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CASE REPORT

Vertebral Langerhans cell histiocytosis in an adult patient : Case report and review of the literature

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Vertebral Langerhans cell histiocytosis, formerly called Histiocytosis X, is rarely seen in adults : a systematic non-quantitative review of the literature yielded only 27 cases. Vertebra plana is often associated in children, but this is not a feature in the adult population. The authors report the case of a 29-year-old woman with a two-month history of lumbar pain. Osteolysis of the right pedicle of L2 was noted on CT and MRI. Transpedicular curettage and fusion L1L3 were performed. Histopathological examination revealed Langerhans cell histiocytosis. Although rare, Langerhans cell histiocytosis should be included in the differential diagnosis of the solitary lytic vertebral lesion in the adult. Good results have been reported, in the absence of systemic manifestations, with the most conservative approach possible.

Keywords : vertebral ; Langerhans cell histiocytosis ; adult ; solitary osteolytic lesion.

INTRODUCTION

Most cases of vertebral Langerhans cell histiocytosis (LCH) concern children (*19*) ; they are almost always associated with vertebra plana (*2*). Adults are rarely affected : only 27 cases were reported in the English literature. Vertebra plana was mentioned in only one of these cases. The authors present the case of a 29-year-old woman with involvement of L2.

CASE REPORT

A 29-year-old female complained of progressively worsening low back pain, radiating to the right sacro-iliac joint for the past two months. Over the counter non-steroidal anti-inflammatory drugs provided no relief. Low-grade fever up to 37.5° C and night sweats raised the suspicion of an underlying infection or malignancy. Nonetheless, physical examination was unremarkable, as were her laboratory tests. The latter included full blood cell count with differential, serum electrolytes, urea and creatinine, liver function tests, alkaline phosphatase (total and bone-specific), erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). No pathologic findings were seen on plain radiographs. A CT-scan revealed an osteolytic lesion with poorly defined margins, 1 cm in diameter, located in the right pedicle of L2, extending to the posterior part of the vertebral body (Fig. 1 & 2). The lesion extended beyond the osseous cortex on the lateral

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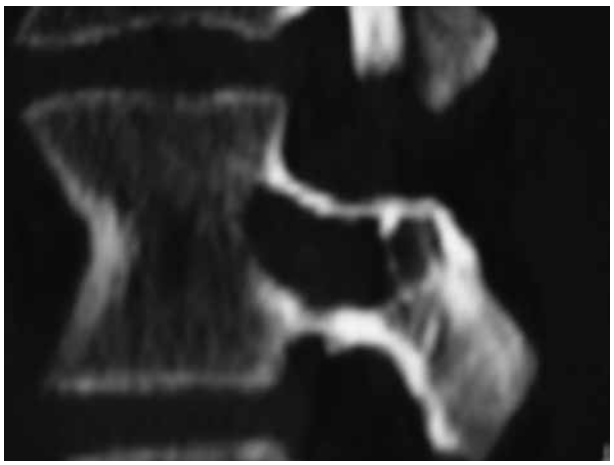

Fig. 1. — Preoperative CT-scan, sagittal view. Osteolytic lesion of the right pedicle of L2.

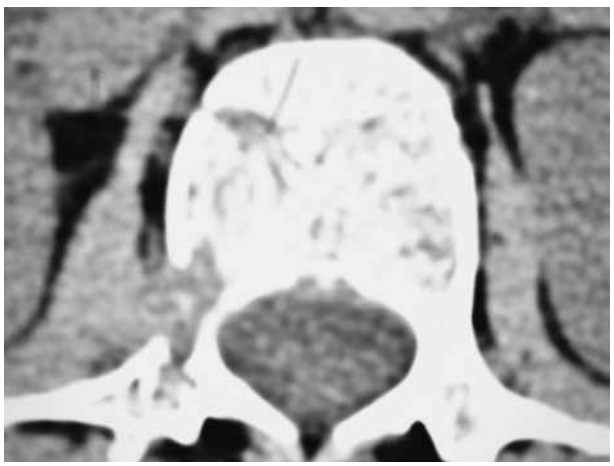

Fig. 2. — Preoperative CT-scan, transverse view. Osteolytic lesion of the right pedicle of L2.

side. A Tc-99 MDP bone scan did not show increased uptake, neither at the site of the lesion nor anywhere else in the skeleton. A CT-scan of head and abdomen did not reveal another space-occupying lesion.

Despite all the imaging studies performed, the precise extent of the lesion could not be determined. Even so, the location and size of the tumour compromised spinal stability. Furthermore, the extra-osseous extension raised concerns of impending neurological involvement, either intra- or extramedullary. It was thus decided to perform an open biopsy of the entire lesion and treat the instability in one stage. The operation therefore consisted of transpedicular removal of the mass and fusion L1L3 with pedicle screws, rods and autologous bone grafts. Histological examination of the specimen showed infiltration of lymphocytes, eosinophils, multinucleated giant cells and Langerhans histiocytes.

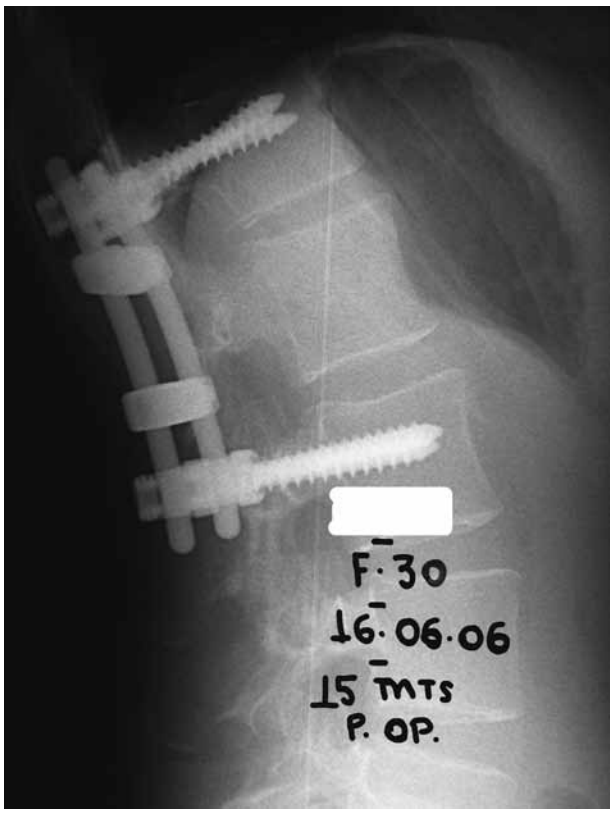

Fig. 3. — Plain radiograph of the lumbar spine, lateral view, 15 months postoperatively. Instrumented arthrodesis L1L3.

Immunohistochemical staining was positive for S-100, CD1a and Langerin proteins, thus authenticating the diagnosis of Langerhans cell histiocytosis. The postoperative course was smooth, and complete pain relief was achieved. No residual disease or recurrence was noted at follow-up more than one year later (Fig. 3).

Table I. — Previously published cases of vertebral LCH in adults

	Age, gender	Site	Neurologic involvement	Treatment	Result
MGH 1954 * (*17,18*)	35M	T11	Yes	S + R	Good
Kaye 1969 (*17,18*)	21F	L2	No	R	Good
Ferris 1974 (*11*)	23M	T6, T7, T8	No	S + R	Good
Casson 1981 (*17,18*)	58F	L5	Yes	S	Good
Sanchez 1984 (*17,18*)	55F	C5, C6	Yes	S + R + C	Good
Herring 1987 (*13*)	20M	C2	No	S + R	Good
Kanterewicz 1988 (*16*)	38F	L1	Yes	S + R + steroids	Good
Padovani 1998 (*17,18*)	20M	T2	Yes	S + R	Good
Dickinson 1991 (*8*)	33F	C2	No	R	Good
Acciarri 1992 (*1*)	44M	T1	Yes	S + R	Good
Villas 1993 (*21*)	20F	T9	No	S	Good
Villas 1993 (*21*)	23F	L2	No	S	Good
Johnson 1993 (*15*)	29M	C2	Yes	S	Not rep.
Cardon 1994 (*7*)	25M	L3	No	R + vertebroplasty	Good
Lauffenburger 1995 (*17*)	35F	C3	No	S + R	Good
Bilge 1995 (*4*)	34F	L5	No	S	Good
Boutsen 1999 (*5*)	36M	T11	No	Conservative	Good
Duarte-Silva 1999 (*9*)	42M	C3, C4, C5	Yes	S + R	Good
Reddy 2000 (*18*)	47M	T11	No	S	Good
Bertram 2002 (*3*)	46M	C2	No	R	Good
Simanski 2004 (*20*)	44M	C4	Yes	S	Good
Garg 2006 (*12*)	25F	L4	No	R	Good

S = surgery, R = radiotherapy, C = chemotherapy.

Islinger *et al* (*14*) report a further 7 cases, but do not mention the characteristics of each individual case.

* MGH = Massachusetts General Hospital. No authors listed : Case Records of the Massachusetts General Hospital. Weekly clinicopathological exercise, case 40342. *N Engl J Med* 1954 ; 251 ; 354-357.

DISCUSSION

Langerhans cell histiocytosis (LCH) is a group of disorders characterized by the proliferation of the similarly named specialized dendritic phagocytes. The disease was formerly known as Histiocytosis X, and its localized osseous form as eosinophilic granuloma. In 1997, the World Health Organization (WHO) Committee on Histiocytic/Reticulum Cell Proliferations presented a classification whereby these older terms were replaced with the collective term "Langerhans cell histiocytosis" ; the term "eosinophilic granuloma" is now defined as single system, solitary site disease (*10*).

Langerhans cells (LC) originate in the bone marrow and travel via the blood stream to the lymph nodes, thymus, lungs and skin. On infection, LC will take up and process microbial antigens and travel to the draining lymph node. There they mature into antigen presenting cells and interact with T-cells, initiating an immune response.

Microscopically, LCH is an aggregate of histiocytes, mature eosinophils and multinucleated giant cells. The most commonly affected organs are bone,

lungs, CNS, liver, thymus, skin and lymph nodes (*2*). In vertebrae, the lesions destroy and replace the normal bone architecture. This may result in partial or even complete collapse of the vertebral body, creating the characteristic vertebra plana (*2*).

A review of the published adult cases demonstrates that the overwhelming majority of patients with vertebral LCH present with pain, localized in the affected area, with or without irradiation (*1,3-5,7-9,11-18,20,21*). Neurologic involvement is variable and not always correlated with the degree of osteolysis or collapse.

The radiologic appearance is that of an osteolytic lesion with either poorly defined or sclerotic margins (*1,3-5,7-9,11,13-18,20,21*). The radiographic sign of vertebra plana, so prominent in pediatric cases, is markedly absent in adults. Only one adult case with vertebra plana has been reported (*12*). CT and MRI findings are non-specific ; however, these modalities help to identify the extent of the disease as well as any soft tissue involvement. Scintigraphy has very low sensitivity (*3*), but can identify small lesions even if radiographs are normal (*2*). The differential diagnosis is that of the solitary osteolytic spinal lesion. It includes metastasis, lymphoma and leukaemic infiltration, Ewing sarcoma, osteoblastoma, aneurysmal bone cyst, intraosseous haemangioma, osteomyelitis, tuberculosis and hyperparathyroidism (brown tumours). The definitive diagnosis can only be made by histological and histochemical examination. Positive immunohistochemical staining for proteins found on Langerhans histiocytes, like S-100 and CD1a, is diagnostic. The management of vertebral Langerhans cell histiocytes is not standardized. The number of published adult cases is too small for safe conclusions (Table I). Solitary osseous LCH is a benign, self-limiting disorder when systemic disease is absent (*6*). It is therefore imperative to confirm the diagnosis and to establish the extent of the disease. A skeletal survey and bone scintigraphy should be employed to this end, as well as to exclude other diagnoses. CT and MRI will reveal an impending neural involvement. Biopsy should always be attempted using the least invasive technique possible. When this is not possible or when spinal stability or neurological function is threatened, surgery is advocated. There is no role for routine radiotherapy, except in cases where the disease continues to progress or when surgical access is particularly demanding (*2,6*). Chemotherapy is reserved for multiple systemic lesions (*6*).

RERERENCES

1. **Acciarri N, Paganini M, Fonda C, Gaist G, Padovani R.** Langerhans cell histiocytosis of the spine causing cord compression : case report. *Neurosurgery* 1992 ; 31 : 965-968.
2. **Azouz EM, Saigal G, Rodriguez MM, Podda A.** Langerhans' cell histiocytosis : pathology, imaging and treatment of skeletal involvement. *Pediatr Radiol* 2005 ; 35 : 103-115.
3. **Bertram C, Madert J, Eggers C.** Eosinophilic granuloma of the cervical spine. *Spine* 2002 ; 27 : 1408-1413.
4. **Bilge T, Barut S, Yaymaci Y, Alatli C.** Solitary eosinophilic granuloma of the lumbar spine in an adult. Case report. *Paraplegia* 1995 ; 33 : 485-487.
5. **Boutsen Y, Esselinckx W, Delos M, Nisolle JF.** Adult onset of multifocal eosinophilic granuloma of bone : a long-term follow-up with evaluation of various treatment options and spontaneous healing. *Clin Rheumatol* 1999 ; 18 : 69-73.
6. **Brown CW, Jarvis JG, Letts M, Carpenter B.** Treatment and outcome of vertebral Langerhans cell histiocytosis at the Children's Hospital of Eastern Ontario. *Can J Surg* 2005 ; 48 : 230-236.
7. **Cardon T, Hachulla E, Flipo RM *et al*.** Percutaneous vertebroplasty with acrylic cement in the treatment of a Langerhans cell vertebral histiocytosis. *Clin Rheumatol* 1994 ; 13 : 518-521.
8. **Dickinson LD, Farhat SM.** Eosinophilic granuloma of the cervical spine. A case report and review of the literature. *Surg Neurol* 1991 ; 35 : 57-63.
9. **Duarte-Silva EB, Noujaim J el-K, Carnevale F.** Cervical spine cord compression by eosinophilic granuloma. Case report. *Arq Neuropsiquiatr* 1999 ; 57 : 498-503.
10. **Favara BE, Feller AC, Pauli M, Jaffe ES, Weiss LM, Arico M *et al*.** Contemporary classification of histiocytic disorders. The WHO Committee On Histiocytic/Reticulum Cell Proliferations. Reclassification Working Group of the Histiocyte Society. *Med Pediatr Oncol* 1997 ; 29 : 157-166.
11. **Ferris RA, Pettrone FA, McKelvie AM, Twigg HL, Chun BK.** Eosinophilic granuloma of the spine : an unusual radiographic presentation. *Clin Orthop Relat Res* 1974 ; 99 : 57-63.
12. **Garg B, Sharma V, Eachempati KK, Malhotra R, Bhan S.** An unusual presentation of eosinophilic granuloma in an adult : a case report. *J Orthop Surg (Hong Kong)* 2006 ; 14 : 81-83.

13. **Herring JA, Johnston CE 2nd.** Lytic lesion of C2. *J Pediatr Orthop* 1987 ; 7 : 481-483.
14. **Islinger RB, Kuklo TR, Owens BD *et al.*** Langerhans' cell histiocytosis in patients older than 21 years. *Clin Orthop Relat Res* 2000 ; 379 : 231-235.
15. **Johnson S, Klostermeier T, Weinstein A.** Case report 768. Eosinophilic granuloma of the cervical spine. *Skeletal Radiol* 1993 ; 22 : 63-65.
16. **Kanterewicz E, Condom E, Canete JD, Del Olmo JA.** Spinal cord compression by a unifocal eosinophilic granuloma : a case report of an adult with unusual roentgenological features. *Neurosurgery* 1988 ; 23 : 666-668.
17. **Lauffenburger MD, Dull ST, Toselli R.** Eosinophilic granuloma of the adult spine : a case report and review of the literature. *J Spinal Disord* 1995 ; 8 : 243-248.
18. **Reddy PK, Vannemreddy PS, Nanda A.** Eosinophilic granuloma of spine in adults : a case report and review of literature. *Spinal Cord* 2000 ; 38 : 766-768.
19. **Scarpinati M, Artico M, Artizzu S.** Spinal cord compression by eosinophilic granuloma of the cervical spine. Case report and review of the literature. *Neurosurg Rev* 1995 ; 18 : 209-212.
20. **Simanski C, Bouillon B, Brockmann M, Tiling T.** The Langerhans' cell histiocytosis (eosinophilic granuloma) of the cervical spine : a rare diagnosis of cervical pain. *Magn Reson Imaging* 2004 ; 22 : 589-594.
21. **Villas C, Martinez-Peric R, Barrios RH, Beguiristain JL.** Eosinophilic granuloma of the spine with and without vertebra plana : long-term follow-up of six cases. *J Spinal Disord* 1993 ; 6 : 260-268.