Eur Spine J
DOI 10.1007/s00586-015-3764-x

CASE REPORT

Vertebroplasty in a 10-year-old boy with Gorham–Stout syndrome

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Received: 5 August 2014 / Revised: 7 January 2015 / Accepted: 9 January 2015

Abstract
Purpose To describe our experience using balloon vertebroplasty with polymethylmethacrylate filler in a 10-year-old boy with Gorham–Stout syndrome.
Summary of background data Gorham–Stout syndrome is a rare disorder of unknown etiology characterized by intraosseous proliferation of fibrovascular or lymphatic tissue that results in progressive osteolysis and bone destruction. It can affect any part of the skeleton and lead to pathological fractures and muscular weakness or atrophy. The literature contains few reports on the surgical management of vertebral osteolysis with risk of fracture in children. Spinal fusion is the treatment of choice.
Methods The patient was asymptomatic until age 10 years, when he began to experience back pain. Annual magnetic resonance scans did not show progression of bone disease. Medical treatment was unsuccessful, and the visual analog scale (VAS) for lumbar back pain was 8–10. Balloon vertebroplasty with polymethylmethacrylate filler was performed at L3 and L4, the vertebrae with an increased risk of pathological fracture.
Results The postoperative course was uneventful. One month after surgery the VAS was 2–3. Four years later, the patient remains free of procedure-related complications, his clinical situation is stable, and no further low back pain has been reported.
Conclusions We report the only application to date of vertebroplasty to treat vertebral osteolysis in a pediatric patient. The outcome and possible complications of this technique remain unknown in children. The patient in the present report underwent vertebroplasty at two levels, and his progress remains satisfactory 4 years after surgery. He has not developed technical complications or changes in spinal growth. Therefore, we propose vertebroplasty for the treatment of vertebral osteolysis in pediatric patients at risk of pathological fracture.

Keywords Vertebroplasty · Gorham–Stout syndrome · Pediatric population

Introduction

Gorham–Stout syndrome is a rare disorder that is characterized by spontaneous massive osteolysis resulting in hemangiomatous or lymphangiomatous tissue proliferation and bone destruction. There is no subsequent bone regeneration. Spinal lesions can be managed with radiation therapy, brace, or halo traction. Surgical stabilization may be required for unstable spinal lesions.

Case report

A 6-year-old boy presented with a lymphatic malformation in his right thigh that extended to the perineum and scrotum. Magnetic resonance imaging (MRI) showed extensive visceral involvement and osteolysis of the vertebral bodies (T1 to L5), iliac bones, ischium, sacrum, and the proximal thirds of both femurs. The excisional biopsy of the thigh mass confirmed the diagnosis of Gorham–Stout syndrome. The annual MRI scans did not show progression of the bone lesions. He gradually developed dysmetria due to excessive growth of the right femur and underwent

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Published online: 14 January 2015

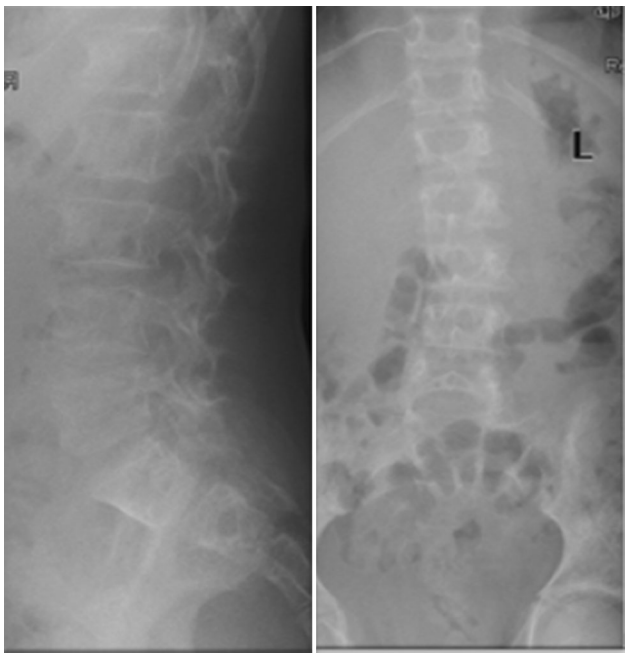

Fig. 1 Plain radiographs showing vertebral osteolysis

temporary epiphysiodesis with eight-plates attached to the distal femur.

The patient was asymptomatic until he was 10 years old, when he began to experience back pain. A plain radiograph of the lumbar spine showed vertebral osteolysis (Fig. 1). Medical treatment consisting of radiation therapy, intravenous zoledronic acid, and interferon alfa-2b was unsuccessful, and the score on the visual analog scale (VAS) for lumbar back pain was 8–10. Balloon vertebroplasty with polymethylmethacrylate filler was performed at L3 and L4, the two vertebrae at increased risk of pathological fracture.

A transpedicular approach was used to introduce a thoracic spine vertebroplasty trocar under x-ray guidance (owing to the size of the patient). Clear liquid similar to lymphatic drainage liquid was observed. The trocar was exchanged for a working cannula to introduce a deflated bone tamp, which was inflated with radio-opaque contrast, thus making it possible to compact the cancellous bone along the margin of the central cavity and reduce the risk of cement leakage. After removing the balloon, we filled the defect with high viscosity cement under low injection pressure.

The postoperative course was uneventful (Fig. 2). One month after surgery the VAS score improved to 2–3. Four years later, the patient remains free of procedure-related complications and low back pain, and his clinical situation is stable. The eight-plates were removed 3 years after their attachment. Final dysmetria was about 1.5 cm. The most recent MRI scan showed no progression of bone disease, only vertebroplasty cement at L3 and L4, with no signs of

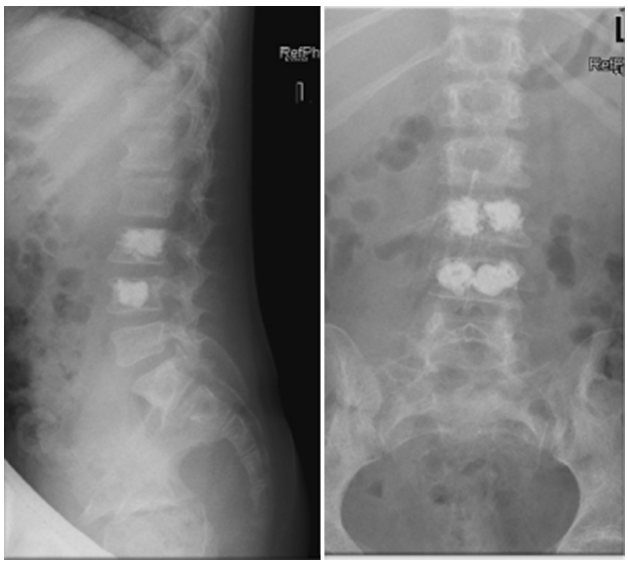

Fig. 2 Immediate postoperative plain radiographs

bone collapse. The morphology and signal of the spinal canal, cauda equina, and thecal sac were normal 4 years after the procedure (Fig. 3a, b). Bone densitometry revealed osteopenia in the femoral neck, with normal values in L1–L2 (80 % increase in mineral density compared to densitometry performed before treatment with bisphosphonates).

Discussion

Gorham–Stout syndrome, also known as disappearing bone disease, phantom bone disease, and vanishing bone disease, was first described by Jackson [1] in 1883, but it was not considered an independent disease until 1955 [2]. Fewer than 200 cases have been reported in the literature. The syndrome can affect men or women of any age, although most cases are diagnosed in childhood and adolescence. No familiar predisposition or genetic transmission has been established. The clinical course is characterized by replacement of normal bone by aggressively expanding fibrovascular or lymphatic tissue without neoplastic features. There is no subsequent osteoblastic regeneration [3]. Plain radiographs show osteolysis or pathological fractures. Computed tomography and MRI are useful for ascertaining disease extension and soft tissue involvement [4].

Medical treatment consists of radiation therapy, chemotherapy, anti-resorptive drugs (bisphosphonates), and hemangiomatosis therapy. Surgical options include resection and reconstruction of the osteolytic lesions and treatment of the fracture. Spinal lesions can be managed with radiation therapy, braces, or halo traction [5]. Surgical stabilization may be required for unstable spinal lesions.

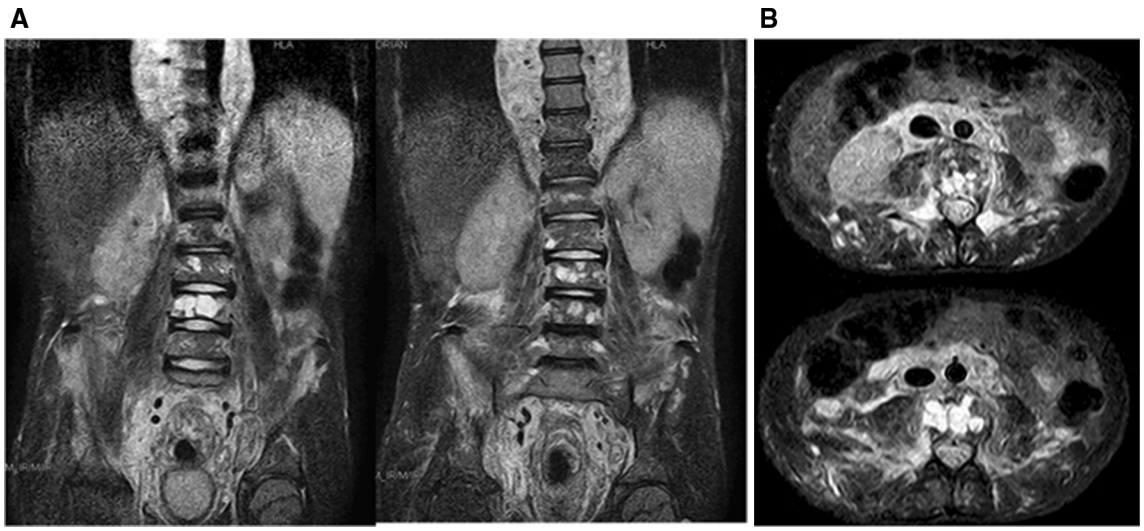

Fig. 3 **a** MRI (coronal view) shows vertebroplasty cement at L3 and L4**. b** MRI (axial view) shows vertebroplasty cement at L3 and L4

Table 1 Review of surgical treatment of vertebral osteolysis in pediatric patients

Author	Patient	Treatment	Results
Espinosa-García et al. [7]	11 years. Thoracic spine osteolysis and spinal cord injury	In situ posterior fusion	Complete motor lesion and incomplete sensory medullary lesion
Aizawa et al. [8]	10 years. Kyphosis >90° due to osteolysis T3–T12	In situ posterior fusion without deformity correction	Complete paraplegia and anesthesia below T6
Drewry et al. [6]	13 years. Osteolysis T10–L1	In situ posterior fusion	Incomplete paraplegia, resolved with anterior fusion with cage and allografts
Livesley et al. [9]	5 years. Osteolysis T6–T10	In situ anterior fusion	Increasing kyphosis, resolved with posterior fusion and halo-jacket (30 months)
Mawk et al. [10]	6 years. Osteolysis C1–C3	Radiation therapy, chemotherapy, and halo traction (3 months)	Remineralization

Few cases of vertebral lesions in children diagnosed with Gorham–Stout syndrome have been reported in the English-language literature [6–10]. Treatment was mainly medical (drugs and/or radiation therapy). Only five patients underwent surgery, because they had intense drug-resistant pain, neurological symptoms, or deformity. The results were generally disappointing (Table 1).

To our knowledge, balloon-vertebroplasty with polymethylmethacrylate filler has not been applied in pediatric patients. Vertebroplasty and kyphoplasty are widely used techniques in the percutaneous treatment of conditions such as vertebral fractures, osteolytic metastases, symptomatic hemangiomas, and multiple myeloma [6]. However, given the low prevalence of these disorders in childhood, no cases of vertebroplasty have been recorded. Therefore, the outcome and possible short- and long-term complications of the technique remain unknown in this population. Anterior vertebral reconstruction with autograft followed by vertebral fusion was ruled out for several reasons: the known consequences of vertebral fusion before skeletal maturity, the aggressiveness of the surgical procedure, and the increased risk of an anterior approach in a patient who had previously undergone abdominal surgery (as in the present case).

Our patient underwent vertebroplasty of two vertebrae at age 10 years, and his progress remains satisfactory 4 years after surgery. He has not developed technical complications or changes in spinal growth.

Conclusions

Gorham–Stout syndrome is a rare entity that can lead to vertebral osteolysis in childhood. When medical treatment is unsuccessful, surgery is necessary. Spinal fusion is the treatment of choice in the literature, although the results are discouraging. We describe a 10-year-old patient who underwent successful balloon vertebroplasty at L3–L4.

This procedure may be considered a viable option for the treatment of vertebral osteolysis in pediatric patients at risk of fracture.

Acknowledgments No funds were received in support of this work.

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

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