Eosinophilic Granuloma of the Spine With and Without Vertebra Plana: Long-term Follow-up of Six Cases (Cast Reports)

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SUMMARY

Vertebral eosinophilic granuloma is a rare condition frequently associated with vertebra plana. In this paper we present six patients with eosinophilic granuloma of the spine; three were without vertebra plana, which represents a diagnostic problem. The mean follow-up was 9 years, (range 2 to 23) and the mean age was 10.8 years at diagnosis. All complained of pain with no neurological deficit. The lesions were located on the vertebral bodies of C4, T9, T10, L1, L2, and L5, respectively. Histologic confirmation of diagnosis was obtained in all patients, two by puncture and four by open biopsy. The patients with vertebra plana (T10, L1, and L5, respectively) were treated conservatively. Long-term follow-up demonstrated total healing of the vertebral body in two and partial rebuilding 8 years after diagnosis in one. Patients without vertebra plana (C4, T9, and L2, respectively) underwent curettage and bone grafting. In the patient with T9 location, a T8-10 anterior arthrodesis with autogenous rib graft was performed. The outcome was satisfactory in all.

KEY WORDS

Granuloma, eosinophilic; Disease, spinal; Histiocytosis X.

INTRODUCTION

Eosinophilic granuloma accounts for ≤75% of all cases of histiocytosis X and affects the skeleton in a monostotic or polyostotic fashion (1,5,7,9,10,13,17, 20). Any bone can be involved, but >70% of the lesions occur in the skull, pelvis, ribs, mandible, femur, and humerus (7,23).

Schajowicz (22) reported that the incidente of vertebral eosinophilic granuloma representa 7.8% of all eosinophilic granulomas of bone. The thoracic vertebrae are involved most often (54%), followed by lumbar (35%) and cervical (11%) vertebrae (7).

Most orthopaedic surgeons have been trained to think of vertebral eosinophilic granuloma of the spine as a disease presenting vertebra plana, but little attention has been given to other radiographic appearances (15). We present six patients affected by vertebral eosinophilic granuloma, three without vertebra plana, with a mean follow-up of 9 years (range 2 to 23) after diagnosis.

CASE REPORTS

Case 1

A 3-year-old boy was referred to our department in January 1969 with a l-month history of gait disturbance and a weakness of the lower extremities. Physical examination demonstrated lumbar pain and no neurological deficit. Lumbar spine radiographs revealed a vertebra plana of L1 (Fig. 1A). The routine blood analysis was normal. A percutaneous biopsy, under radiographic guidance, comfirmed the presumptive diagnosis of eosinophilic granuloma. The patient was treated with a plaster corset for 3 months. Subsequent radiographs demonstrated progressive increase of the vertebral body height (Fig. I B and C). Twenty-three years later, the patient is asymptomatic with a complete reconstruction of the vertebral height

Case 2

A 20-year-old woman with a clinical picture of 6 months of dorsal pain consulted the department in April 1981. Physical examination demonstrated a tenderness to percussion of lower dorsal vertebrae and absence of neurological deficit The sedimentation tate was 60 mm/h. Other blood tests were normal. Radiographs and CT scan (Fig. 2A and B) showed a lytic lesion of the anterior portion of the T9 vertebral body without vertebral collapse. Technetium bone scintigraphy was normal. The preoperative diagnosis was aneurysmal bone cyst. An anterior approach biopsy (Fig. 2C) and curettage of the lesion were done, and an T8-T10 arthrodesis was performed using autogenous rib graft. Postoperatively, a plaster cast was indicated for 4 months. Ten years later there was a complete bone fusion at the arthrodesis level without functional deficit (Fig. 2D).

Case 3

A 5-year-old girl presented at our department in April 1981 with lumbar pain after a bicycle trauma. There was no neurological deficit. A radiograph of the lumbar spine showed a vertebra plana of L5 (Fig. 3A). Sedimentation cate was 65 mm/h; myelography, electromyography, and technetium bone scintigraphy were normal. An anterior approach biopsy confirmed the diagnosis of eosinophilic granuloma. The patient was treated with a plaster corset for 3 months. A year later a lytic lesion appeared in the parietal bone, resulting in the same histological diagnosis. Blood analysis detected diminution in T-lymphocyte activity. Radiotherapy (2000 cGy) was indicated to control the skull lesion and intramuscular injections of thymic extracts were indicated for the treatment of the lymphocyte abnormality. Long-term follow-up of the vertebral lesion (10 years) demonstrated a total reconstitution of the vertebral height without any sequelae (Fig. 3B and C).

Case 4

An 8-year-old boy came to the emergency service with a painful torticollis of 2 months evolution. No numbness, weakness, or paresthesias of the extremities were noted. On

physical examination, the patient presented a slight limitation in neck motion. No sensory or motor deficits were detected. Results of laboratory blood tests and chest radiographs were unremarkable. Plain radiographs, tomograms (Fig. 4A), and CT scan showed a lytic lesion involving the posterior portion of the C4 vertebral body. Technetium bone scintigraphy was normal. An anterior approach biopsy (Fig. 4C) and curettage of the lesion were performed. A tricortical iliac bone graft was placed in the defect, preserving the vertebral end platel. A Minerva jacket was indicated for 3 months. Two years later there was a reconstruction of the vertebral body (Fig. 4B), and the patient was asymptomatic.

Case 5

A 6-year-old boy with dorsal pain of 1 year evolution was referred to the department in July 1983. Physical examination denoted pain in the lower dorsal vertebrae on spinous percussion. No neurological deficit was detected. Plain radiographs showed a T10 vertebra plana, without kyphosis (Fig. 5A), and technetium bone scintigraphy demonstrated an increased uptake in a lower thoracic vertebra. The blood tests were normal except for a low number of supressor lymphocytes. Punction biopsy was performed. The pathologic diagnosis was eosinophilic granuloma of bone. It was treated with thymic extracts and radiotherapy in another center. After 8 years, a 40% vertebral reconstitution of the vertebral height was noted (Fig. 5B).

Case 6

A 23-year-old woman with a lumbar pain of 3 months evolution consulted our service during September 1989. Physical examination showed no neurological deficit Plain radiographs were normal, CT scan showed a lytic lesion involving the posterior portion of the L2 vertebral body (Fig. 6B), and MRI showed a high signal intensity arca located in the posterior—inferior comer of the L2 vertebral body (Fig. 6A). Technetium bone scintigraphy was normal. By a posterior approach biopsy (Fig. 6C), curettage and bone grafting of the lesion were performed. Two years later the patient remained asymptomatic, and a complete healing of the vertebral body was observed.

DISCUSSION

The term “vertebra plana” corresponds to the condition described by Calvé in 1925 as a vertebral osteochondritis (3). Several histopathological studies have shown that eosinophilic granuloma is the commonest causative lesion of vertebra plana (6,14,16). The collapse of the vertebral body is often considered pathognomonic of eosinophilic granuloma in young patients (2,4,6,14). Support for this conclusion is the involvement of only 1 vertebra, preservation of the disc spaces, and consistent radiographic density of the collapsed body. Some authors (11,15,24) had reportad eosinophilic granuloma of the spine without platyspondylia, but most of these cases had extrabody location of the lesions. In our series, three patients with vertebral body involvement (C4, T9, and T10, respectively) did not show vertebral collapse. This is not often seen and causes a diagnostic problem.

Eosinophilic granuloma with vertebral body involvement can progress to vertebra plana depending on the extension of the lesion and its location in the spine. As we had witnessed in our series, extensive vertebral body lesions in high weight-bearing areas of

the spine progress to platyspondylia. Neither in our series nor in the literature did we find any relationship between patient age and the development of vertebra plana.

Neurologic impairment secondary to spinal eosinophilic granuloma has been infrequently reported in the literatura (8,10-12,17-19,24,25,27). This complication can be dueto several mechanisms: Yabsley and Harris (27) believe it is caused by vertebral collapse itself. Green et al. (11) report that extradural spread of the lesion may also be an important factor. Recently, Mafulli et al. (19) reported a C5 eosinophilic granuloma with a secondary C5-C6 segmentary instability that caused spinal cord compression.

Technetium bone scintigraphy was done in five of six patients. Only one patient with a T10 vertebra plana demonstrated an increased uptake. Technetium scintigraphy as a screening method seems to have poor sensitivity. In comparison, we believe that a lytic radiographic image without vertebra plana with normal Tc scintigraphy orienta to the diagnosis of a benign lesion. Nevertheless, in these cases, biopsy must be performed.

Treatment is still controversial. Eosinophilic granuloma is a self-limited disease, and a simple biopsy and curettage alone are frequently sufficient stimuli to induce the lesion to regress. The goals of the treatment are spinal stability and preservation of neurological function. The treatment of a typical vertebra plana without neurological deficit must be conservative (4,14). In the three patients of our series with platyspondylia, we observed gradual increase of the vertebral height. In two of them, rebuilding was almost total (L1 and L5, respectively); and in the other (T10), the reconstitution of the height was almost 40%. This patient had not reached skeletal maturity at the time of the last control. None of these these patients had pain nor vertebral deformity at the long-term follow-up.

In patients with detected immunologic alterations, such as the presence of lymphocytes spontaneously cytotoxic or T-lymphocyte deficiency, thymic extract treatment can reverse these abnormalities as demonstrated by Osband et al. (21).

Radiation therapy alone, in dosages ranging from 500 to 1500 cGy, has frequently been employed in the treatment of solitary lesions or to control disseminated ones (9,11,22,24-27). No postradiation sarcomas have been described in the literature (26). Although we believe Chis treatment in most cases is not necessary, we agree with Yabsley and Harris (27) that radiation therapy can only be justified in the treatment of difficult surgical access lesions associated with neurological damage.

In our opinion, surgical treatment can be indicated when the lesion compromises spinal stability with secondary neurological impairment and when open biopsy must be performed, especially in those cases with atypical radiologic presentation.

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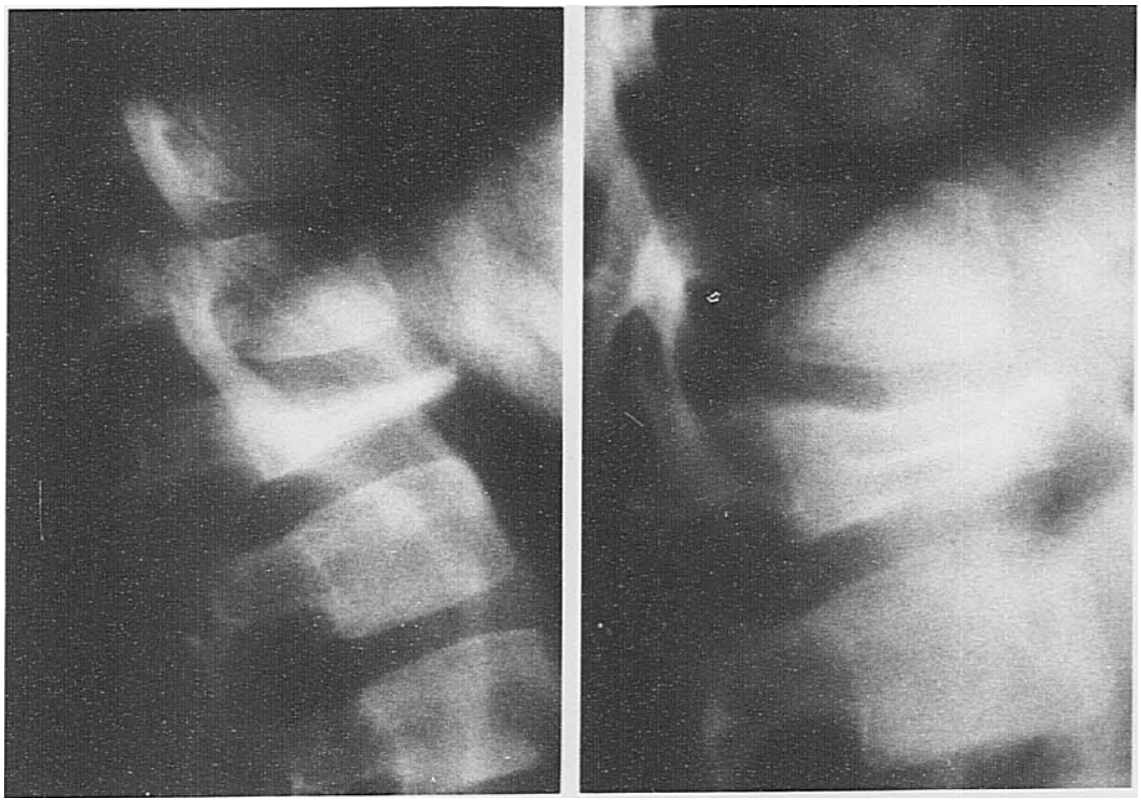


Figure A **Figure B**

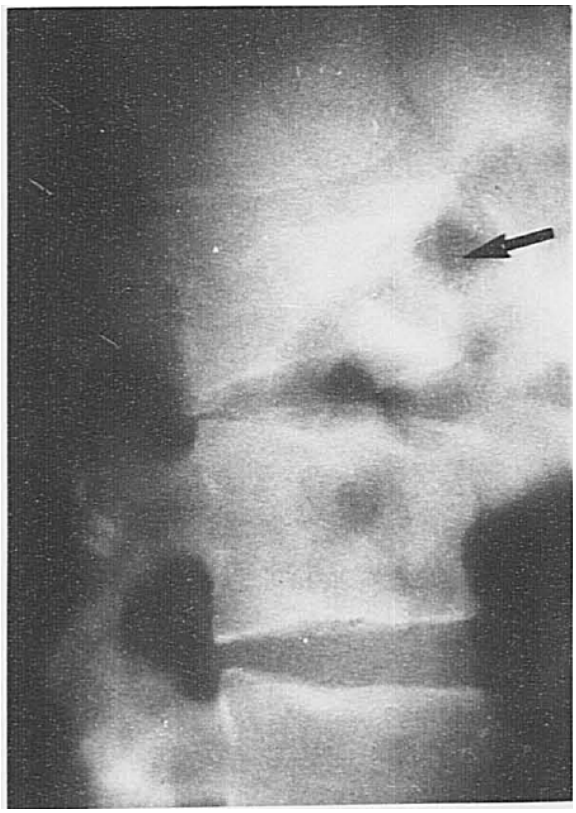
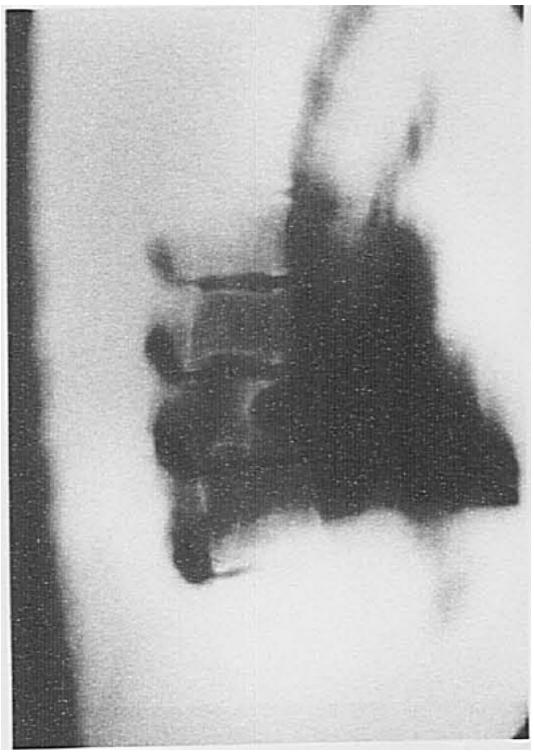


Figure C

Figure 1. A: Collapse of L1 with loss of the vertebral height in a 3-year-old boy. **B:** Five years alter diagnosis showed pardal restorage of the vertebral height. **C:** Twenty-three years later the collapsed vertebra (arrow) had almost recovered its height.



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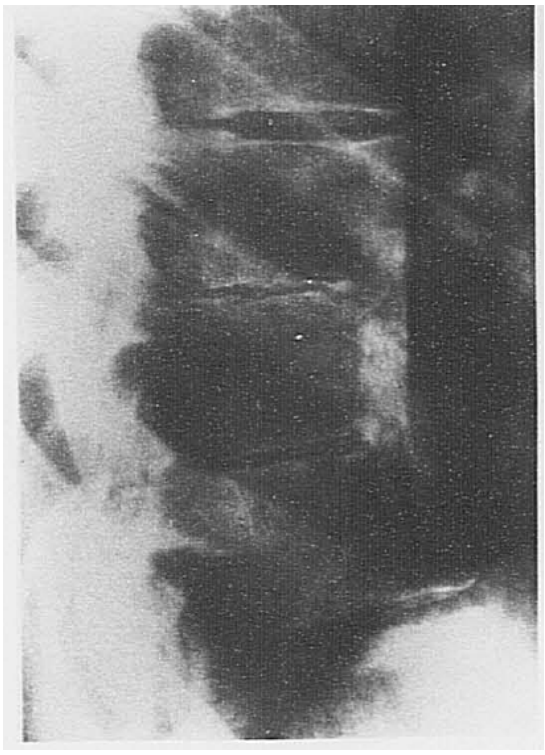

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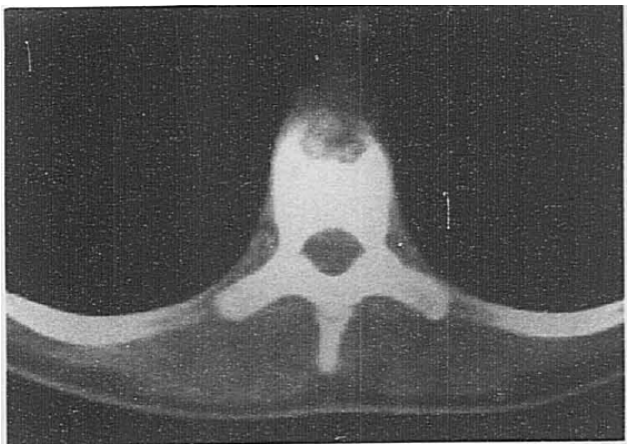

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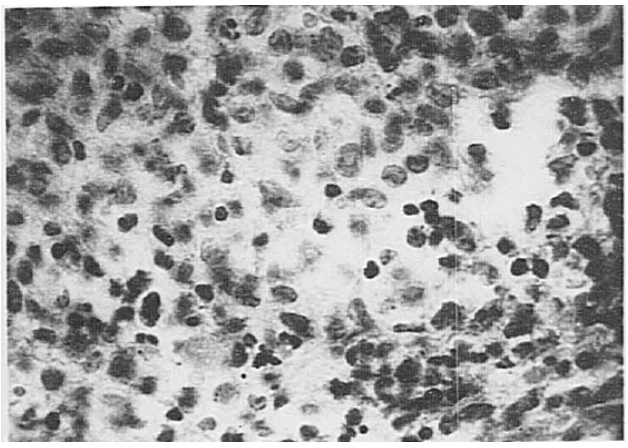
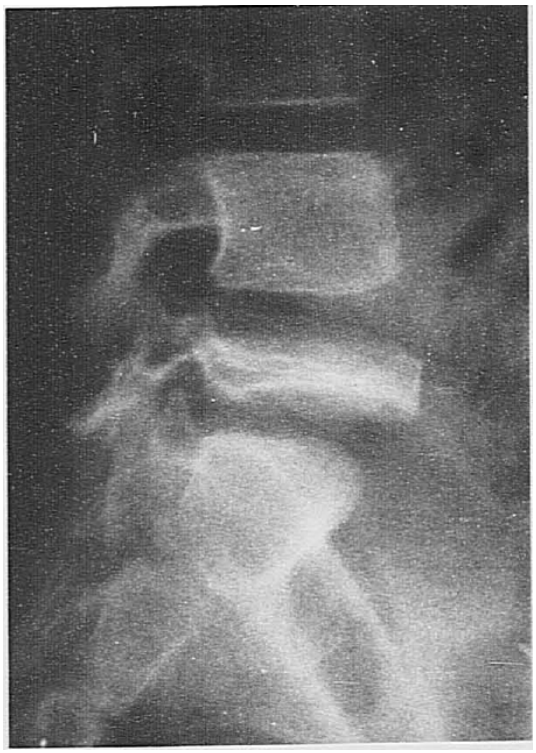


Figure C

Figure 2. A Lateral radiograph of the dorsal spine showing anterior osteolysis of T9 vertebral body. **B:** CT scan through T9 vertebral body showing anterior destruction and extension to soft tissue. **C:** Histologic confirmation of the diagnosis. **D:** Radiograph obtained 10 years alter surgery showing incorporation of the autogenous rib graft.



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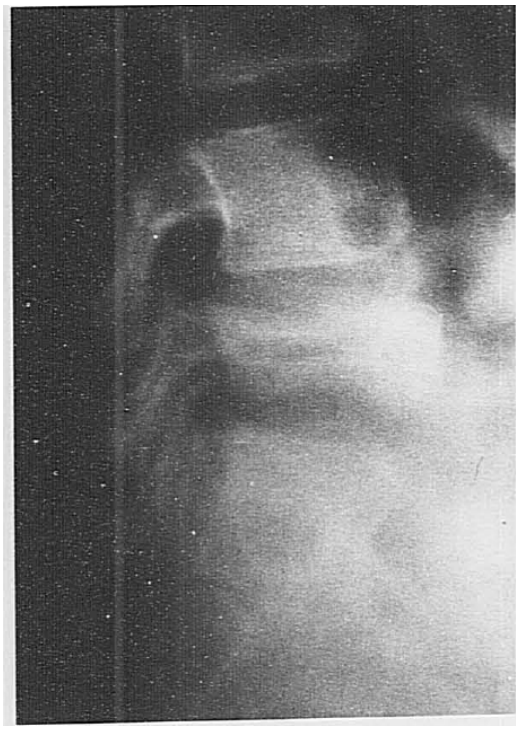

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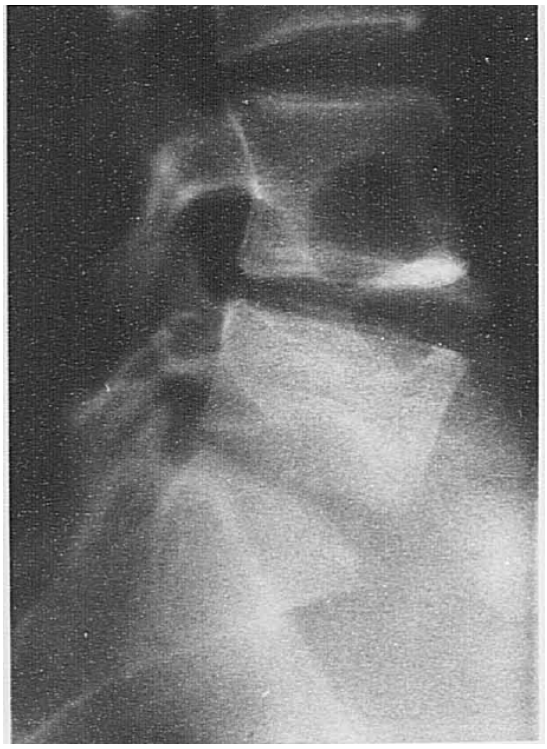


Figure C

Figure 3. A: Lateral radiograph of a 5-year-old girl showing vertebra plana of L5. **B:** Five years after diagnosis partid reconstruction of L5 vertebral body is shown. **C:** Ten years later there was reconstitution of the vertebral height.

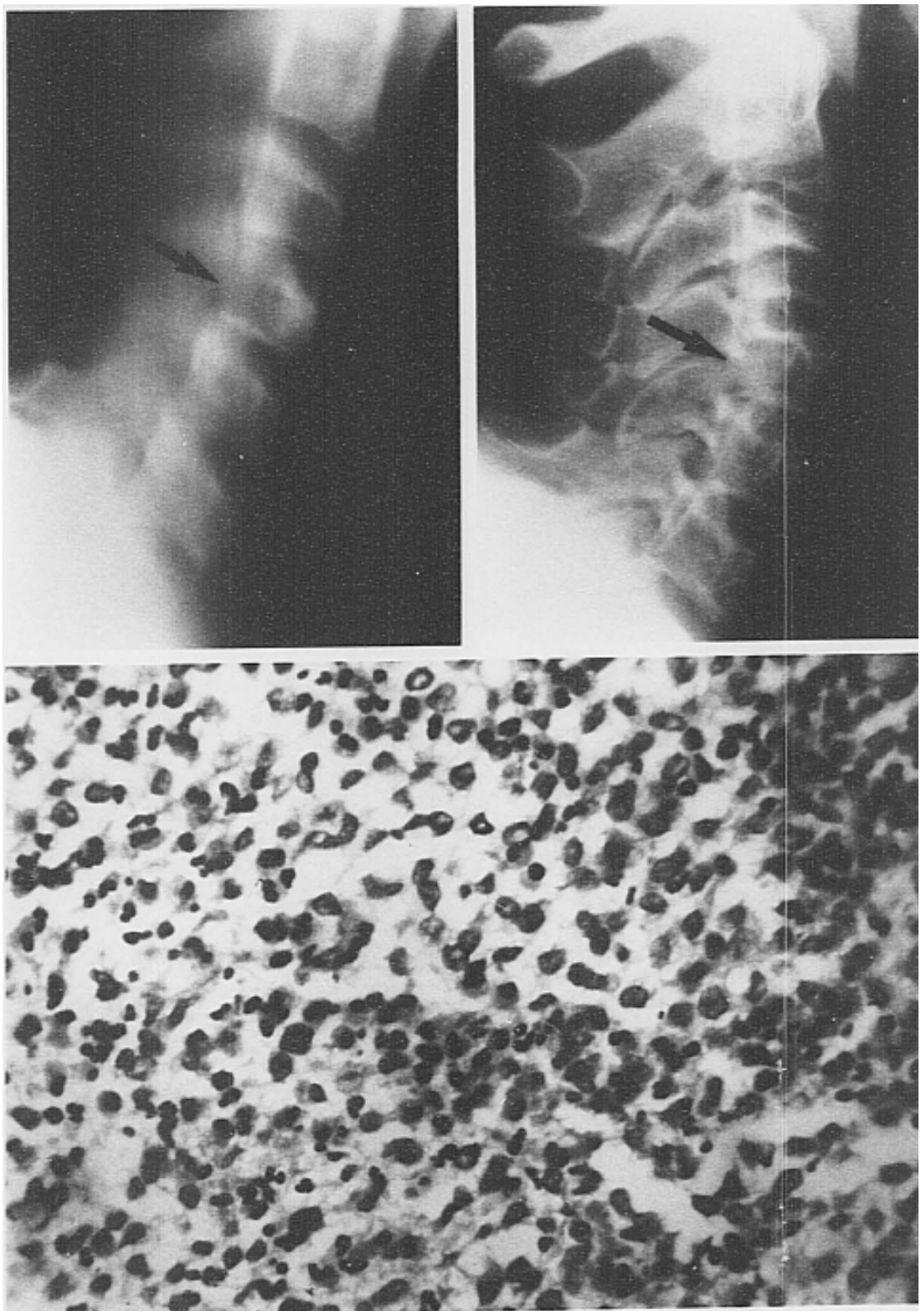


Figure 4. A: Lateral tomograms showing a lytic lesion in the body of C4. **B:** One year after surgery there was incorporation of the iliac tricortical bone graft at the defect (arrow). **C:** Ilistologic confirmation of the diagnosis.

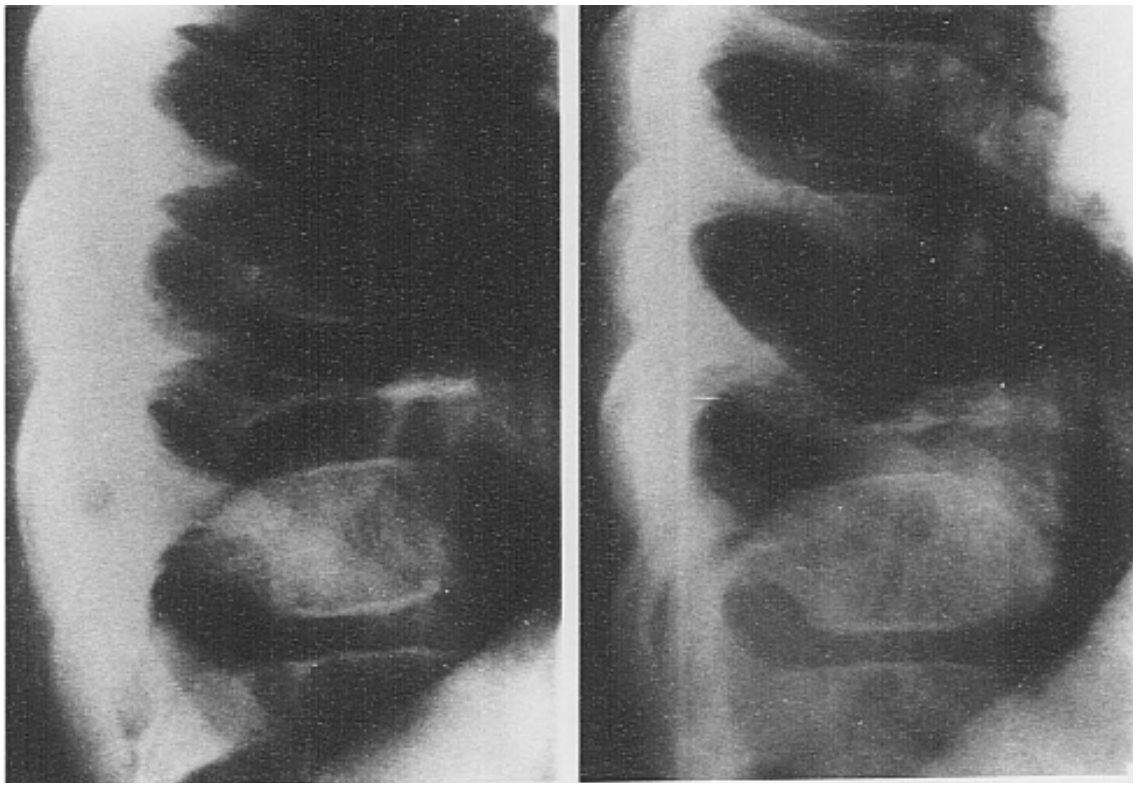
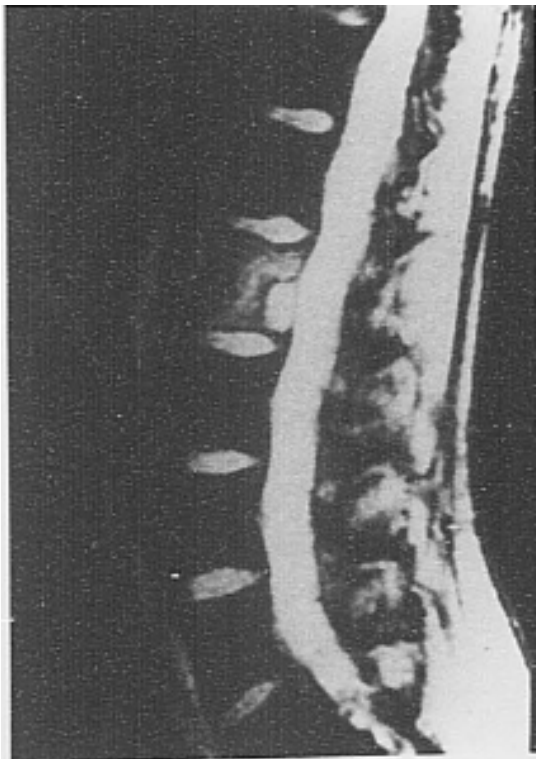


Figure 5. A: Lateral radiograph at diagnosis of a 6-year-old boy with a T10 vertebra plana. **B:** Eght years later, the disc spaces were of normal thickness, and the collapsed vertebra had recovered 40% of its height.



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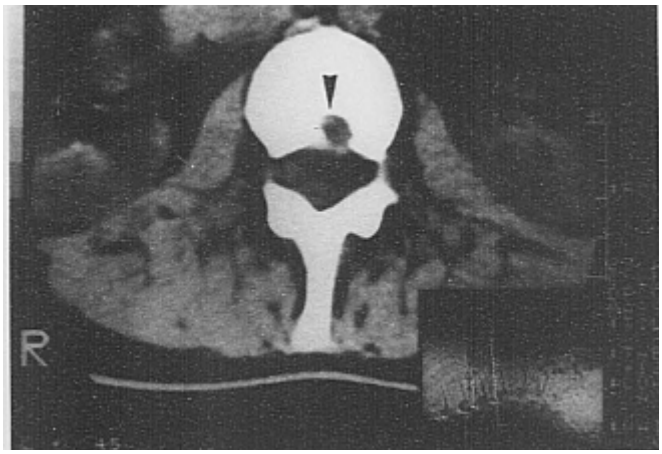

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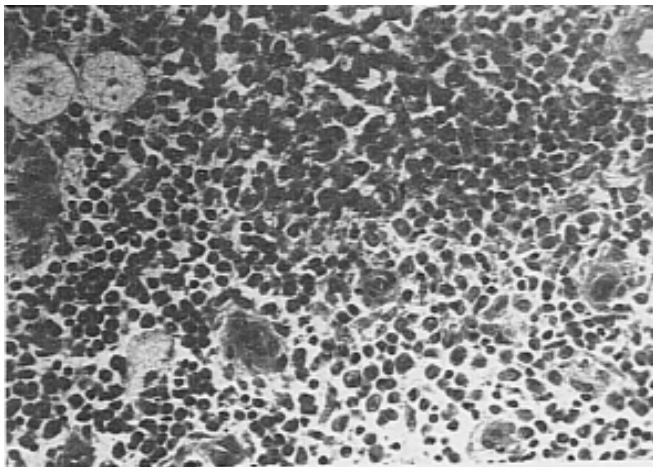

Figure C

Figure 6. A: An MR image weighted in T2 with myelographic effect showing a high signal intensity area surrounded by an intermediate signal area located in the posteroinferior corner of the L2 vertebral body corresponding to an eosinophilic granutoma with secondary edema. **B:** A CT scan at L2 level showing a lytic lesion (arrow) in the posterior area of the vertebral body. **C:** Histologic confirmation of the diagnosis.