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Langerhans Cell Histiocytosis of the Cervical Spine

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Manuscript Text

31-year-old male with history of diffuse Langerhans cell histiocytosis (LCH) involving the left mastoid base status post resection and radiation presented with progressive severe right-sided neck pain. CT and MRI revealed an osteolytic lesion at C5 (Figure 1) and due to concern for mechanical instability, the patient underwent a two-stage anterior C5 corpectomy and posterior cervical fusion with instrumentation (Figure 2). Final pathology of the surgical tissue confirmed LCH (Figure 3). Post-operatively, the patient's pain improved and he continues to undergo chemotherapy for his systemic disease. LCH is a rare disease commonly affecting the bones and spinal involvement occurs in 6.5-25% of all skeletal cases with 11% affecting the cervical spine[1]. Patients commonly present with localized neck pain, restricted range of motion or neurological deficits[2-4]. Imaging frequently reveals an osteolytic lesion, though osteoblastic lesions can occur[2]. LCH of the spine may be treated conservatively with observation, immobilization, NSAIDS and casting or with radiation, steroids or chemotherapy—especially in systemic disease or multifocal spine disease[1-2, 4-5]. Surgical intervention—i.e. curettage with or without bone grafting, internal fixation and fusion or percutaneous vertebroplasty—may be indicated in cases of severe mechanical instability, deformity and/or neurological deficits caused by compression[1, 2].

Figure 1: MRI (A) and CT (B) imaging showing an expansile lytic lesion associated with erosive changes within the C5 vertebral body

Figure 2: Post-operative CT imaging, with and without contrast, showing stable corpectomy at C5 with posterior fusion of C4-C6

Figure 3: Histopathology of LCH involving the cervical spine. A histiocytic lesion with numerous admixed eosinophils is seen diffusely involving the vertebral body and partially eroding bone (A, H&E, 20X lens, bone indicated by '*'). The histiocytic cells were strongly CD1A positive (B, immunoperoxidase, 20X lens). Closer examination of the neoplastic cells revealed characteristic Langerhans cells with elongated and folded nuclei (C, H&E, 40X, arrow indicating cells with more prominent nuclear features).

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