Unusual Bleeding of Aneurysmal Bone Cyst in the Upper Thoracic Spine

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Aneurysmal bone cyst (ABC) is a benign bone lesion and commonly affects young adolescents. It usually grows rapidly with hypervascularity. In the spine, it can cause extensive bone destruction and compress neural structures with eventual vertebral collapse. Operative management of such a lesion may be complicated by profuse hemorrhage. Herein, we report a 15-year-old boy who had an acute exacerbation of spinal cord compression because of an ABC in the upper thoracic region. With a two-staged operation that complicating with profuse intraoperative bleeding, decompressive curettage and stabilization of the vertebral column were assured. However, abrupt neurologic deterioration occurred because of rebleeding with spinal cord compression 1 month postoperatively. Secondary decompressive curettage and following local radiotherapy were undertaken to cure the disease. Neurological recovery and healing of spinal ABC could be expected if the lining of a cyst had been totally removed. At the 18-month follow-up examination, he was neurologically intact and without any backache, leg pain or gait disturbance. (Chang Gung Med J 2002;25: 183-9)

Key words: aneurysmal bone cyst, upper thoracic spine, rebleeding.

An aneurysmal bone cyst (ABC) is a benign, locally proliferative vascular disorder of non-neoplastic osseous lesions in children and young adults. These cysts can occur in any part of the skeleton, and are most commonly seen in the metaphyseal regions of tubular bones, comprising 1.4% of all primary bone tumors.¹ Seventy-five percent of ABCs occur before the age of 20 years and 90% are seen before the age of 30 years.² Approximately 15-20% of ABCs occur in the spine.³,⁴ They arise from the neural arch and occasionally invade the pedicle and vertebral body.⁵ These paravertebral masses grow rapidly with expansion and osteolysis of the adjacent vertebrae, and they exacerbate compression of the spinal cord or nerve roots with eventual vertebral collapse. Treatment modalities include surgery, radiation, transarterial embolization and injection of a variety of agents.¹,³⁰⁴† Surgical excision and bone reconstructive procedures for spinal ABCs may be complicated by profuse hemorrhage.⁴ However, recognition of unusual bleeding has not been adequately addressed in the orthopaedic literature.

ABC was distinguished from other bone cysts and giant cell tumors on the basis of its clinicopathological characteristics.¹⁰ In fact, ABCs are not aneurysms because they occur in bones rather than in arteries, and they are not bone cysts because they have no epithelial lining as unicameral bone cysts do.²,¹⁰ They do have cyst-like walls of predominant-
ly fibrous tissue. In Taiwan, this disease is rare, as reported over a 10-year period at the authors' institution. Controversy concerning treatment persists as regards to ABC localization, growth pattern, proximity of the spinal cord, spinal instability and focal recurrence. Some spinal ABC with aggressive biological behavior can produce rapid osseous destruction, vertebral fracture and profuse hemorrhage. Total excision is difficult because of its inaccessibility. The role of transarterial embolotherapy is not definite within a critical collapse in the cervicothoracic spine. Surgical treatment of this rare lesion becomes challenging when there is abrupt exacerbation of myelopathy and postoperative rebleeding.

CASE REPORT

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paresis developed two weeks prior to admission. A spastic gait and fecal incontinence were noted for 2 days. Physical examination revealed tenderness over the upper thoracic paravertebral region and bilateral lower extremities weakness (grade 4/5 muscle strength). The deep tendon reflexes of both knees and ankles were increased, and a positive Babinski sign was present bilaterally. Radiographs of the upper thoracic spine demonstrated destruction of the T4 lamina, spinal process, right pedicle, and collapse of the vertebral body (Fig. 1). Magnetic resonance imaging (MRI) showed an expansile soft tissue mass over the right posterior elements of the T4 level extending along the pedicle to the vertebral body with a heterogenous high density in T2 weighted images (Fig. 2A). It caused extradural compression of the spinal cord. After gadolinium injection, there was little septate enhancement, fluid level of content, and a rim of lower signal all around the mass (Fig. 2B).

An emergency decompressive laminectomy of T4 and upper T5 was done with posterior spinal

![Fig. 1](image1.png)  
**Fig. 1** Anteroposterior (A) and lateral (B) radiographs of the upper thoracic spine show destruction of the T4 lamina, spinal process, right pedicle and a collapsed vertebral body (arrows).
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instrumentation using an Isola device (AcroMed, Cleveland, OH, USA) from T1 to T8. Somatosensory evoked potential monitoring was used throughout the operation. Grossly, the yellow-brownish tumor was friable and spongelike. A marginal excision could be achieved as the spinal cord was adequately decompressed. The lesion consisted of an unclotted vascular bed under pressure which filled the conspicuous space and caused expansion of the affected bone. A blood loss of 1,900 cc was noted during surgery. Postoperatively, there was improvement of sphincter function, and he regained sensation and motor power in the bilateral lower limbs. The histopathological examination revealed multiple blood-filled lacunae lined by an indistinct epithelium, and the connective tissue stroma was filled with dilated capillaries, spindle-shaped cells and osteoclastic giant cells near the cavernous channels (Fig. 3). Few hemosiderin-laden macrophages were seen in the cystic wall and reactive osteoid was deposited along the thin fibrous septa. This was consistent with a diagnosis of ABC.

A second stage operation was done via an anterior approach 5 days later. T4 and T5 corpectomy including resection of the vertebral tumor, and interbody fusion from T3 to T6 with an iliac strut graft were performed using a video-assisted thoracoscopic procedure. Indistinct venous pooling occurred in the friable bone after ligation of the T4 and T5 vertebral segmental arteries and packing of the lesion. Total blood loss of 2,300 cc was noted in this procedure. The chest tube was removed on the fourth postoperative day. He had a complete recovery from paraparesis.

Unfortunately, acute onset of severe paraparesis (grade 2/5 strength of both lower limbs) occurred 4 weeks after surgery when he was scheduled to start radiotherapy after a radio-oncological consultation. An emergency myelogram plus computed tomography disclosed a nearly complete blockage of contrast at the T5 level and extradural compression of the spinal cord by a soft tissue mass from the right pos-

Fig. 2 (A) MRI shows an expansile soft tissue mass over the posterior elements of the T4 level, extending along the right pedicle to the vertebral body. Extradural compression of the spinal cord is noted. (B) After contrast enhancement, there is little septate enhancement of the lesion, fluid level of the content and a rim of lower signal around the mass. The T4 vertebral body is collapsed and a contiguous T5 vertebra is involved.
terolateral aspect of the spinal canal (Fig. 4). The iliac strut graft remained without any displacement. No transarterial embolization of spinal angiography was attempted on account of acute myelopathy and high risk of interference with the blood supply to the spinal cord. The previous laminectomy of T3-4 was enlarged immediately to evacuate the unclotted blood. During the procedure of curettage on the cortical rim, the peridural tissue bled very easily. A total blood loss of 3,000 cc was encountered intraoperatively. With poor delineation of the cortical outline, the possibility of a coexisting primary lesion had to be considered until there was no proof of malignancy. The patient promptly regained neurologic function after operation. Radiotherapy with a dose of 2,000 rads for ABC followed. At the 18-month follow-up, the patient was neurologically intact without any backache, leg pain or gait disturbance. Radiographic examinations showed solid fusion without any evidence of recurrence.

**DISCUSSION**

ABC is an expansile, osteolytic lesion which poses an aggressive vascular disorder in the bones. It is eccentrically blown out, like the saccular protrusion of an aneurysm, and strands of bone form a soap-bubble appearance on radiographs. The blood-filled space undergoes pressure with marked ballooning of a thinned cortex and exhibits hypervasularity. Pathological fracture of ABC (25%) is frequent in contrast to other bone cysts (10%) particularly with a marked expansile and destructive lesion. The period between the onset of symptoms and treatment is less than 4 months especially in an aggressive cyst. It is usually treated successfully by surgical en bloc resection, curettage and bone graft at the metaphyseal region of the extremities. Spinal ABC may be more serious because of the proximity to the spinal cord and pathological fracture with a sequential vertebral collapse.

ABC has an uncertain pathogenesis and therapy is not well defined, particularly because of profuse hemorrhage and focal recurrence. Theories about pathogenesis include local hemodynamic disturbance, arteriovenous anomaly and abnormal endochondral bone remodeling secondary to coexisting bone lesions. A primary ABC is defined histologically as anastomosing fibrous-walled channels that have a complete or incomplete lining of endothelial cell, but in contrast to true blood vessels, they contain no elastic lamina or muscular layer. A secondary ABC has the same microscopic characteristics, but with the additional findings of a coexist-
ing lesion\cite{13} - either benign or malignant. The hemorrhagic and cystic changes of these lesions can also be found in giant cell tumors, chondroblastoma, osteoblastoma, hemangioma, fibrous dysplasia, nonossifying fibroma, fractured bone cyst and telangiectatic osteosarcoma.\cite{2,16} Eighty percent of associated lesions dominate the radiological criteria of diagnosis especially when they are malignant.\cite{11,13} Poor delineation of the cortical outline should raise the index of suspicion until there was no proof of malignancy. A benign process is delimited by a calcified periosteal membrane which aids in differentiating ABC from a bony cyst, fibrous dysplasia or nonossifying fibroma.

The spinal lesions of ABC usually have nonspecific clinical findings such as diffuse pain accompanied by stiffness in the back. A palpable mass or aggravating backache in the supine position alerts the physician to this problem.\cite{1,3,4} Depending on the level of involvement and the extent of spinal cord compression, a wide variety of neurological symptoms and signs are noted later, ranging from mild radiculopathy to complete paraplegia or tetraplegia. Radiographically, most spinal ABCs disclose pure osteolysis with little trabeculation on the neural arch. The extensive type may invade along the pedicle to the vertebral body with collapse, and across the disc space to involve the adjacent vertebrae.\cite{3,6} Internal septation with varying density of fluid content and lower signal in the rim surrounding the lesion may be evident on MR images due to sedimentation of uncoagulated blood. However, they are not consistent findings of the pathophysiological changes in accordance with the proliferative nature of ABCs. Therefore, Capanna et al\cite{2,3} classified the activity of ABC as inactive, active or aggressive, depending upon its state of evolution. Recurrence (19\%) always occurs early in aggressive or active cysts.\cite{10}

In our patient, the preoperative MRI indicated an aggressive stage of lesion with little reparative osteogenesis, contiguous vertebral involvement and infiltration of the surrounding soft tissues. Removal of the huge tumor and concomitant bone reconstruction in the upper thoracic spine must be accomplished by a combined anterior and posterior procedures.\cite{5,12} There is intense bleeding once the thin outer shell of bone has been opened. Preoperative transarterial embolization to reduce bleeding amount is risky in a case of collapsed spine with severe cord compromise.\cite{7,8} Urgent spinal reconstruction is strongly indicated to prevent irreversible cord damage in spite of profuse hemorrhage.\cite{12} Bleeding seems to come from the sinusoid lining of the thin-walled vascular space, and may be difficult to control until all the lining has been removed.\cite{1} Neurological deterioration within 4 weeks of surgery reflects its aggression after the marginal excision. Further curettage and radiotherapy can prevent recurrence, as confirmed in our patient.

As noted in the literature, the natural course of spinal ABCs can vary from one of a rapidly destructive lesion to that of a small, quiescent lesion which spontaneously regresses after a simple biopsy.\cite{14} The recommended methods of treatment include en bloc resection, curettage, radiotherapy, transarterial embolization or a combination, such as excision with postoperative radiotherapy or presurgical embolization and excision.\cite{11,3,4} Total excision is the treatment of choice, but it is feasible when it involves only the lamina or transverse process. Curettage and bone grafting are the standard treatment for the central lesions of most spinal ABCs. If there is vertebral body involvement, combined anterior and posterior approaches for tumor removal and fusion are required to maintain the structural integrity. Surgical excision may become difficult because of extensive hemorrhage and inaccessibility, so selective embolization after spinal angiography has been suggested before a tumor removal.\cite{7,8} However, it is risky in the face of acute vertebral collapse. A bone reconstructive procedure seems to be more important than embotherapy. The addition of postoperative radiotherapy has been proposed if some infiltrative tissue remains in patients after partial excision of a huge tumor.\cite{1,2} Hey et al reported no recurrence in cases of total excision, 25% recurrence in cases of partial excision, 6% recurrence in cases of partial excision followed by radiotherapy, and 11% recurrence in cases of radiotherapy alone.\cite{10} The prognosis is good, as there is no recurrence after a second curettage if all the spaces are opened.

Some reports on spinal angiography demonstrated diffuse opacity in an arteriogram late phase, arteriovenous shunt and indistinct venous pooling of contrast in the cystic areas as characteristics of ABC.\cite{2,4} No definite nutrient vessels can be identi-
fied, so the effect of embolotherapy is not predictable. The potential complications include embolizing normal adjacent tissue, iatrogenic pulmonary emboli and catheter-related complications.\(^9\) The authors did not use embolotherapy, due to the danger of interference with the blood supply to the spinal cord in an event of abrupt myelopathy within a critical collapse in the cervicothoracic region. Radiotherapy has been advocated as an adjuvant method to diminish the blood supply of tumor when complete surgical excision is not possible. Radiation-induced complications include growth plate disturbance, myelopathy, gonade damage and sarcomatous changes.\(^1,2\) The radiation risk of 2.9\% is much less than that of catastrophic bleeding and spinal cord compression when attempting to excise a large tumor.\(^1,3,4\) It appears that radiotherapy not exceeding 2,000 rads is safe. Intralesional injection of spinal ABCs with thrombogenic agents and calcitonin has been reported,\(^9\) but further clinical experience is necessary to establish their efficacy.

ABC may be the development of a vicious hemodynamic cycle in a bone. The growth rate of a lesion is inversely dependent upon the venous drainage within the lining of the cyst. Recurrence after surgery is related to the cavities not being fully opened. Neurological recovery and radiographic healing of spinal ABC can be expected if the lining of a cyst has been totally removed. A residual small cyst with radiolucency is of concern and requires regular follow-up over 1 year.\(^6\)

**REFERENCES**

上胸椎動脈瘤骨囊腫合併異常出血

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動脈瘤骨囊腫是一良性，但會迅速局部擴張，血管增生、骨融解的骨病變，主犯二十一歲前的青少年，多發生在四肢長骨的幹板近端。當病灶侵犯脊椎部位，侵犯破壞胸弓、椎體使其骨折、多量出血，並會壓迫脊髓神經，致有多樣的臨床表徵。手術切除骨囊腫、植骨後、血管栓塞術及術後放射治療，常因囊腫生長部位、速度、出血程度、神經壓迫輕重而有不同。文獻上少有強調頸胸椎處的動脈瘤骨囊腫，再發出血使神經壓迫再度惡化時，如何應變之道。我們提出的病例報告是一名15歲男孩，因上胸椎動脈瘤骨囊腫出血壓迫脊髓症狀，先以後位囊腫清除、減壓、胸椎內固定手術，再以影像輔助之胸腔鏡手術，切除囊腫及前方椎體融合。不幸一個月內再發出血，急速下肢無力，緊急後位清除血腫、囊腫界面徹底剝除，使神經症狀復原，再補以術後放射治療，目前十八個月追蹤，並無再發現象。對此上胸椎處巨大、有侵襲性的動脈瘤骨囊腫，再發性異常出血壓迫脊髓，並不適合血管栓塞，以免脊髓缺氧之虞。重建穩定脊椎架構，前後位徹底剝除囊腫是首要課題。因有再發出血之虞，術後放射治療，密切追蹤一年以上是必要的。（長庚醫誌 2002;25:183-9)

關鍵字：動脈瘤骨囊腫，上胸椎，再出血。