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Case Report

Gorham Disease of the Spine: A Case Report and Treatment Strategies for This Enigmatic Bone Disease

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AIZAWA, T., SATO, T. and KOKUBUN, S. *Gorham Disease of the Spine: A Case Report and Treatment Strategies for This Enigmatic Bone Disease*. Tohoku J. Exp. Med., 2005, **205** (2), 187-196 —— Gorham disease is an extremely rare condition of unknown etiology characterized by progressive osteolysis. Only 28 cases of its spinal involvement have been reported, and some of those cases showed kyphosis, kyphoscoliosis, subluxation or dislocation. No definite regimen of treatment has been established yet. A 10-year-old boy presented with a severe and progressive kyphosis over 90 degrees caused by Gorham disease from T3 to T12. In situ posterior fusion with a hook and rod system and iliac bone grafts were performed, but after surgery, he had complete paraplegia and its cause was uncertain. Based on the unfortunate consequence of the present case and the review of the literature, we propose the treatment strategies for spinal Gorham disease. ———— Gorham disease; massive osteolysis; spine; surgical treatment; kyphosis

Gorham disease is an extremely rare and mysterious bone disease of unknown etiology, which is histopathologically characterized by a proliferation of thin-walled vascular channels in bone (Vinée et al. 1994). Since first described by Jackson (1838), it was called by various names: vanishing bone disease, acute absorption of bone, phantom bone, primary lymphangioma, progressive osteolysis and disappearing bone (Gorham and Stout 1955). Gorham and Stout tabulated 24 previously reported cases in 1955 and the condition became known as Gorham disease thereafter. Heffez et al. (1983) suggested the following diagnostic criteria: 1) a positive biopsy for angiomatous tissue, 2) absence of cellular atypia, 3) minimal or no osteoblastic response and absence of dystrophic calcification, 4) evidence of local progressive osseous resorption, 5) non-expansile, non-ulcerative lesion, 6) absence of visceral involvement, 7) osteolytic radiographic pattern, and 8) negative hereditary metabolic, neoplastic, immunologic, or infectious etiology. Any bone can be affected (Chung et al. 1997). However, only 28 cases of spinal involvement, to our knowledge, have been reported in English literature. The progressive osteolysis causes kyphosis, kyphoscoliosis, subluxation or even dislocation of the spine (Castleman and McNeely 1964; Ellis and Adams 1971; Heyden et al. 1977; Woodward et al. 1981; Bohlman et al. 1986; Joseph and Bartal 1987; Drewry et al. 1994; Aoki et al. 1996; Livesley et al. 1996; Chung et al. 1997; Bode-Lensniewska et

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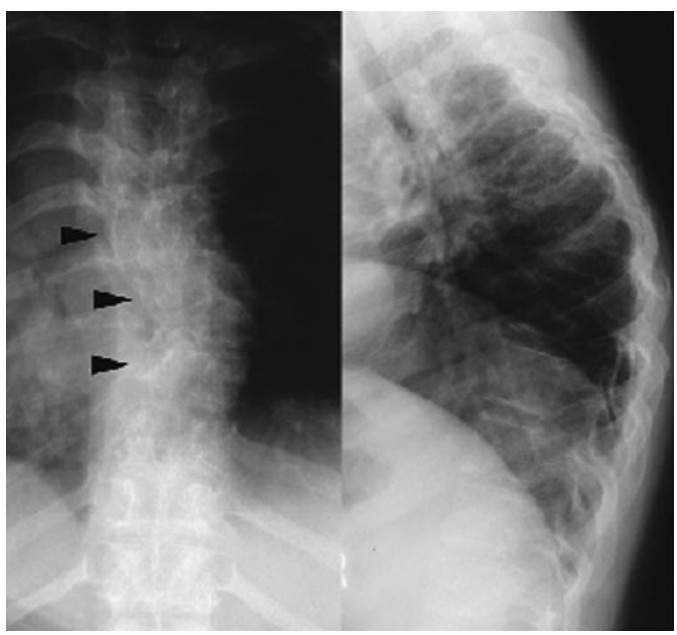
al. 2002; Chong and Shell 2003; Ceroni et al. 2004). No answer has been given to the question how to treat the spinal Gorham disease with progressive deformity.

We report here a case of a patient with Gorham disease in the thoracic spine having a progressive kyphosis over 90 degrees. In situ posterior fixation with a rod and hook system was performed, but after surgery, he had complete paraplegia. This tragedy raises a question what should be performed for this enigmatic bone disease in the spine.

Case Report

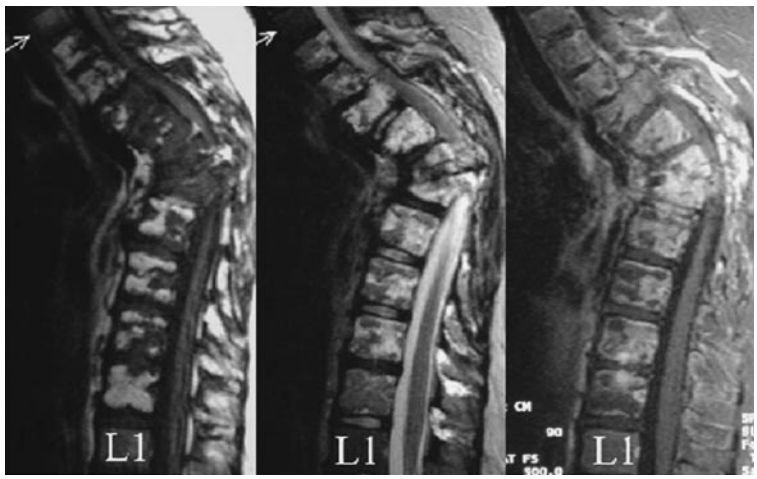
A 10-year-old boy came to our hospital because of a back pain and a deformity in May, 2000. On plain radiographs, his thoracic spine showed a 55-degree kyphosis from T4 to T10 (Fig. 1). Neurological examination revealed slight muscle weakness of the left great toe. Knee and ankle jerks were bilaterally exaggerated. The T3-T12 vertebrae had lesions showing high-signal intensities mixed with low-signal intensities on T1-weighted magnetic resonance (MR) images (T1WI) and heterogeneously high intensities on T2-weighted images (T2WI). The lesions were unevenly enhanced by gadolinium-diethylenetriamine-pentaacetic acid (Gd-DTPA) (Figs. 2a-c). From those MR images, a possible diagnosis of his condition was Gorham disease of the spine, but bone tumors or infectious diseases were not completely ruled out. His back pain decreased for several days without treatment.

The patient had a high fever on July 11, 2000. The white blood cell count was 10,500/μl and C- reactive protein (CRP) was 19.0 mg/μl. A pleural effusion was demonstrated on the right. The fever decreased gradually and CRP became negative on July 24. But the next day, he had gait disturbance because of increased spasticity of both legs, which spontaneously improved in the following weeks. As a definite diagnosis could not be made, open biopsy was performed on August 8, 2000. The specimen showed irregular ectatic vascular formation among bone trabeculae and osteoblastic rimming in reactive bone forma-



(a) (b)

Fig. 1. Plain radiographs on admission.
a) Anterior-posterior (A-P) view showing destructive changes in the mid-thoracic spine and right convex scoliosis (arrowhwads).
b) Lateral view showing osteolytic changes and 55-degree kyphosis in the mid-thoracic spine.



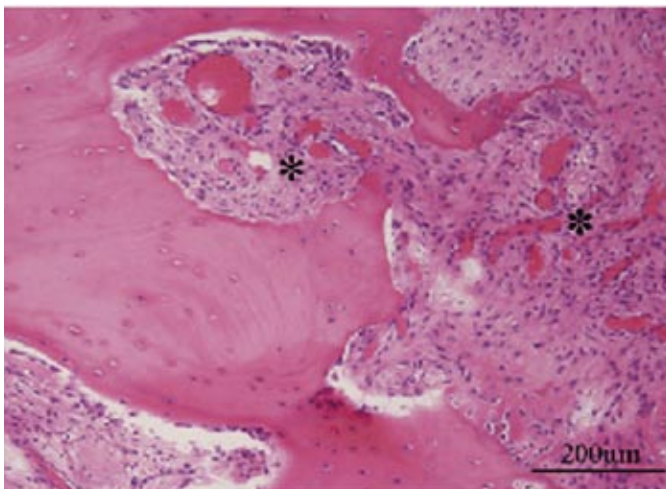
(a) (b) (c)

Fig. 2. MR images on admission.
a) T1WI showing high-signal intensities mixed with low intensities in the vertebral bodies and spinous processes form T3 to T12.
b) T2WI showing heterogeneous high-signal intensities from T3 to T12.
c) The affected vertebrae are well enhanced by Gd-DTPA.

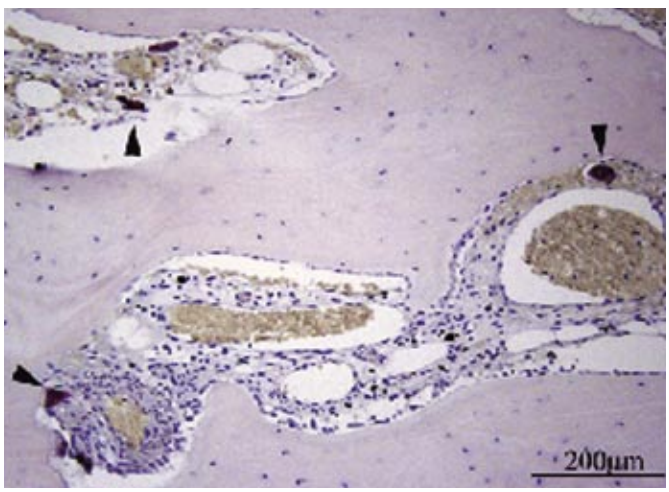
tion (Fig. 3a). Osteoclasts were also detected, but their number did not show much increase, as shown by tartrate-resistant acid phosphate (TRAP) staining (Fig. 3b). Biopsy material was negative for malignancy and bacterial or fungal growth. Histologically, his condition was diagnosed as Gorham disease. Treatment with bisphosphonate was started on September 4, 2000 and gait improved gradually although slight spasticity of both legs continued.

The kyphosis increased gradually and it measured 85 degrees on supine position and 100 degrees on standing position on October 19, 2001 (Figs. 4a and 4b). Three-dimensional computed tomograms (3D-CT) showed destruction of the vertebrae from T3 to T12 and several ribs (Fig. 4c). In situ posterior fusion was performed on December 4, 2001 with expectation of preventing the deformity from further progression and the thoracic myelopathy from worsening (Masini and Maranhão 1997; Sato et al. 1997; McMaster and Singh 1999). Before operation, the patient was placed awake prone on a frame for ten minutes, which caused no power loss or worsening of the spasticity in the legs. At operation, the laminae from T3 to T12 had many erosions, ulcers and holes. In some of them, only thin inner cortical tables had remained. Claw hooks and rods were applied between T1 and L2. No spinal correction was added. Finally, sliver grafts from an iliac crest were laid on the laminae (Figs. 5a and 5b).

After recovering from general anesthesia, the patient was found completely paraplegic and anesthetic on and below T6 dermatomes. The hook and rod system was immediately removed and MR images were taken. On T2WI, the spinal cord had a wide high-signal intensity lesion around the apex of the kyphosis, which indicated intramedullary edema of the spinal cord (Fig. 6). High-dose methylpredonisolone was injected according to the protocol of the National Acute Spinal Cord Injury Study (Bracken et al. 1990). Anterior decompression from T6 to T8 was performed through a left cost-transversectomy approach with removal of the 7th and 8th rib heads the next day (Wilkinson 1955). The removed hook and rod system was reset. Neither bony fusion nor osteosclerosis was detected on plain radiographs and CT scans, and no neurological improvement was detected 3 years after operation though the spine remained stable.



(a)

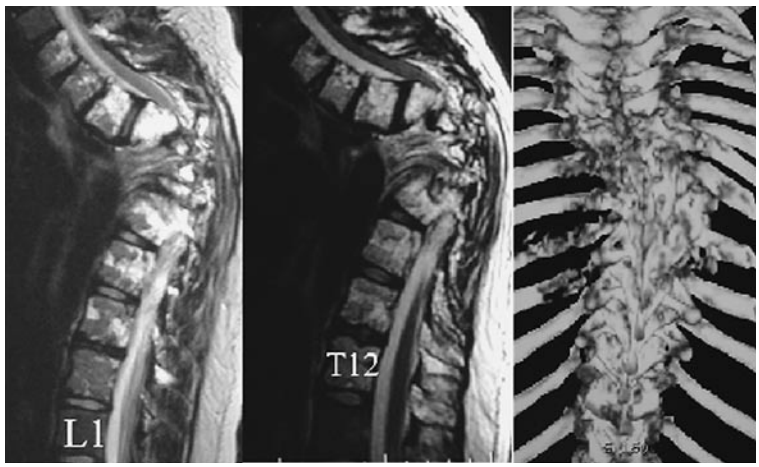


(b)

Fig. 3. Histology of the specimen.
a) Hematoxylin and eosin staining. Irregular, abnormal vessel formation is detected among bone trabeculae (asterisks). The bar is 200 μm.
b) TRAP staining. The number of osteoclasts, which shows positive staining, is not large (arrowheads). The bar is 200 μm.

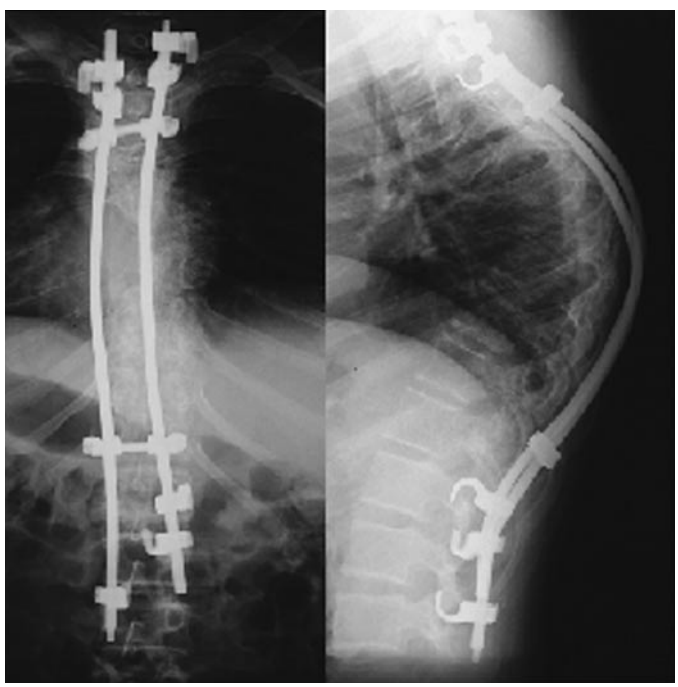
Discussion

Gorham disease of the spine is extremely rare. Only 28 cases, to our knowledge, have been reported in the English literature (Table 1) (Bickel and Broders 1947; Gorham and Stout 1955; Falkmer and Tilling 1956; Hambach et al. 1958; Castleman and McNeely 1964; Ellis and Adams 1971; Thompson and Schurman 1974; Heyden et al. 1977; Woodward et al. 1981; Heffez et al. 1983; Brown et al. 1986; Bohlman et al. 1986; Hejgaard and Olsen 1987; Joseph and Bartal 1987; Mitchell et al. 1993; Drewry et al. 1994; Halliday et al. 1994; Stöve and Reichelt 1995; Aoki et al. 1996; Livesley et al. 1996; Chung et al. 1997; Hagberg et al. 1997; Mawk et al. 1997; Benhalima et al. 2001; Bode-Lesniewska et al. 2002; Chong and Shell 2003; Ceroni et al. 2004). Eighteen patients were males and 10 patients were females. The age made diagnosis ranged from 5



(a) (b) (c)

Fig. 4. Follow-up radiographs before surgery.
a) T2WI on February 2, 2001. Kyphosis from T4 to T10 measures 78 degrees.
b) T2WI on October 19, 2001. The kyphotic deformity has progressed to 85 degrees.
c) 3D-CT. Laminae and several ribs show osteolytic or destructive changes.



(a) (b)

Fig. 5. Plain radiographs after in situ fusion using a claw hook and rod system. (a) A-P view, (b) Lateral view.

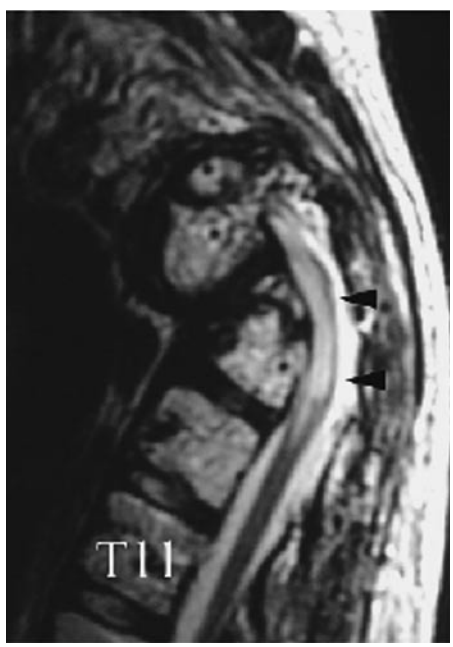


Fig. 6. T2WI immediately after operation. The spinal cord has a wide T2 high lesion, which might indicate an intramedullary edema or swelling (arrowheads).

TABLE 1. *Review of Cases of Spinal Gorham Disease*

First author	Age	Sex	Affected vertebrae	Neurologic deficits	Treatments for Spine	Pulmonary effusion	Results
Bickel, W.H. (1947)	5	F	L4-Sacrum	None	Biopsy	No discription	Condition arrested for 10 yrs
Gohram, L.W. (1955)	16	M	Lower cervical-Upper thoracic	None	Biopsy	(+)	Died after 2 and 1/2 yrs
Falkmer, S. (1956)	55	M	L3-5	None	Removal	No discription	No changes after 12 months
Hambach, R. (1958)	16	F	C3-4	None	None	(+)	Died after 14 months
Castleman, B. (1964)	24	M	C4-6 (C5 subluxation)	Myelopathy	Posterior fusion with iliac bone → destruction of grafted bone → halo-collar fixation for 1 month → posterior fusion with tibial bone	None	Excellent fusion
Halliday, D.R. (1964)	23	M	Sacrum	None	Removal + radiation	None	No discription
	27	M	Thoracic spine	Myelopathy	Radiation + Bradford frame	(+)	Died after 7 yrs
Ellis, D.J. (1971)	72	F	Cervical	None	None	(+)	Death after one month
Thompson, J.S. (1974)	48	M	L5-S1	None	Biopsy	No discription	No discription
Heyden, G. (1977)	34	M	C7-T3	Myelopathy	Radiation + Milwaukee brace	None	New bone formation Fused the involved vertebrae after 3 and 1/2 yrs
Woodward, H.R. (1981)	69	F	C2-4 (C2 dislocation)	Myelopathy	Anterior fusion with iliac bone → absorption of grafted bone → posterior fusion with iliac bone and wire → halo traction for 3 months	None	Complete collapse of C2-C6 Died after 11 yrs by a cerebral vascular accident
Heffez, L. (1983)	13	M	C1, C4-C6	None	Radiation	No discription	Recalcification after 6 months
Bohlman, B.H. (1986)	8	M	C2-4 (C3 subluxation)	Quadriparesis	Biopsy + radiation	(+)	Died after 8 yrs
Brown, L.R. (1986)	30	M	T4-7	None	Removal	(+)	Died 2wks postoperatively
Hejgaard, N. (1987)	9	M	Sacrum	No discription	None	(+)	Mild progression of osteolysis
Joseph, J. (1987)	7	F	T9-10	None	Increasing kyphoscoliosis → radiation → paraplegia	(+)	Died after one and 1/2 yrs
Mitchell, C.S. (1993)	17	F	T4	No discription	Dexamethasone → radiation	No discription	No discription

Drewry, G.R. (1994)	13	M	T10-L1	None	Biopsy → back brace → radiation → posterior fusion with instrument → incomplete paraplegiae → anterior fusion with cage and allografts	(+)	Neurologic status improved Spine stable after 22 months
Stove, J. (1995)	49	F	Sacrum	None	Radiation + calcitonin → etidronacid and clodron acid	No discription	Subjective improvement only
Aoki, M. (1996)	19	F	T9-L3	Paraplegia	Corpectomy + Anterior spinal fusion → halo-pelvic apparatus for 8 months → posterior fusion with instrument	(+)	Free from pain and paresis for 2 yrs
Livesley, P.J. (1996)	5	M	T6-10	None	Anterior fusion → increasing kyphosis → posterior fusion + halo jacket + chemotherapy	No discription	Absorption of grafted bone and adjacent vertebrae Remained halo jacket for 30 months
Chung, C. (1997)	48	F	C5-T3	Radiculopathy	Radiation	(+)	Progression of osteolysis
Hagberg, H. (1997)	19	M	C7-T2	None	Stabilization surgery on three occasions → alfa-2b interferon + clodronate	(+)	No signs of progressive disease
Mawk, J.R. (1997)	6	M	C1-3	None	Radiation + halo traction for 3 months	(+)	Remineralization of affected vertebrae
Benhalima, H. (2001)	17	M	C1	No discription	Biopsy → radiation	No discription	No discription on C1 lesion
Bode-Lesniewska, B. (2002)	65	F	C4-T2	Monoplegia	Sugery was interrupted (impossible to stabilize) → radiation	(+)	Died after 15 months
Chong, L. (2003)	49	M	C2-5 (C3 dislocation)	Quadriparesis	Open reduction + inernal fixation with lateral mass plates → futher anterior dislocation → Anterior fusion with iliac bone → continued osteolysis → posterior fusion with instrument → rod broken → vertebral cage + replacement of the rods	None	Full neurologic recovery No signs of progressive disease
Ceroni, D. (2004)	8	M	C5-T4	Paraplegia	None	(+)	No discription

to 72 years and 15 of them were younger than 20 years. Though it arises at any level from the cervical spine to the sacrum, the thoracic spine was most frequently involved amounting 13 cases. Eight patients died with pulmonary effusion considered as chylothorax. Ten patients had neuropathy: eight myelopathy, one radiculopathy and one monoplegia of the left upper extremity.

The mechanism by which bone absorption occurs in Gorham disease remains unclear. Heyden et al. (1977) proposed a theory that perivascular cells might be osteoclast precursors and associated with bone absorption as they demonstrated strong acid phosphatase and leucine aminopeptidase activities. In TRAP staining of the present case, the number of osteoclasts was not large. The affected bones, therefore, might not be simply absorbed only by osteoclasts but by other cells, too. Or other mechanisms such as mechanical pressure caused by increased vascular tissues might act on this bone absorption process (Joseph and Bartal 1987).

Radiotherapy, medication and surgical treatment were chosen alone or in combination for Gorham disease of the spine but most of them never led to satisfactory results. Some authors stated that radiotherapy could not cease the progression of osteolysis (Cannon 1986; Stöve and Reichelt 1995; Livesley et al. 1996; Chung et al. 1997; Bode-Lesniewska et al. 2002), and others reported favorable outcomes of radiotherapy alone, or in combination with a brace or halo traction, which not only prevented further progression of the disease but also lead to new bone formation in involved vertebrae (Heyden et al. 1977; Heffez et al. 1983; Mawk et al. 1997). In the present case, radiotherapy was not chosen because its effect was controversial and there were too many vertebrae involved.

Medication has been performed for Gorham disease in long bones and spines in combination with some of the following drugs: vitamin D, parathyroid hormone, androgen, calcium, adrenal extracts, vitamin B12 and bisphosphonates (Bullough 1971). Their effects, however, were uncertain. Liversley et al. (1996) used parmidronate, calcitonin and calcium carbonate after spinal fusion. As no evidence of bony healing or no obvious progression of the disease was detected, the patient had remained in a halo jacket for 30 months. Clodronate and α-2b interferon prescribed after operation with radiotherapy ceased a progressive spinal deformity and decreased a pleural chylous effusion (Hagberg et al. 1997). In the present case, bisphosphonate was used expecting prevention of the spine from further resorption (Hagberg et al. 1997). However, its effect could not be confirmed radiographycally. The spinal deformity did not progress although neither bony fusion nor osteosclerosis was detected on plain radiographs and CT scans.

The difficulty in choosing treatments arises from the possibility of spontaneous arrest in Gorham disease reported in some cases (Campbell et al. 1975; Woodward et al. 1981). For this reason, several authors described that conservative treatment should be recommended first or surgical treatment should be performed as late as possible (Bullough 1971; Hejgaard and Olsen 1987). In the present case, the surgical treatment was selected when the kyphosis had increased to 100 degrees and caused myelopathy.

Surgical treatment has been chosen for spinal Gorham disease in 8 cases. Their results were discouraging (Castleman and McNeely 1964; Woodward et al. 1981; Drewry et al. 1994; Aoki et al. 1996; Livesley et al. 1996; Hagberg et al. 1997; Bode-Lesniewska et al. 2002; Chong and Shell 2003). Every spine was not fused by the initial operation and the grafted bones were absorbed. After multiple operations, 5 cases could obtain stable spine (Castleman and McNeely 1964; Drewry et al. 1994; Aoki et al. 1996; Hagberg et al. 1997; Chong et al. 2003). In affected bones in the extremities, amputation or massive prosthetic replacement can be chosen to eradicate their destructive processes (Cannon et al. 1986; Joseph and Bartal 1987; Livesley et al. 1996). Therefore, total spondylectomy was recommended if the number of involved vertebrae is small (Tomita et al. 1997). In the present case, however, it was not considered, because 10 vertebrae were affected.

In situ posterior fusion with a hook and rod

system was a treatment of choice in the present case. Less invasive surgery supposed to be better as it was uncertain whether spinal fusion would be completed. Therefore, we avoided major surgeries such as corrective osteotomy. Unfortunately, the patient had complete paraplegia though no neurologic changes were observed in the prone position test before surgery. The mechanism of this tragedy was uncertain. The spines cranial and caudal to the vertebrae forming kyphos might be over-stretched under general anesthesia, which increased the spinal cord compression against the internal kyphos through the tethering effect.

Conclusion

Even today, there is no reasonable answer to the question how to treat Gorham disease of the spine. Considering the review of the literature and the unfortunate consequence in the present case, we propose the following guidelines. Namely, if the affected vertebrae show no deformity such as kyphosis or subluxation, radiotherapy and medication should initially be chosen independently or in combination. Observation might be another choice expecting spontaneous arrest. If the number of affected vertebrae is small with a progressive deformity in the thoracic or lumbar spine, total spondylectomy is recommended. If the number is large as in this case or if the lesion occurs in the cervical spine with a progressive deformity, long posterior fusion with rigid instruments should be performed before the spinal cord has been compressed greatly.

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