Original Article

First technical report of a pediatric case with thoracic Langerhans cell histiocytosis: Gross total tumor removal, corpectomy, and 360° stabilization via posterolateral approach at a single stage

ABSTRACT

Background: Langerhans cell histiocytosis (LCH) is a rare nonmalignant disease characterized by a clonal proliferation of mononuclear cells called Langerhans histiocytes and infiltrates surrounding tissues, mostly self-limiting and usually occurring in the first two decades of life. Vertebral involvement is rare, mostly seen in the thoracic region, and involves the anterior elements of the corpus. In the literature, several treatment options and surgical approaches have been reported concerning the treatment of this disease and surgery.

Case Presentation: We report an 18-month-old male with thoracic LCH who underwent surgery due to progressive neurological deficit. Gross total removal of the tumor with one level corpectomy in this patient was achieved via a posterolateral approach with postoperative functional improvement. The surgical cavity was supported by corpectomy cage and unilateral screw-rod fixation system at the same stage.

Conclusion: Gross total tumor removal, corpectomy, and 360 stabilization via posterolateral approach at a single stage are safe, effective, and definite neurosurgical methods in terms of providing neurological recovery, long-term tumor-free survival, and spinal stability.

Keywords: Corpectomy, eosinophilic granuloma, Langerhans cell histiocytosis, spinal tumor

INTRODUCTION

Langerhans cell histiocytosis (LCH) is an uncommon disorder characterized by uncontrolled proliferation and accumulation of lipid-containing histiocytes from the reticuloendothelial system.[1] Proliferation of histiocytes in medullary bone results destruction of cortical bone with extension into adjacent soft tissue. Eosinophilic granuloma (EG) is a unifocal benign local process of LCH presents with a solitary bone lesion, accounts for 75% of LCH cases but composes <1% of all bone tumors[2] and often seen in the first or second decades of life. It is thought that the lesion causes bone destruction due to the secretion of interleukin (IL)-1 and PGE2.[3] Skull is the most frequent site for occurrence in the skeletal system.[4] Vertebral involvement occurs in 10%–17% of affected patients with a male predominance mostly seen in the thoracic region and involves the anterior elements of the corpus.[5] In this article, we present a pediatric case with thoracic EG who has progressive symptoms of spinal cord compression. The surgical technique, approach, and procedures described in this report are of high importance to the pediatric neurosurgery due to their implementation for the first time.

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CASE

An 18-month-old male presented with a complaint of gait disturbance due to progressive loss of strength in both legs. On neurological examination, paraparesis, slightly increased deep tendon reflexes in the lower extremities, bilateral positive Babinski signs, and clonus were observed. Spinal computed tomography (CT) scan showed the total collapse of T3 vertebral body [Figure 1]. Magnetic resonance imaging of the spine revealed that collapsed body of T3 vertebra surrounding by enhancing lesion compressing the spinal cord anteriorly with uninvolved adjacent disc spaces [Figure 1]. Following the intubation of the patient, neuromonitoring modalities including somatosensory-evoked potential, motor-evoked potential, and electromyogram were placed. Then, the patient placed in the prone position and the head was fixed in a clamp. The cervical spine maintained in a neutral position. Pathological level was identified using C-arm fluoroscopy and T1-T5 segments were marked. After image acquisition, a midline skin incision was performed with a T-shape over the left third rib. Fascia was divided and paraspinal muscles were dissected. The muscles are then elevated off over the transverse processes and ribs to expose the costotransverse articulations. Under an operative microscope, T2-3-4 total laminectomy and then left T3-4 costotransversectomy were performed by bone-cutting device (Piezo Surgystar®, Dmetec Piezo Bone Surgery, South Korea) [Figure 2a, c and d]. Dirty gray-colored tumor tissue that destructed the corpus of T3 vertebra but was not invaded into the dura was encountered [Figure 2b]. Total T3 corpectomy, two-level discectomy, and gross total excision of tumor were performed via left posterolateral approach under the microscope [Figure 2e and f]. End plates were preserved. Anterior longitudinal ligament was seen as the ventral limit of corpectomy. Decompression of spinal cord and nerve roots was observed. T2-4 interbody fusion was performed with corpectomy cage placement and stabilized with a screw-rod fixation system under fluoroscopic guidance. Total excision was achieved with no additional neurological deficits in the postoperative period. Frozen section was reported to be compatible with LCH. The patient was discharged with a cervicothoracic rigid brace and approximately 20% strength regain in the lower extremities in the early postoperative period. Full neurological recovery was achieved 3 weeks after the surgery. Histopathologic examination revealed diffuse infiltration of histiocytic cells with eosinophils and lymphocytes. The histiocytic cells had irregularly folded vesicular nucleus and large eosinophilic cytoplasm. Immunohistochemical analysis showed positivity for S100, CD1a, and langerin [Figure 3].

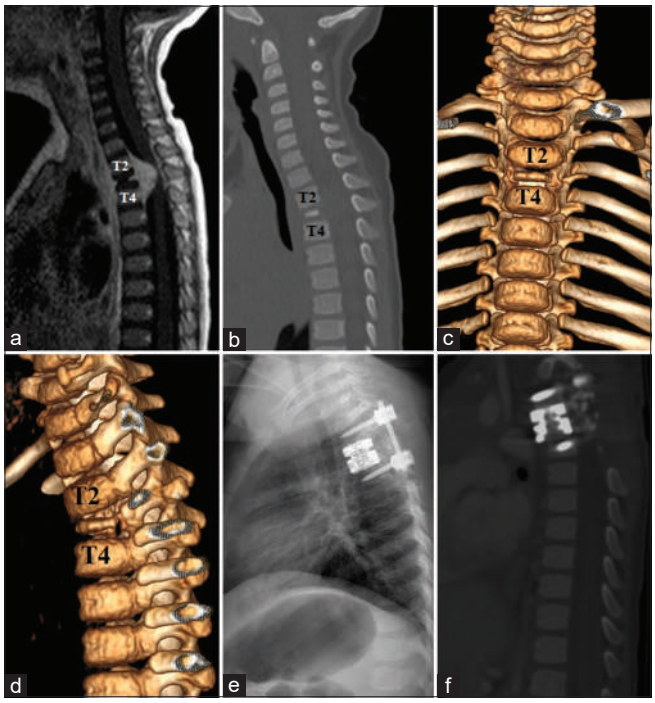

Figure 1: (a) Gadolinium-enhanced sagittal image of cervicothoracic magnetic resonance imaging demonstrated osteolytic destruction of the T3 vertebral body and contrast enhancement of paravertebral soft tissue with compression of the spinal cord. (b-d) Collapse of T3 vertebral body. (e and f) Instrumentation was placed between T2 and T4

DISCUSSION

Osteomyelitis, neuroblastoma, chondroma, Ewing sarcoma, osteosarcoma, leukemia, and lymphoma comprise the differential diagnoses for EG that may also present with vertebral collapse.[6] Histopathologic diagnosis of EG requires the demonstration of S-100 or CD1a and langerin (CD207)-positive Langerhans cells or ultrastructural demonstration of the presence of cytoplasmic tennis racquet-shaped Birbeck granules in lesional cells.[7] If a diagnosis of EG is confirmed, observation with conservative management and immobilization is recommended for patients without spinal instability or neurological deficit.[8-10] The condition seems to be self-limiting, regressing spontaneously with good prognosis and recurrence ratings are low in the pediatric population.[11] Patients who have partially regained the vertebral body height have been reported.[9,12] CT-guided intralesional injection of corticosteroids has been proved effective and safe with good responses in the literature, recommended for patients who are neurologically intact and without a structural deformity or surgery could not be performed.[5,13] Corticosteroids are thought to inhibit the release of secretion of ILs and prostaglandins as bone resorptive agents from Langerhans cells.[3] Low-dose radiation has also proven effective in the treatment of EG but care must be taken in children about the development

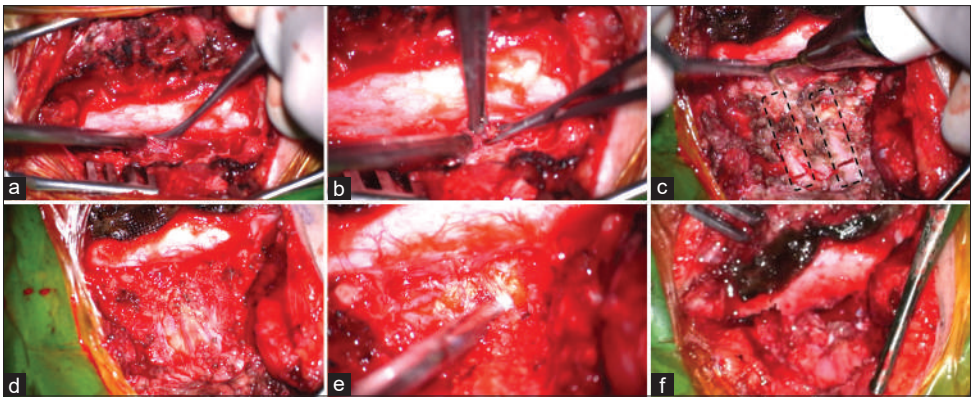

Figure 2: (a) Wide and elongated laminectomy preventing spinal cord compression. (Left side of the picture represents the cranial direction and right side of the picture represents caudal direction.) (b) The biopsy sampling and verification of the lesion. (c) Exposure of unilateral (left side) two-level costotransverse joints, costae and performing the costotransversectomies using a bone scalpel. (Black dotted rectangular areas represent 3rd and 4th ribs.) (d) The surgical cavity after costotransversectomy. (e) Tumor resect. (f) The final view of the surgical field after total tumor removal and before the corpectomy cage placement

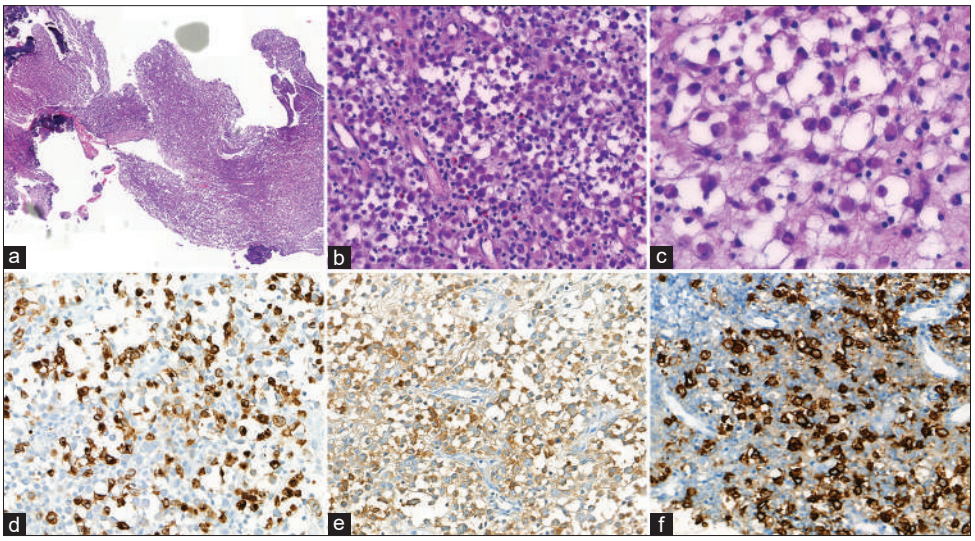

Figure 3: (a) HE × 3 (b) HE × 25 (c) HE × 40 (d) CD1ax25 (e) Langerin × 25 (f) S100 × 25

of radiation-induced tumors in irradiated fields and impair endochondral ossification epiphyseal plates or radiation myelitis.[14,15] Chemotherapy is only recommended for multiple lesions or multisystem disseminated disease and whenever surgery and radiotherapy have failed.[3,16,17] Surgery is rarely indicated for cases who have progressive neurologic deficit due to spinal cord or nerve root compression from the collapsed vertebra or tumor tissue and structural deformity like severe kyphosis or spinal instability when external orthosis has failed. Lu *et al*. reported in their study that posterior approach and fusion with instrumentation is a safe preferable surgical choice for pediatric age patients with thoracic EG for minimal surgical trauma.[18] According to our recent knowledge after available literature was reviewed for thoracic EG in pediatric age population, the case we present in this article is the first and the only one who underwent gross-total removal of the tumor and total corpectomy, interbody fusion with corpectomy cage, and stabilized with screw-rod fixation system through costotransversectomy via posterolateral approach at a single stage.

CONCLUSION

Surgery is recommended immediately for cases with neurological deficit or spinal cord compression with or without spinal instability or deformity. In the literature, although there are numerous neurosurgical procedures about the scheme of treatment to be applied, the posterolateral approach can be considered primarily in these patients due to its safety, effectiveness, and allowing 360° stabilization with gross total tumor removal at a single stage.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's parents have given their consent for patient's images and other

clinical information to be reported in the journal. The patient's parents understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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