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CASE REPORT

Spontaneous and complete regeneration of a vertebra plana after surgical curettage of an eosinophilic granuloma

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Abstract
Purpose The eosinophilic granuloma is a unifocal or multifocal Langerhans cell histiocytosis characterized by an expanding proliferation of Langerhans cells in bones. Skeletal LCH is a rare condition, and vertebral regeneration in cases of vertebral body collapse is even rarer. We report the case of a girl with spontaneous complete healing.
Methods ad results A 3-year-old girl was referred for nighttime back pain, with no fever and no neurologic signs. Within a few days, she developed sudden painful restriction of all spine movements. X-ray and computed tomography (CT) of the spine showed reduced T7 vertebral body height (vertebra plana).The patient underwent T7 curettage and the histopathological exam was suggestive of LCH. Two additional skull lesions were found and therefore she underwent chemotherapy. After 7 years of follow-up, total vertebral reconstruction was observed.
Conclusions Despite the rarity of the condition and despite the rarity of vertebral body lesion resolution, total vertebral body reconstruction was observed over a 7-year period. Long-term follow-up is necessary for a better understanding of the final outcome of patients with EG.

Keywords Eosinophilic granuloma · Vertebral collapse · Case report

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Abbreviations

LCH	Langerhans cell histiocytosis
EG	Eosinophilic granuloma

Background and purpose

Langerhans cell histiocytosis (LCH) is a rare disease (one new case per 2,000,000 persons per year) of unclear etiology involving the clonal proliferation of Langerhans cells, i.e., abnormal dendritic cells. LCH is part of a group of syndromes called histiocytoses that includes three clinical entities: eosinophilic granuloma (EG), Hand–Schuller–Christian syndrome and Letter–Siwe disease [1, 2]. An eosinophilic granuloma is a unifocal or multifocal LCH characterized by an expanding proliferation of Langerhans cells in bones; Hand–Schuller–Christian syndrome is characterized by the triad of diabetes insipidus, exophthalmos and lytic bone lesions, while Letter–Siwe disease is a multifocal multisystem LCH in which Langerhans cells proliferate in many tissues.

The most common presentations of skeletal LCH (EG) are lytic bone lesions, mainly in the skull, followed by the femur, ribs, pelvis and spine [1, 3, 4]. Concerning a spinal location, there is a high prevalence of lesions in the cervical spine [1, 5, 6]; vertebral involvement is reported in 7.8–25% of cases [7, 8].

X-rays of children and adolescents with spinal LCH usually reveal different grades of vertebral body collapse, which can be symmetric or asymmetric, sometimes associated with lesions of the posterior elements [1, 5].

Although not diagnostic, vertebra plana on radiographs is considered a typical radiological feature [1, 2]. Vertebral body collapse can undergo some regeneration, but generally full-body height is not regained. Spontaneous

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resolution of vertebral body lesions is very rare, and in several patients a local deformity has been found [9, 10]. This report describes the case of a patient with a thoracic EG, characterized by the radiographic picture of vertebra plana, which resolved spontaneously over long-term follow-up.

The CARE guidelines were followed while preparing this case report.

Case presentation

In 2009, a 3-year-old girl presented sudden nighttime back pain, with no fever and no neurologic signs. The patient had a negative medical/family history and no co-morbidities. A first X-ray of the spine was prescribed, but it was negative. The hematological examination was normal. Within a few days, she developed further painful restriction of all spine movements, so she was evaluated once more with X-ray, and a computed tomography (CT) scan was ordered as well. These imaging exams showed reduced T7 body height (a vertebral plana not present in the previous X-ray 2 weeks before) (Fig. 1). The diagnostic suspicion was EG, but a differential diagnosis between other neoplastic causes or infective causes was necessary. The patient was admitted to the hospital for T7 surgical curettage: a transpedicular approach was pursued, the curettage was not aggressive and the ring apophysis were preserved, no bone grafting was taken up. The subsequent histopathological examination of the lesion confirmed the diagnostic hypothesis of EG. After the curettage, a Milwaukee brace was prescribed and worn during day for 3 months. After this period, the patient was asymptomatic. A couple of months after discharge, the patient underwent full-body radiographic controls and two additional skull lesions due to EG were identified. No other tissues were involved, thus the diagnosis became multifocal EG. Chemotherapy (vinblastine 3.7 mg IV) was started and was carried on for 1 year. The patient has been followed up at the Oncology Institute for 7 years, and no relapses have been reported. In 2010, a magnetic resonance imaging (MRI) exam confirmed the vertebral deformation without the appearance of altered signals in the bones or in the soft tissue (Fig. 2). Consecutive X-rays of the spine showed partial vertebral reconstruction from 2009 to 2011 and total vertebral reconstruction in 2015 (Fig. 3).

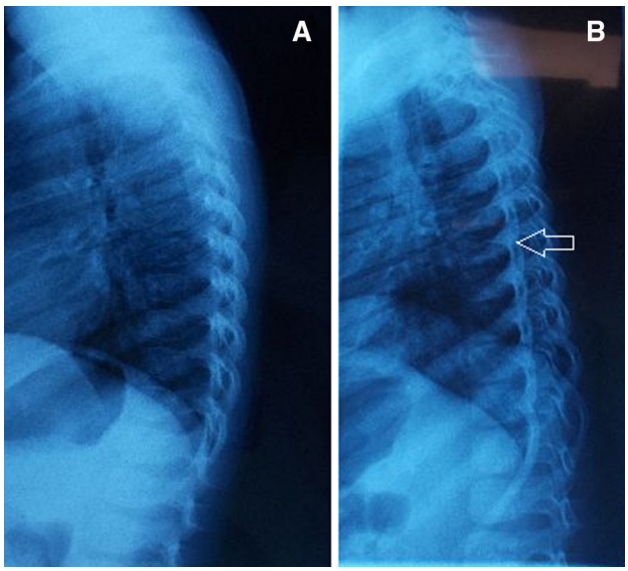

Fig. 1 **a** X-ray lateral view of thoracic spine showing normal vertebral morphology, first exam in March 2009. **b** X-ray lateral view of thoracic spine, repeated after 2 weeks, showing collapse of the T7 vertebral body (*arrow*)

Discussion

The clinical presentation of patients with LCH involving the spine includes neck or back pain, restricted motion of the spine, neurologic symptoms and deformity [1]. Radiologic evaluation of a patient who presents with osseous vertebral lesions often includes radiography, CT and MRI [2, 8]. The radiographic features of a typical LCH spinal lesion consist of complete or incomplete collapse of the vertebral body (vertebra plana is very common in pediatric patients compared to adults), preservation of adjacent disk spaces and the absence of an adjacent paravertebral soft-tissue shadow [7, 8, 11]. Data concerning the involvement of the vertebral posterior elements are discordant: some authors report that preservation of these elements is typical, while others report considerable involvement of posterior elements, i.e. from 40% to more than 60% of cases [1, 7–9, 11]. CT is the most accurate method for evaluating

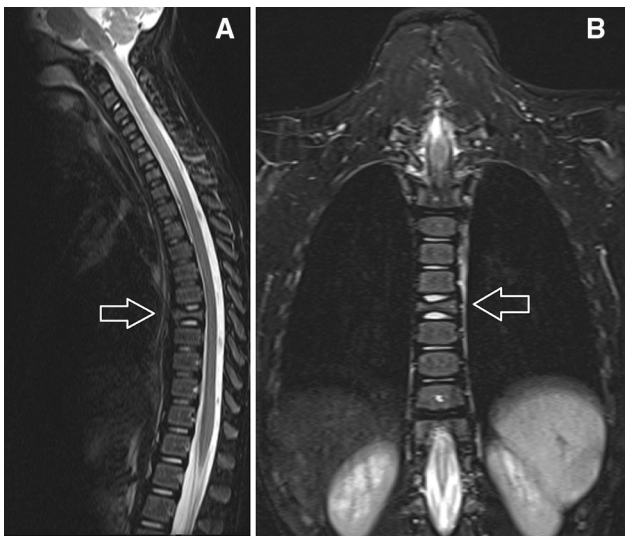

Fig. 2 **a**, **b** RMI exam in 2010. STIR sagittal and coronal images show T7 vertebral body collapse (*arrows*)

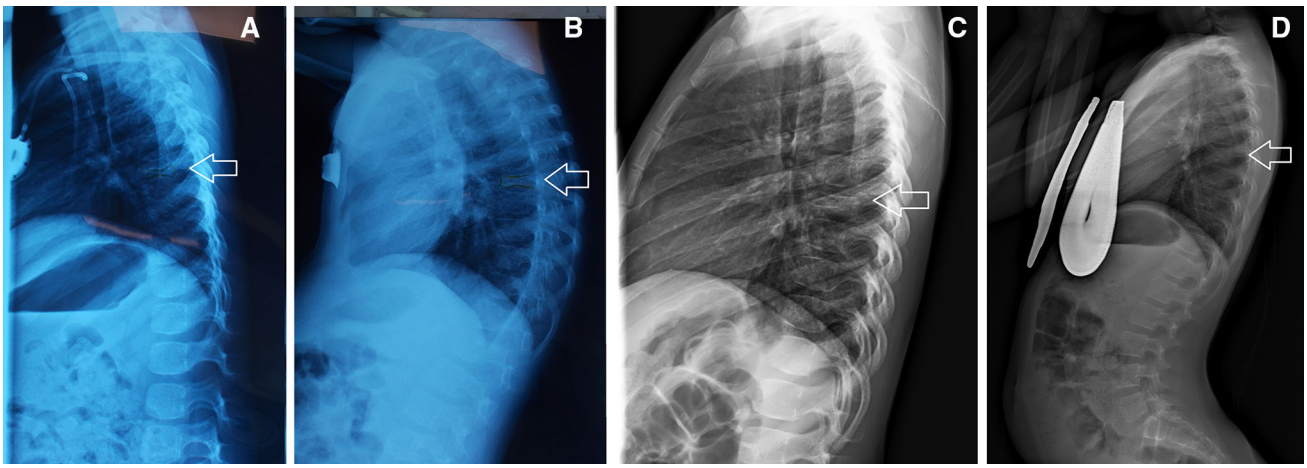

Fig. 3 X-ray lateral view of thoracic spine showing a progressive T7 vertebral reconstruction from 2009 to 2015 (*arrows*); **a** 2009 6 months after the diagnosis, **b** 2010, **c** 2011, **d** 2015

the anatomic involvement of the vertebrae and the degree of cortical bone loss; it also helps to estimate vertebral body collapse. MRI is the best imaging modality for the evaluation of soft-tissue involvement, showing a low to intermediate signal on T1-weighted imaging and a hyperintense signal on T2-weighted imaging [1, 8].

In the pediatric population, acquired pathological vertebral collapse could refer to a range of differential diagnoses. First of all, we have to differentiate between a pathological fracture and a post-traumatic one, but this clearly depends upon the patient's history. Then, it is important to discern between infectious causes, such as pyogenic osteomyelitis or tuberculous osteomyelitis, between neoplastic causes such as osteosarcoma, Ewing sarcoma, fibrous dysplasia, an aneurysmal bone cyst, acute lymphoblastic leukemia/lymphoma and metastases (even if less common than in adults), and between miscellaneous causes such as idiopathic osteoporosis, Langerhans cell histiocytosis and chronic recurrent multifocal osteomyelitis [12].

Concerning vertebral EG, treatment options could be bracing, radiation therapy and chemotherapy, used alone or in combination. A solitary bone lesion can be treated with observation, excision or local radiotherapy, while disseminated disease needs chemotherapy [2, 9, 11, 12]. In cases of solitary EG of the spine, some authors do not suggest observation and recommend open biopsy rather needle aspiration biopsy in order to exclude Ewing's sarcoma, neuroblastoma or bony manifestations of leukemia [13].

Total spontaneous recovery of vertebral body lesions is quite unusual [9–11]. There are data concerning vertebral remodeling in EG of the spine associated with conservative orthopedic treatment with immobilization using a brace (from 1 year up to 5 years of bracing); in a group of 14 patients, reconstitution of vertebral height was reported to be between 18.2 and 63.8% of the height of the adjacent vertebrae before skeletal maturity [14]. Even though this patient wore a Milwaukee brace for 3 months after surgical curettage, complete vertebral reconstruction can be defined as spontaneous following years of brace use.

Conclusions

This case describes a patient with vertebral collapse caused by EG with total vertebral reconstruction over a period of 7 years. The scientific literature reports that, in cases of vertebral body collapse, complete vertebral regeneration is rare. Long-term follow-up period is necessary for a better understanding of the final outcome of patients with EG.

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Compliance with ethical standards

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Conflict of interest None of the authors has any potential conflict of interest.

Informed consent The patient's parents provided informed consent for the publication of this case report.

References

1. Huang WD et al (2013) Langerhans cell histiocytosis of spine: a comparative study of clinical, imaging features, and diagnosis in children, adolescents, and adults. Spine J Off J N Am Spine Soc 13(9):1108–1117

2. Sadashiva N, Rajalakshmi P, Mahadevan A, Vazhayil V, Rao KN, Somanna S (2016) Surgical treatment of Langerhans cell histiocytosis of cervical spine: case report and review of literature. Childs Nerv Syst ChNS Off J Int Soc Pediatr Neurosurg 32(6):1149–1152
3. Bunch WH (1981) Orthopedic and rehabilitation aspects of eosinophilic granuloma. Am J Pediatr Hematol Oncol 3(2):151–156
4. Howarth DM, Gilchrist GS, Mullan BP, Wiseman GA, Edmonson JH, Schomberg PJ (1999) Langerhans cell histiocytosis: diagnosis, natural history, management, and outcome. Cancer 85(10):2278–2290
5. Garg S, Mehta S, Dormans JP (2004) Langerhans cell histiocytosis of the spine in children. Long-term follow-up. J Bone Jt Surg Am 86-A(8):1740–1750
6. Sanchez RL, Llovet J, Moreno A, Galito E (1984) Symptomatic eosinophilic granuloma of the spine: report of two cases and review of the literature. Orthopedics 7(11):1721–1726
7. Yeom JS, Lee CK, Shin HY, Lee CS, Han CS, Chang H (1999) Langerhans' cell histiocytosis of the spine. Analysis of twenty-three cases. Spine 24(16):1740–1749
8. Rodallec MH et al (2008) Diagnostic imaging of solitary tumors of the spine: what to do and say. Radiogr Rev Publ Radiol Soc N Am 28(4):1019–1041
9. Floman Y, Bar-On E, Mosheiff R, Mirovsky Y, Robin GC, Ramu N (1997) Eosinophilic granuloma of the spine. J Pediatr Orthop Part B 6(4):260–265
10. Bavbek M, Atalay B, Altinörs N, Caner H (2004) Spontaneous resolution of lumbar vertebral eosinophilic granuloma. Acta Neurochir (Wien) 146(2):165–167
11. Levine SE, Dormans JP, Meyer JS, Corcoran TA (1996) Langerhans' cell histiocytosis of the spine in children. Clin Orthop 323:288–293
12. Han I, Suh ES, Lee S-H, Cho HS, Oh JH, Kim H-S (2009) Management of eosinophilic granuloma occurring in the appendicular skeleton in children. Clin Orthop Surg 1(2):63–67
13. Immenkamp M (1985) Eosinophilic granuloma of the spine. Z Orthop Ihre Grenzgeb 123(2):227–234
14. Raab P, Hohmann F, Kühl J, Krauspe R (1998) Vertebral remodeling in eosinophilic granuloma of the spine: a long-term follow-up. Spine 23(12):1351–1354