging including bone scan and computed tomography of chest, abdomen and pelvis revealed loco-regional metastases in the left shoulder, lymph nodes as well as multiple metastases in the liver. Therefore, a palliative treatment was introduced. At three-months of follow-up the patient was alive with disease.

Conclusion: Tumours of histiozytes are among the rarest of tumours affecting the lymphoid tissues. Histopathology and immunohistochemistry are necessary for diagnosis. The immunophenotype of the LCS is identical to that of Langerhans cell histiocytosis (LCH) and can be distinguished from LCH by malignant cytological features, such as high mitotic rate with atypia. The prognosis in patients with experienced multi-organ involvement is bad, solitary lesions showed favourable outcome. The therapy includes: surgery, chemotherapy as well as radiation and has to be adapted from case to case. To achieve the correct diagnosis of this rare high grade entity and thereby the adequate treatment, a multidisciplinary approach is essential.