

Lymphangiomas of the Spine

Two Cases Requiring Surgical Intervention

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Study Design. Two cases of lymphangiomas of the spine are presented.

Objectives. To report two cases of lymphangiomas of the spine requiring surgical intervention and to review the literature.

Summary of Background Information. Lymphangiomas are a rare childhood disease characterized by abnormal lymph tissue at multiple sites. Skeletal and visceral involvement are both common. Prognosis depends on the extent of extraskelatal disease.

Methods. Two cases of lymphangiomas causing neural compression and instability at the cervicothoracic junction are presented. Both patients underwent surgical decompression and stabilization.

Results. One patient died, whereas the other regained full function and activity.

Conclusions. Surgery is indicated when lymphangiomas cause neural compression and instability of the spine. Surgical outcome is strongly influenced by extraskelatal involvement. [Key words: cervicothoracic spine, lymphangiomas, surgery] *Spine* 2003;28:E45–E50

Lymphangiomas are a childhood disease characterized by abnormal lymph tissue at multiple sites. Lymphangiomas have been reported in every type of tissue except neural tissue. Skeletal and visceral involvement are both common.¹¹ Prognosis depends on the extent of extraskelatal disease.^{3,15,18,23,24} Spinal involvement usually consists of replacement of bone with lymph tissue, resulting in direct neural compression or bony instability.

Two cases of lymphangiomas causing neural compression and instability in the cervicothoracic spine are presented. Both cases required surgical intervention. Surgical outcome was strongly influenced by extraskelatal involvement.

Case Reports

Case 1. D.M. presented to the authors as a 15-year-old girl with a history of lymphangiomas. She previously had undergone radiotherapy for mediastinal involvement. Her only

disorder at presentation was limited neck motion. She was neurologically intact.

Radiographs showed severe lower cervical kyphosis with compensatory upper cervical hyperlordosis (Figure 1). Flexion and extension views showed stiffness in the lower cervical segment, with mobility above and below. Radiographs from 10 years previously showed no deformity (Figure 2). Magnetic resonance imaging (MRI) from 8 months previously showed a 29° kyphosis, with mild basilar impingement and mild spinal canal narrowing.

While awaiting a repeat MRI to check the status of the basilar impingement and spinal cord for preoperative planning, the patient experienced the development of pain radiating to her left arm and subjective sensory loss. The results of physical examination remained normal. An MRI demonstrated collapse at C6 and C7, the apex of the kyphosis, with no cord edema. The kyphotic deformity had progressed from 29° to 48° over 13 months. The decision was made to schedule the patient for anterior decompression with plating and posterior fusion.

Within the same week, the patient experienced shortness of breath and dyspnea. She underwent thoracentesis to drain a left chylothorax. The patient experienced two similar bouts of respiratory distress caused by the mass effect from the mediastinal involvement. After a brief trial of interferon therapy, she underwent resection of the left thoracic lymphangioma. After surgery and 6 weeks of draining the chylothorax with a chest tube, the patient went home in stable condition.

Within 1 month of the resection, the patient returned with right upper and lower extremity weakness and left upper and lower extremity sensory loss. She also had hyperreflexia and positive Babinski signs bilaterally. Bowel and bladder function were intact. The patient was admitted to the hospital.

An MRI showed severe compression from C5 to T1 (Figure 3), and CT scan showed significant bony destruction. Halo traction was attempted but was terminated after 24 hours because the patient experienced progressive neurologic loss.

The patient underwent an anterolateral approach with corpectomy and decompression from C4 to T2 with arthrodesis using structural allograft. The fibular allograft was slotted at the upper and lower ends and wedged into place. The patient was secured in a halo vest. Somatosensory-evoked potential did not change throughout the surgery. She tolerated the procedure well without complications. After surgery, she was transferred to the intensive care unit, with plans for posterior stabilization when she became medically stable.

During the next 2 weeks, the patient had worsening respiratory distress secondary to destruction of her left lung and persistent chylothorax. Chylopericardium developed, which required drainage. Systemic perfusion was maintained with multiple cardiovascular ionotropes. Her mental status waxed and waned. Occasionally, she was able to move her upper extremities against gravity. The patient died of cardiorespiratory failure 12 days after surgery.

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Figure 1. Case 1. Cervical radiograph at presentation. Note severe lower cervical kyphosis with compensatory upper cervical lordosis.

Case 2. S.D. was a 13-year-old boy referred with a diagnosis of cervicothoracic kyphosis. His father reported that the deformity, which was first noticed 1 year previously, had progressively worsened. The patient reported neck pain in the region of the deformity, and experienced occasional falling as a result of his legs giving way. He presented with hyperreflexia in all extremities and clonus in the bilateral lower extremities as the abnormal neurologic physical findings.

Radiographs and MRI showed a 125° kyphosis from C7 to T3 with wedging of T1–T2, narrowing of the canal with the cord draped over the kyphotic deformity and absence of CSF signal, and abnormal signal in the bone marrow from C4 to T3 (Figure 4). In addition, there was a similar lesion in the L5 body. A skeletal survey also showed a rib lesion, which was biopsied with CT guidance.

Histology confirmed a diagnosis of lymphangiomatosis. At this point, the patient and his parents agreed to surgical intervention to prevent progressive spine deformity and neurologic compromise.

The patient underwent a left fourth rib thoracotomy with anterior discectomy and morselized rib autograft from C6 to T4, as well as posterior exploration with facetectomies from C6 to T5. Lymph fluid was encountered on entrance to the left hemithorax. Furthermore, there was extensive involvement of lymphangioma in the vertebral bodies and lamina from C6 to T4. Biopsy confirmed the diagnosis.

After surgery, the patient was placed in 8 pounds of halo suspension traction. Initially, he had weakness in his left quadriceps and dorsiflexors. After the weight was decreased to 2 pounds, the patient's strength partially returned. The traction was slowly increased back to 8 pounds, with the patient's mo-

tor strength fluctuating. An MRI with the patient in the halo showed compression of the cord at the cervicothoracic junction.

Because of the cord compression and neurologic changes, the patient was taken back to the operating room for anterior vertebrectomies from C6 to T4 and fusion with iliac crest autograft, fibular strut allograft, and anterior titanium plate fixation from C4 to T4. The fibular allograft was inserted into the body of C5 through the inferior endplate and slotted into the body of T4 laterally. The patient received Decadron intraoperatively. Somatosensory-evoked potentials did not change during the case.

After surgery, the patient was unable to move his toes bilaterally and lacked extension of his right wrist and fingers. Emergent MRI showed edematous changes within the cord, with no distinct compression. The patient was stabilized in the ICU and maintained on Solu-Medrol.

A CT scan demonstrated residual bony compression anteriorly at the cervicothoracic junction. Therefore, the patient was taken back to the operating room for further decompression. Through an anterior supraclavicular approach, the plate and allograft were removed and the bodies of C6 to T2 were further decompressed posterolaterally. The anterior plate and fibular strut allograft then were revised from C4 to T4. After surgery, motor function improved in all extremities with residual weakness of the right hand and left leg.

Subsequently, posterior stabilization was performed with iliac crest bone graft and instrumentation from C2 to T7 (Figures 5 and 6). Lymph fluid leaked from the posterior elements of C4 to T3.

The patient's 2-month hospital course was complicated by



Figure 2. Case 1. Cervical radiograph 10 years before presentation. Note normal alignment with no deformity.

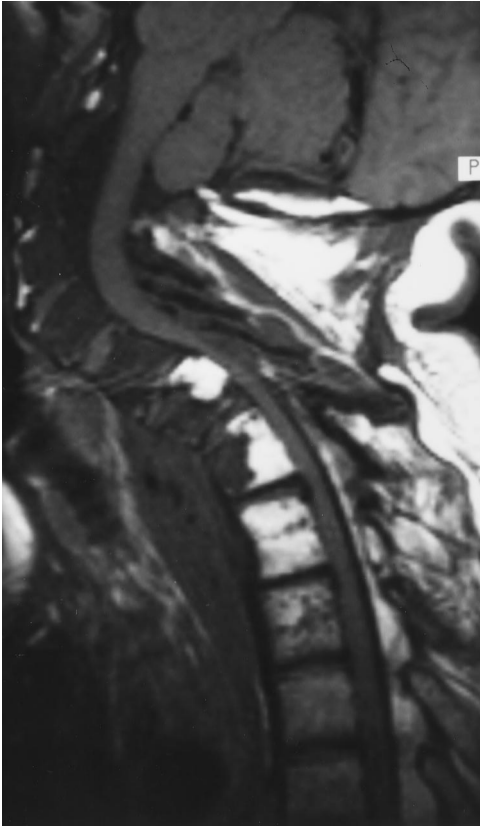


Figure 3. Case 1. Magnetic resonance imaging of the cervicothoracic spine. Note severe kyphotic deformity with cord compression from C5 to T1.

transient mesenteric artery syndrome and right chylothorax. Both conditions resolved, and the patient was discharged in stable condition. He remained in a halo brace for 3 months.

The patient was started on interferon 3 months after surgery for treatment of lymphangiomatosis. By 4 months after surgery, the patient had full function of both lower extremities, with mild residual weakness of his right hand. At the 1-year follow-up visit, complete hand function had returned. At this writing, the patient continues to receive interferon therapy and remains neurologically intact.

■ Discussion

Lymphangiomatosis is a rare disorder that usually affects children. In some studies, up to 65% of the subjects are children.^{8,11,14,18} Cases have been described with patients ranging in age from birth to 80 years.¹¹ Lymphangiomatosis is commonly believed to be a sporadic disease, but familial associations have been established.²⁴ Lymphangiomas are generally considered to be congenital malformations of the lymphatic system, although a neoplastic process is still a possibility.^{6,8,11-13,17,18,26-28}

Lymphangiomatosis involves multifocal collections of endothelial-lined tissue containing eosinophilic stained material or lymph.^{6,8,11,13,31} There is confusion over the distinction between lymphangiomas and hemangiomas. Hemangiomas are collections of endothelial-lined tissue containing red blood cells. Because many patients have both lymph and blood in cystic spaces, some authors

have grouped the two entities together as cystic angiomas or skeletal angiomas.^{3,7,13,15,24,28} Furthermore, Gutierrez and Spjut¹⁵ found that whether lesions were hemangiomas, lymphangiomas, or mixed hemangiomas, there was no difference in the symptoms, clinical course, prognosis, or treatment. In contrast, others have reported that unlike congenital hemangiomas, spontaneous involution of lymphangiomas is rare.^{9,18,21}

Other names that have been attached to this disease include generalized lymphangiectasis, cystic lymphangiectasis, diffuse skeletal angiomas, hamartous hemangiomas, and massive osteolysis.^{6,12,20,29} The distinction between lymphangiomatosis and Gorham disease, or “disappearing bone disease,” is worth noting. Gorham disease is unifocal, without skull involvement and without extraosseous origin.^{7,15,23}

Lymphangiomas have been reported in every tissue type, except nervous tissue. They usually present in the neck region.^{1,13} Commonly affected organs include the spleen, liver, and lungs. Multiple organs are involved in 75% of cases.¹¹ Skeletal involvement has been documented in up to 70% of cases.¹¹ The axial skeleton, including skull, ribs, shoulder girdle, spine, and pelvis is the typical site for lymphangiomas.¹⁶ Angiomas have been reported at distal sites such as the hands.^{24,28}

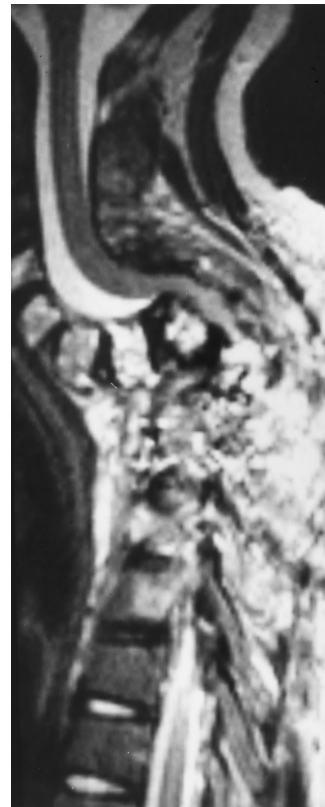


Figure 4. Case 2. Magnetic resonance imaging of the cervicothoracic spine. Note narrowing of the spinal canal with the cord draped over the kyphotic deformity and abnormal signal in the bone marrow from C4 to T3.

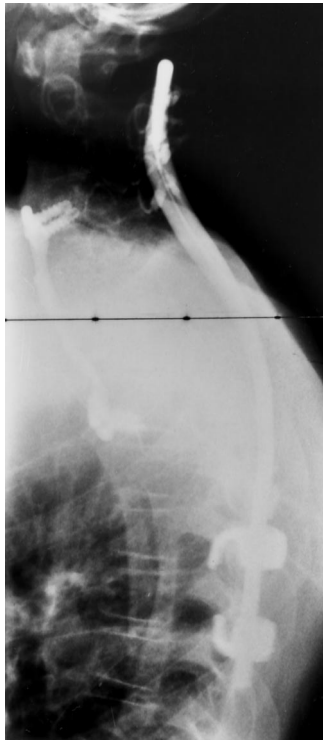


Figure 5. Case 2. Postoperative lateral radiograph of the cervicothoracic spine. Note correction of sagittal alignment with anterior fixation from C4 to T4 and posterior instrumentation from C2 to T7.

Lymphangiomatosis usually presents with soft tissue swelling caused by lymph blockage, especially in the neck region. Skeletal lesions may be discovered as a result of deformity or pain from a pathologic fracture. If this diagnosis is suspected, a skeletal survey is warranted.

Radiographically, the lesions are typically radiolucent cysts surrounded by a sclerotic rim.^{3,30} They usually are located in the medulla, but may appear in the cortex. The cortex may be expanded and thinned, but is never breached unless there is a pathologic fracture. There is no periosteal reaction.^{3,31} A foamy or honeycombed appearance is sometimes seen.^{12,27} The amount of surrounding sclerosis may increase with age of the lesion.^{12,16} Furthermore, a predominantly sclerotic variant, mimicking an osteoblastic process, has been described.^{12,16} In vertebrae, an area of rarefaction may be traversed by thick, bony trabeculae.⁴ Radiographic differential diagnosis includes histiocytosis X, fibrous dysplasia, hyperparathyroidism, Gaucher disease, congenital fibromatosis, neurofibromatosis, enchondromatosis, and metastatic bone lymphangioma.^{16,27}

Magnetic resonance imaging helps to determine soft tissue involvement. Heterogeneous bone marrow with intermediate to high signal intensity is seen on T1- and T2-weighted images.³ Computed tomography scan provides excellent visualization of lymphangiomas and their relation to adjacent structures.²¹ Brown *et al*³ reported evidence of intraosseous gas within lymphangiomas on CT scan.

The natural history of lymphangiomas involves a slowly growing mass without malignant change.^{13,28} Histologically, they are composed of mature cells.¹¹ The destruction is caused by replacement of inherent tissue as well as blockage of lymphatics and blood vessels. This may result in organ failure, occasionally associated with chylothorax or chylopericardium.^{11,13} In bones, this may cause loss of stability with pathologic fracture or deformity. Ramani and Shah²³ reported an unfavorable prognosis with involvement of the cervical spine.

A poor prognosis is associated with extraskeletal involvement because of systemic complications.^{3,18,23,24} In 36 cases of skeletal angiomatosis, Gutierrez and Spjut¹⁵ showed significant mortality associated with extraskeletal involvement. Among 11 patients without extraskeletal involvement, there was only 1 known death, which may not have resulted from the disease. Of 25 patients with extraskeletal involvement, 11 patients died.

Treatment depends on the degree of involvement with normal structures. Most bony lesions eventually stabilize without intervention.¹⁴ Pathologic fractures and instability are stabilized appropriately. Internal fixation may be difficult because of bone loss.⁶

Surgical resection is indicated when lesions compress vital structures causing dysfunction. Alternative treatment methods have been used for unresectable lesions including radiation, chemotherapy, embolization, and sclerotherapy.^{7,11,17,21,25} Recombinant interferon therapy has shown promising results for disseminated lymphangiomatosis.^{17,25}

In both of the reported cases, surgical decompression was indicated to minimize neurologic loss. In the first

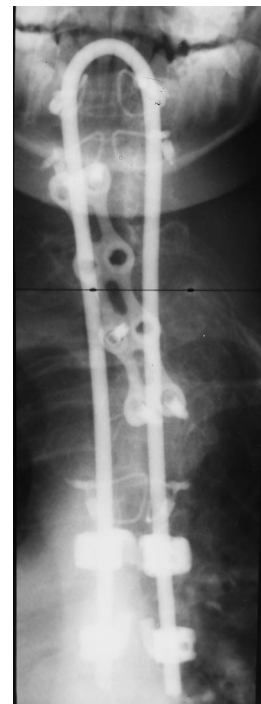


Figure 6. Case 2. Postoperative anteroposterior radiograph of the cervicothoracic spine.

case, the patient (D.M.) underwent anterior decompression as an urgent procedure. The anterior column was supported with fibular allograft. The plan was to supplement the allograft with posterior instrumentation and fusion. In the authors' experience, anterior decompression and allograft, when used in conjunction with posterior instrumentation, usually is successful in obtaining fusion. Fracture through the allograft has not been a problem so long as solid fusion is obtained posteriorly. The reported patient died before the posterior stabilization could be performed.

In the second case, the patient (S.D.) underwent initial anterior discectomy and soft tissue release with posterior facetectomies was done to see whether correction of the deformity would release the pressure on the spinal cord. In some cases of severe deformity, the authors have been able to decompress the spinal cord simply by straightening the deformity and then fusing in that position. However, in this case, when traction was applied, weakness developed in the patient's left leg. He was taken back to the operating room for vertebrectomies to remove the offending structures. Unfortunately, the patient's neurologic status was worse after this second procedure. A CT scan showed residual anterior bony compression. Therefore, further posterolateral decompression of the vertebral bodies was performed. After the fourth procedure, posterior fusion and instrumentation, the patient recovered all neurologic function and maintained solid fusion. This second case supports the authors' experience that anterior decompression and allograft can be successful when used in conjunction with posterior stabilization.

Several studies have confirmed the observation that anterior strut allograft with posterior stabilization has a high success rate.^{2,4,5,19} Fernyhough *et al*¹⁰ reported on multiple-level anterior cervical fusion. Of 67 patients with fibular autograft, 73% achieved fusion. Of 59 patients with fibular strut allograft, only 59% achieved fusion. However, in a follow-up study, these authors improved the allograft fusion rate to 74% with posterior stabilization.⁹

Bridwell *et al*² prospectively studied 24 adult patients with kyphosis or anterior column defects treated with anterior allograft and posterior stabilization and autogenous grafting. Only two patients showed some collapse of their anterior allograft. The other 22 patients attained fusion and maintained their correction.

There have been concerns that allograft may not incorporate sufficiently enough to maintain stability. Meding and Stambough¹⁹ and Buttermann⁵ have reported that it takes allograft longer than autograft to incorporate. However, Olson *et al*²² showed in canines that stand-alone allograft is able to incorporate at the margins and partially in the center.

Bridwell *et al*² reported that in most of his cases, radiographs showed remodeling and incorporation of strut allograft. He stated that anterior strut allograft with posterior stabilization is effective because there is a good vascular bed anteriorly, the strut graft is in compression,

and the posterior instrumentation reduces micromotion between the grafts. Furthermore, he used anterior instrumentation to hold the graft in place instead of providing structural support.

■ Conclusion

Two patients with neural compression and instability in the cervicothoracic region secondary to lymphangiomatosis are presented. Both underwent surgical decompression and stabilization. One patient died of extensive extraosseous involvement of the disease, whereas the other regained full function and activity.

■ Key Points

- Lymphangiomatosis is a rare childhood disease characterized by abnormal lymph tissue at multiple sites.
- Surgery is indicated when lymphangiomatosis causes neural compression and instability in the spine.
- Surgical outcome is strongly influenced by extraosseous involvement of the disease.

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