Spinal giant cell tumor in children
Report of two cases

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Abstract
The spinal giant-cell tumour is a benign neoplasm with aggressive local behaviour which rarely occurs in the paediatric population. Pain in association with neurologic compromise represents the tumour typical onset in children. Ample resection and the decompression of the nervous elements in association with instrumented fusion result in a stable spine, the preservation or restoration of the nervous function and avoidance of tumour recurrence. We present two patients with spinal giant-cell tumours in immature skeletons that were subject to resection and stabilization by instrumented arthrodesis, with no late postoperative recurrence. These patients' clinical onset, their age and their long-term surgical results give extreme importance to the report of these cases in immature skeletons.

Key words: Giant-cell tumor; spinal; immature skeleton.
Level of Evidence: III

Tumor de células gigantes espinal en niños. Reporte de dos casos

Resumen
El tumor de células gigantes espinal es una neoplasia de estirpe benigna y comportamiento local agresivo, de presentación rara en la población pediátrica. El dolor asociado al compromiso neurológico es la presentación típica en niños. La resección amplia del tumor y la descompresión de los elementos neurales asociada a fusión instrumentada permiten obtener una columna estable, preservar o restaurar la función neural y evitar la recidiva tumoral.
Se presentan dos pacientes con tumor de células gigantes espinal en esqueleto inmaduro, sometidos a resección y estabilización mediante artrodesis instrumentada, sin recidiva en el posoperatorio alejado.
La presentación clínica, la edad de los pacientes y los resultados quirúrgicos a largo plazo hacen de extremo valor el reporte de estos casos en esqueletos inmaduros.

Palabras clave: Tumor de células gigantes; espinal; esqueleto inmaduro.
Nivel de Evidencia: III

Conflict of interests: The authors have reported none.

Introduction

The giant-cell tumour (GCT) is a benign neoplasm characterized by locally aggressive behavior. Pain is the most frequent initial symptom. In spite of its apparently benign histological looks, between 1% and 11% of GCT cause lung metastasis.[1]

Typical age of GCT onset is between the second and the fifth decades of life—less than 10% occurs before 19 years old and, exceptionally, it occurs in immature skeletons.

The most frequent location of the GCT is the vertebral body, and less frequently it affects dorsal elements. One-18% of GCT occurs in the sacral bone. In X-rays what prevails is its osteolytic aspect with great destruction of local bone architecture. The presentation of these tumours is usually isolated, although there are documented reports on multiple presentations.[1-4]

From a histological point of view, the GCT is characterized by the presence of giant multinuclear cells and fusiform cells. The tumor tissue is highly vascularized and, in general, it has no connective tissues.[5]

Definite treatment in the spine consists of ample surgical resection with ample tumour removal. In selected cases, some neoadjuvant therapies based on selective embolization, biphosphonates and monoclonal antibodies have achieved some degree of efficiency.

Postoperative radiation is controversial because it is associated with potential risk of malignization.[6,7]

The aim of this study is to describe clinical onset, diagnostic methods, treatments and long-term results in a rare tumor in the paediatric population, with a >2-year follow-up with no recurrence.

Case 1

Nine-year-old prepuberal girl who consults for >30 day-history of incapacitating low back pain, with no traumatic history and progress to paraplegia with preserved sensitivity in lower limbs, no sphincter control, and 48-hour ASIA B history. No history of diseases or treatments.

In initial X-rays of lumbosacral spine, we detect a pathologic fracture which involves upper and lower vertebral end plates, body collapse, and bulge on the wall of the third lumbar vertebral body (Figure 1).

MRI shows an L3 collapse image which is hypointense in T1 and hyperintense in T2, with >75% invasion of spinal canal and preservation of adjacent intervertebral discs (Figure 2), 6-7C in the Weinstein, Boriani and Biagini classification and 2 G0 T0 M0 in Enneking classification.

Within 48 hours of neurologic symptoms onset, we carry our surgery for tumor resection and spine stabilization by left lumbotomy. For reconstruction we use structural fibular autograft between L2 and L4 in association with double-bar Kaneda-like instrumentation between L2 and L4. We do not detect intraoperative complications. We

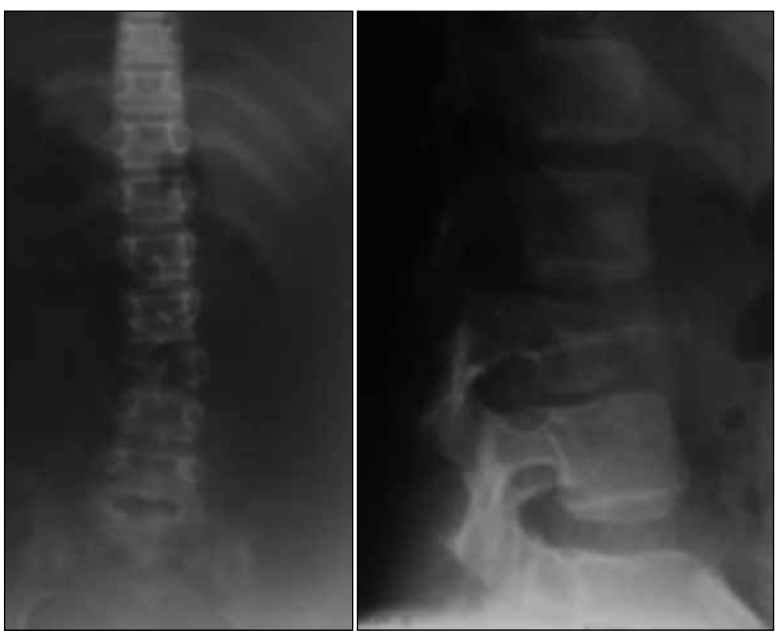

Figure 1. Lumbosacral spine X-rays. Crush fractures of the third lumbar vertebral body, with compromise of the dorsal wall.

send tumour resection material (intraoperative biopsy) for histopathology study, whose result is GCT.

At immediate follow-up, the patient's neurological status progresses to ASIA E. We have complete L2-L4 fusion four months after the surgery. Nowadays the patient has neither symptoms nor signs of tumor recurrence or metastasis (Figure 3).

Fourteen years after the surgery the patient continues asymptomatic, she has no evidence of tumor diseases and carries a normal lease of life with no limitations in daily

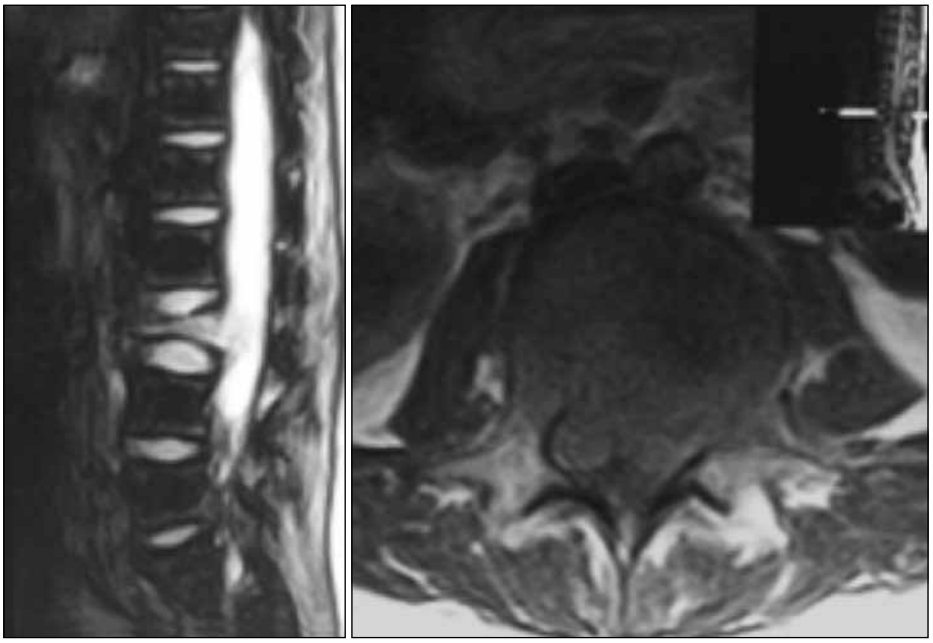

Figure 2. MRI. Crash fracture with compromise of the 75% of the spinal canal and preservation of adjacent intervertebral discs.

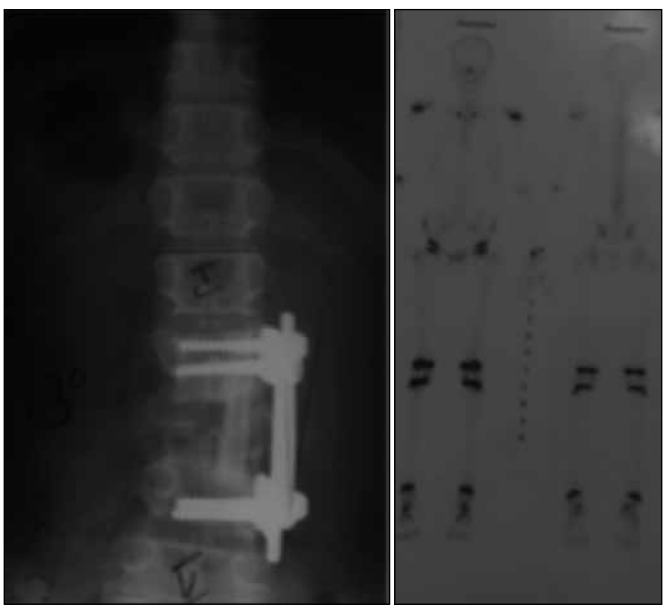

Figure 3. A. After the surgery we can see fusion of the L2-L4 arthrodesis. **B.** Scintigraphy does not show recurrence of the disease.

activities or sports (Figure 4).

Case 2

Three-year-old male who consults for 7-day history of progressive paraparesis; he shows ASIA C, preserved sphincters, patellar and achillean hyperreflexia, and associated dorsolumbar pain.

At the ER he is evaluated by doctors from the Neurosurgery Department who prescribe spine X-ray, which shows normal radiological parameters (Figure 5), and MRI,

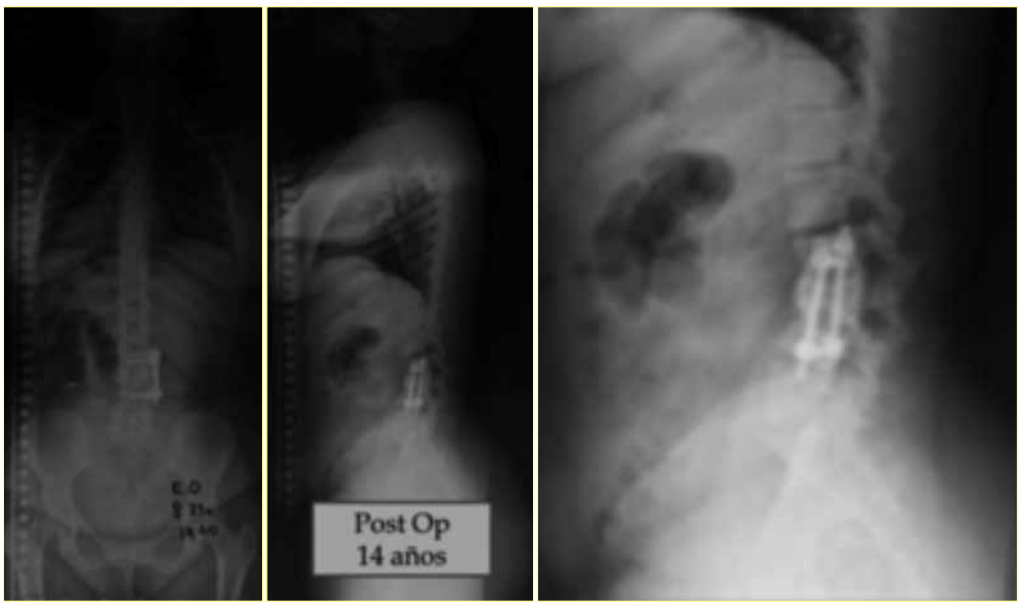

Figure 4. X-rays 14 years after the surgery. Complete fusion, residual local kyphosis.

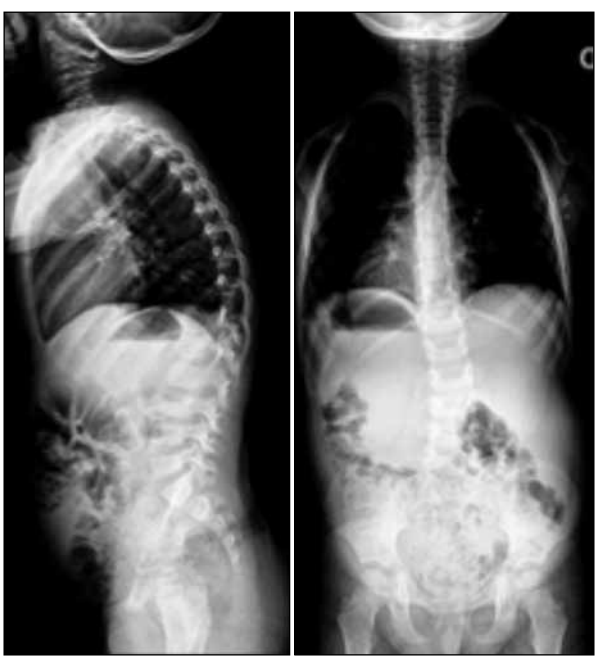

Figure 5. X-rays showing normal radiologic parameters.

which shows and extradural, intracanal tumour mass that spreads to T7 and T9 involving the dorsal arch and the medial column of the T7 body. In view of such progressive neurological condition, we carry out decompression by laminectomy from T6 to T9, and partial tumour resection. This material is sent to the Histopathology Department, which informs GCT. The patient's neurological status improves to ASIA E two weeks after the surgery.

One month after the surgery, with no complications, the patient is asymptomatic with no neurological findings and diagnosis of partial resection of GCT.

In the patient's consultation with the Spinal Surgery Department, he is reassessed with CT scan, MRI and gamma camera. We find remaining tumour in the medial column of the T7 vertebral body, with no metastatic injuries (Figures 6 and 7).

He is scheduled a new surgery two months after the first one. We carry out the resection of the dorsal third of the T7 vertebral body by posterolateral approach by left costotransversectomy and extracelomic approach. We

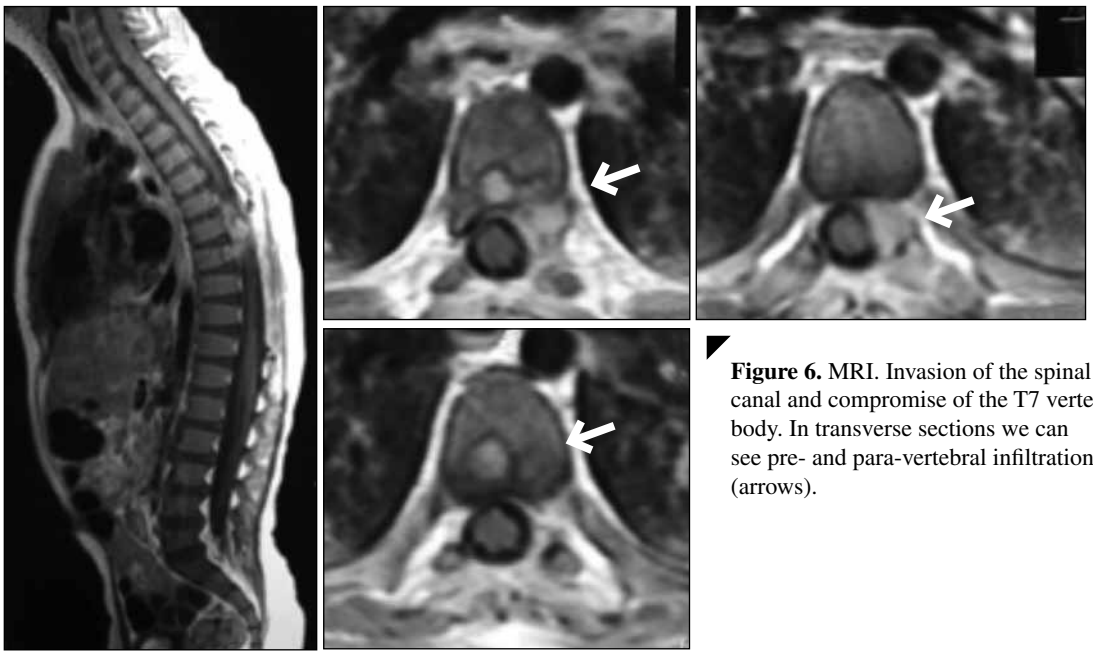

Figure 6. MRI. Invasion of the spinal canal and compromise of the T7 vertebral body. In transverse sections we can see pre- and para-vertebral infiltration (arrows).

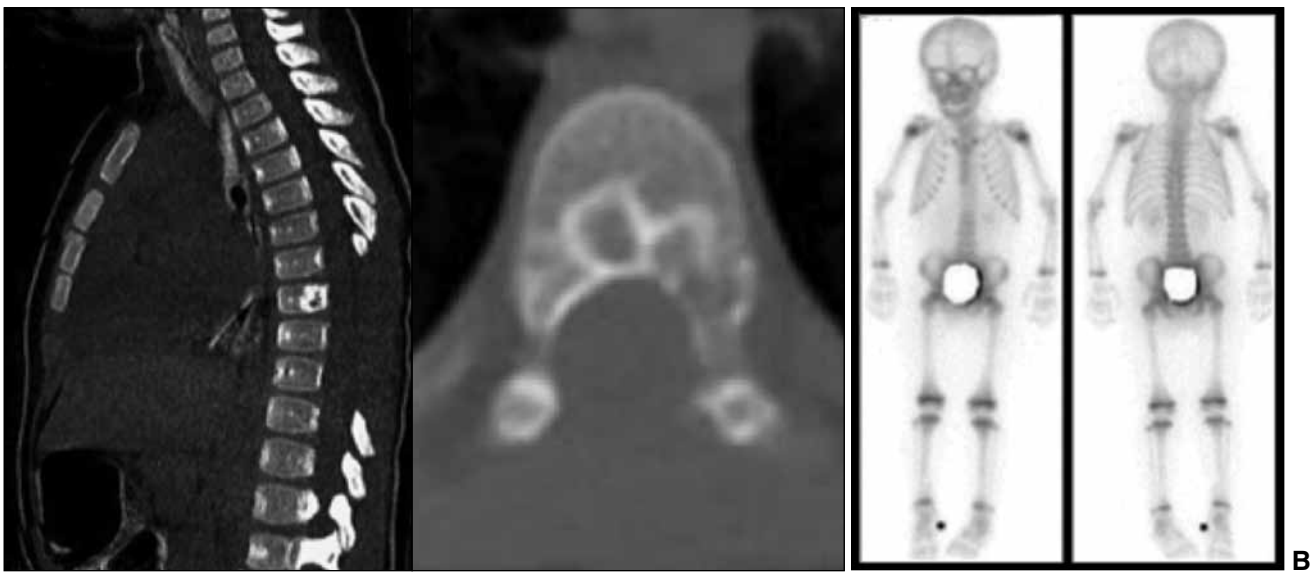

Figure 7. A. Sagittal and transverse CT scan sections. Osteolytic images with sclerotic edges and ample laminectomy. **B**. Gamma camera with no evidence of disease spread.

verify intracanal tumour tissue which spreads from T7 to T9, T7: 6C-T8: E6-T9:6D in the Weinstein, Boriani and Biagini classification, and 2 G0 T0 M0 in the Enneking classification. After ample tumour resection, we carry out posterior instrumented arthrodesis from T5 to T12 and intervertebral filling with bank allograft in the defect left by the body tumour resection. There are no intraoperative complications. The patient uses TLSO corset brace during four months. The patient does not undergo either early or late postoperative complications.

Two years after undergoing decompression surgery and fused dorsal instrumented arthrodesis, the patient shows neither symptoms nor X-ray evidence of tumor recurrence or metastasis (Figure 8).

The Histopathology Department confirms GCT with the second sample.

Discussion

The GCT is a scarcely frequent bone neoplasm; 7% of them are detected in the spine and it is more frequently located in the sacral bone. It is rare before the third decade of life. We present these two cases that we operated on just because GCT is exceptional in patients with immature skeleton.

Likewise patients with closed cartilages the referential pattern for the treatment of GCT is the "block" resection of the tumour; sometimes, however, due to anatomic location, this is impossible and we suggest carrying out ample radical resection.

Schütte and Taconis documented a series of 462 patients with GCT, out of who 49 were <19 years old, and only 9 out of them were located in the vertebral column—one cervical tumour, one thoracic tumour, two lumbar tumours and five sacral bone tumors.[8]

Fidler reported nine adult patients with GCT treated by block resection with good results.[9]

Junming et al. published a series of 22 cases showing better tumour control rates with block resection. They report local recurrence rates of 28%.[10]

It is estimated that the risk of lung metastasis increases six-fold after the first local recurrence.[11]

Miszczyk reported a series of GCT in adults treated with surgery plus radiotherapy vs. surgery in isolation. Local control rates were 83% when they used surgery plus radiation as adjuvant and 69% when they gave patients surgery in isolation. However, differences were not statistically significant.[12]

As of late, it has been shown that biphosphonates such as pamidronate and zoledronic acid reduce tumour local recurrence and are useful as efficient adjuvant therapy.[13]

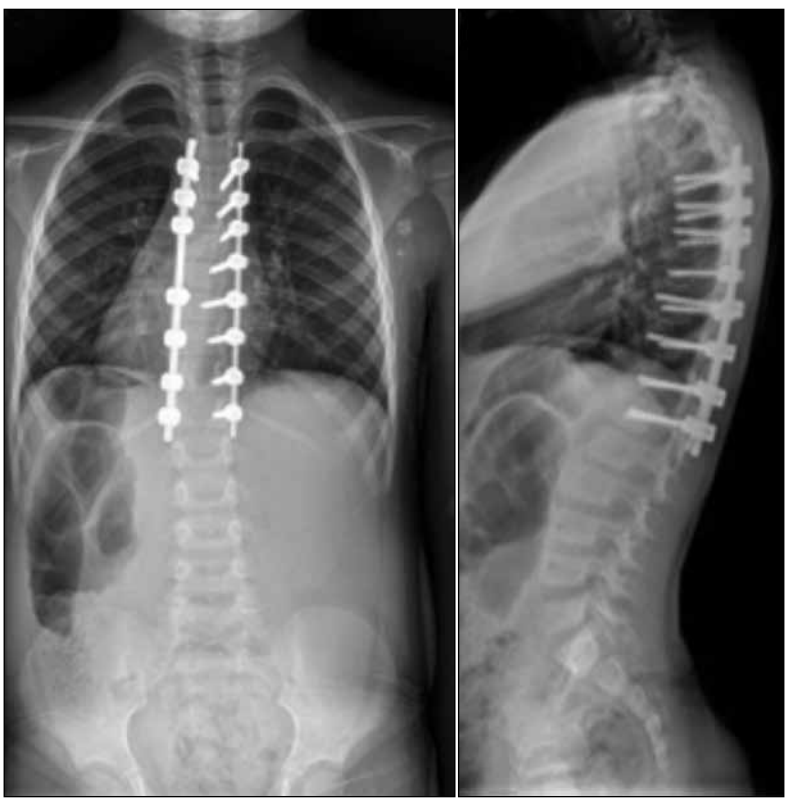

Figure 8. X-ray one year after surgery. T5-T12 dorsal instrumented arthrodesis.

Gille et al.[14] reported the case of a 46-year-old male with recoil of a cervical spine GCT after treatment with zoledronic acid by itself. Diverse authors recommend avoiding radiotherapy to reduce the risk of sarcoma induced by radiation.[15,16] Ma et al.[17], on the contrary, affirm that radiotherapy may represent a valid alternative for the treatment of non-removable tumours with no risk of malignization.

The monoclonal antibody denosumab against the RANK ligand in the fusiform cells of the GCT has reduced the size of the tumour and induced bone formation and, consequently, it has improved preoperative neurologic symptoms caused by tumour compression, and it has even facilitated surgical procedures.[18] Treatment with denosumab in adults with GCT reduced significantly or eliminated proliferating tumour contents replacing them with bone tissue.[19]

As far as we know, there are no reports in current bibliography on the use of denosumab in children with GCT, just reports on isolated cases treated with denosumab for aneurismal bone cysts. [20,21]

Conclusions

The GCT is a benign neoplasm with aggressive local behaviour which rarely occurs in the paediatric population. Pain in association with neurologic compromise represents the tumour typical onset in children. Ample tumour resection and the decompression of the nervous elements in association with instrumented fusion result in a stable spine, the preservation or restoration of the nervous function and avoidance of tumour recurrence.

These patients' clinical onset, their age and their long-term surgical results give extreme importance to the report of these cases in immature skeletons.

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