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Pathology Page

Langerhans cell histiocytosis of the bone

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A 1-year-old male child was brought to the clinic by his mother with progressive right shoulder swelling for 3 weeks. His mother denied any history of specific exposure or trauma. No relevant birth or developmental history or family history was reported. He has followed the recommended immunizations.

Physical examination showed a firm, tender, 2.5 cm mass in the right proximal clavicle area. Laboratory data were within reference levels. Magnetic resonance imaging revealed a 2.8 cm × 2.4 cm × 2.4 cm mass in the right proximal clavicle [Figure 1]. It had lobulated and solid patterns with relatively homogeneous contrast enhancement. Focal anterior cortical destruction with periosteal soft-tissue edema was present. A bone biopsy was performed. Histopathologic examination revealed round to oval-shaped tumor cells with reniform nuclei, fine chromatin, and focal nuclear grooving, admixed with eosinophilic and other inflammatory infiltrates [Figure 2a]. The tumor cells were diffusely positive for S-100 protein and CD1a [Figure 2b]. Langerhans cell histiocytosis (LCH) was diagnosed. A whole-body bone scan showed a solitary bone tumor in the right clavicle with no other bone lesion. The family shifted the patient to another hospital for further management. Further image study confirmed that his disease was limited to one organ system. He had received chemotherapy, and the screenings have demonstrated decreased tumor size. He has had scheduled treatment and regular follow-up.

LCH is a rare histiocytic disorder that typically occurs in children but may occur at any age. LCH mostly develops in one organ system but may present with multisystem disease. It can involve the bone, skin, lymph nodes, lungs, or other rare locations. The etiology of LCH remains unknown. According to molecular study, LCH is not a disease of the epidermal Langerhans cells, but rather one of the myeloid dendritic cells with mononuclear phagocyte dysregulation. The clinical signs and symptoms of LCH vary depending on the organs and extent of involvement. LCH is diagnosed based on pathologic and immunohistochemical evaluation. Histologic features are not predictive of the clinical outcome. Tumor cells are positive for CD1a and CD207 (langerin). Birbeck granules are demonstrated by electron microscopy.

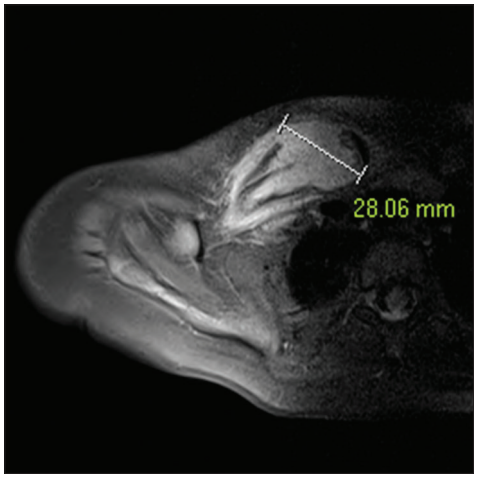

Figure 1: Magnetic resonance imaging reveals a mass in the right proximal clavicle

According to the patient presentation, pretreatment examinations may include laboratory and radiographic

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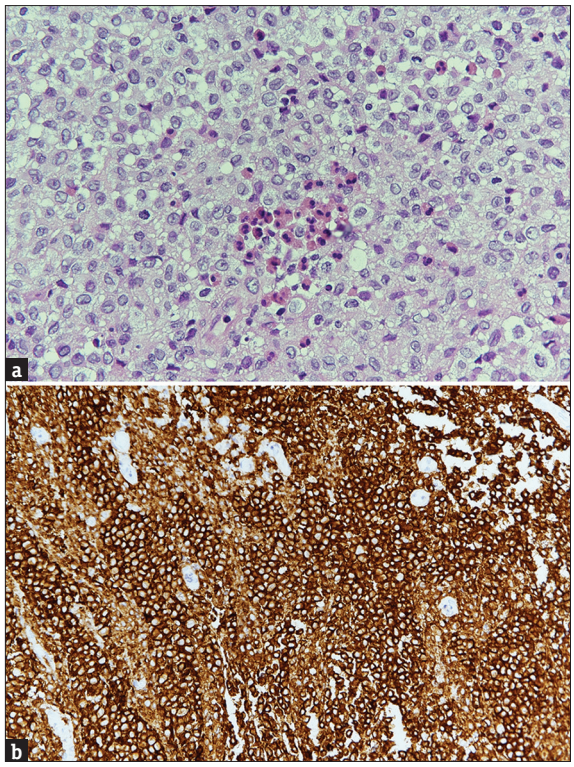

Figure 2: (a) Histopathology shows tumor cells in an inflammatory background (H and E, ×400). (b) CD1a immunostain (×200)

studies to determine the extent of disease. The choice of treatment is based on the disease severity, and whether there is single-system or multisystem involvement. High-risk organs include the bone marrow, liver, and spleen and denote a worse prognosis. *BRAF V600E* mutation has been detected in the majority of patients with multisystem LCH. A solitary bone lesion can be treated with curettage. A combination of cytotoxic chemotherapy and steroids are indicated for polyostotic bone lesions and multisystem disease.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Declaration of patient consent

The authors certify that the patient has obtained appropriate patient consent form. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initial will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Further reading

1. Allen CE, Li L, Peters TL, Leung HC, Yu A, Man TK, et al. Cell-specific gene expression in Langerhans cell histiocytosis lesions reveals a distinct profile compared with epidermal Langerhans cells. J Immunol 2010;184:4557-67.
2. De Young B, Egeler RM, Rollins BJ. Langerhans cell histiocytosis. In: Fletcher CD, Bridge JA, Hogendoorn PC, Mertens F, eds. WHO Classification of Tumours of Soft Tissue and Bone. 4th ed., Lyons: International Agency for Research on Cancer; 2013. p. 356.
3. Gadner H, Grois N, Potschger U, Minkov M, Aricò M, Braier J, et al. Improved outcome in multisystem Langerhans cell histiocytosis is associated with therapy intensification. Blood 2008;111:2556-62.