Aneurysmal Bone Cyst of the Lumbar Spine in a Patient with Turner Syndrome: A Case Report

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The patient is a 19-year-old female with Turner syndrome. She presented at our hospital after acute exacerbation of low back pain that had been present for one month. Her body height was 140 cm, and her weight was 39 kg. Tenderness was noted in the left buttock, but no lower limb muscle weakness or sensory disorders were observed. Plain radiographs showed an osteolytic lesion involving the left L5 vertebra with a winking owl sign observed on anteroposterior(AP) views (Fig. 1a). Computed tomography (CT) revealed pathological fractures of the L5 vertebral body and left pedicle along with osteolytic changes (Fig. 1b). Magnetic resonance imaging (MRI) revealed a lobulated lesion with fluid-fluid levels in the left vertebral arch, pedicle, and vertebral body of the L5 vertebra with high and low signal intensities on T2- and T1-weighted images, respectively. (Fig. 2a, b).

An aneurysmal bone cyst (ABC) of the L5 vertebra was suspected from the image findings. We performed preoperative arterial embolization of the bilateral L4 lumbar arteries and the left iliolumbar artery to prevent intraoperative hemorrhage (Fig. 3a). Using intraoperative navigation to identify tumor margins, a thorough curettage was performed to remove the tumor (Fig. 3b). Rapid intraoperative diagnosis supported the diagnosis of ABC. The final pathological diagnosis of ABC was made by confirming the fibrous connective tissue accompanied by capillary proliferation and partial fibrin deposition, with no signs of malignancy (Fig. 3 c, d). Pedicle screws were inserted into L4 and S1, and an expandable cage packed with autologous bone chips was placed between the vertebral bodies to reconstruct the lumbar spine's anterior strut (Fig. 4a).

After surgery, she had complete pain relief. At two years follow up, CT and MRI showed no apparent signs of recurrence (Fig. 4b, c).

ABC frequency is reported to be approximately 1.4% of all primary bone tumors, with 3-30% of the cysts originating in the spinal column[1]. In the spine, ABCs mostly occur in the thoracic or lumbar spines of young patients aged 10-20 years, with approximately 60% affecting the posterior spinal column components[2]. The treatment options for ABC are surgical curettage and bone transplantation[2-4], embolization[5], radiotherapy[6], and drug injections[7]. Recently, the effective treatment of ABC with embolization alone was reported[5], but there is a risk of spinal cord infarction due to inadvertent embolization of the Adamkiewicz artery. Therefore, surgery has been selected in most cases to treat ABC that arise in the spine, but there are reports of severe intraoperative hemorrhage[1,6]. Therefore, recent reports on ABC treatment often include accounts of surgical treatment with preoperative arterial embolization[3,4]. Along with the therapeutic effects of embolization, the reduction in intraoperative hemorrhage allows the surgeon to operate safely and precisely. In this case, arterial embolization was suitable because the ABC was localized in the L5 vertebra, where there is a low risk of embolizing the Adamkiewicz artery. Since she had a pathologic fracture of L5, we selected surgery instead of observing the effects of embolization.

This case is rare because of the coexistence with Turner syndrome. Patients with Turner syndrome are mainly characterized by a short stature and lack of secondary sexual characteristics[8]. The risk of gonadoblastoma, colon cancers, and central nervous system tumors have been reported to be greater in patients with Turner syndrome than in the normal population, possibly due to the allelic losses of the X chro-

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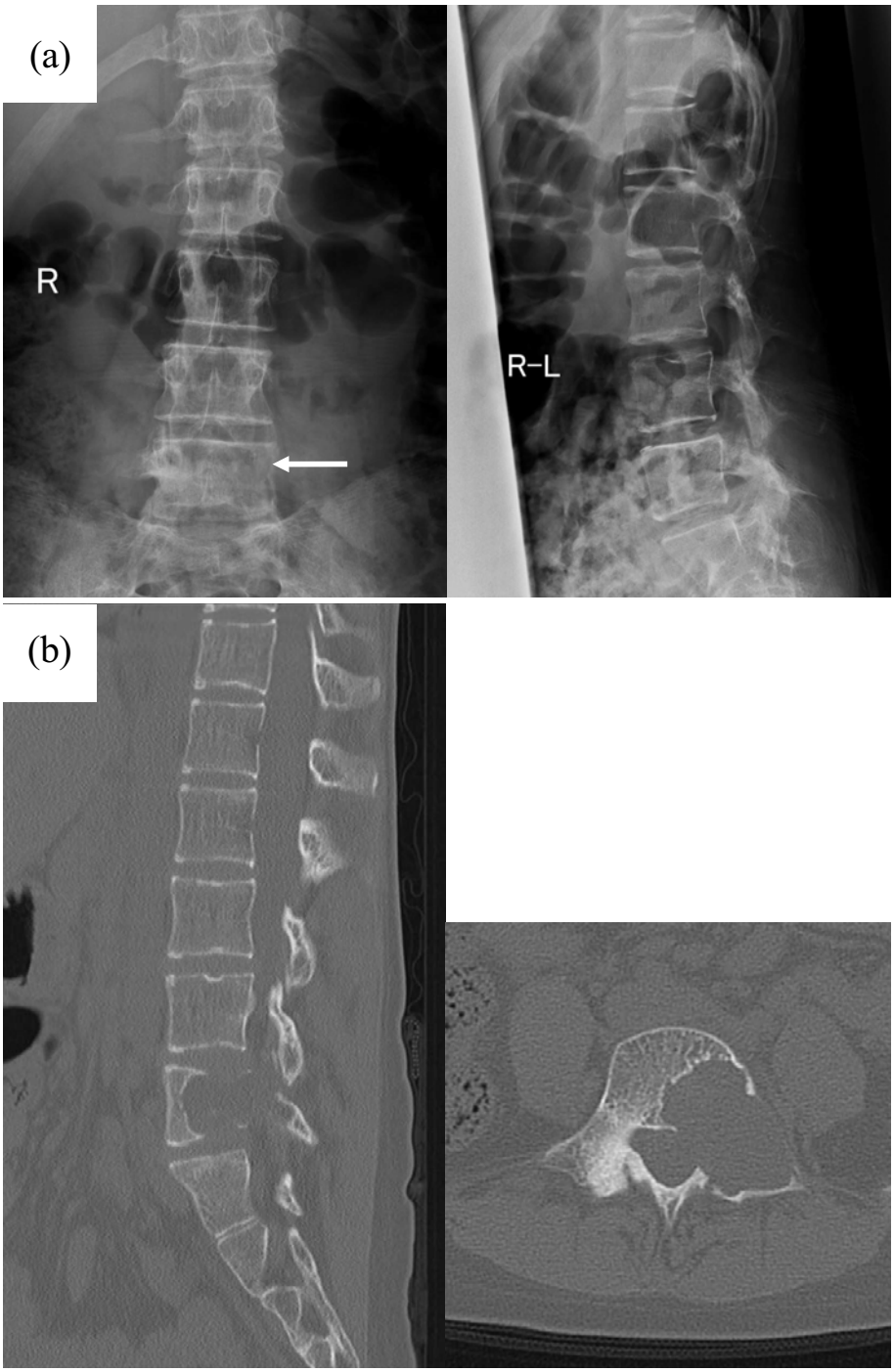

Figure 1. Preoperative X-ray and CT.
(a) X-ray AP view showing a lytic lesion involving the left L5 vertebral body illustrating a winking owl sign (arrow) (b) The reconstructed lumbar CT scan reveals a lytic expansive lesion involving the L5 vertebral body and left pedicle. Left: sagittal view, and right: axial view.

mosome which has been shown to contains genes related to immunity and cancer. Growth hormone therapy, to treat these patients' short stature, may also increase the risk of cancer[9]. Since this patient had received previous growth hormone therapy to increase her growth rate, it is possible that the growth hormone therapy may have lead to the onset of ABC. However, there has been no report of ABC occurring in a patient with Turner syndrome, so the relationship between Turner syndrome and ABC may not be strong. The height of the present case was 140 cm, which is short compared with the mean body height (157.7 cm) of 19-year-old Japanese females. Since she was without neurological symptoms, we were able to preserve the nerve roots by locally adjusting the cage height with an expandable cage to reconstruct the anterior column[10].

Here we reported a case of ABC occurring in the spinal

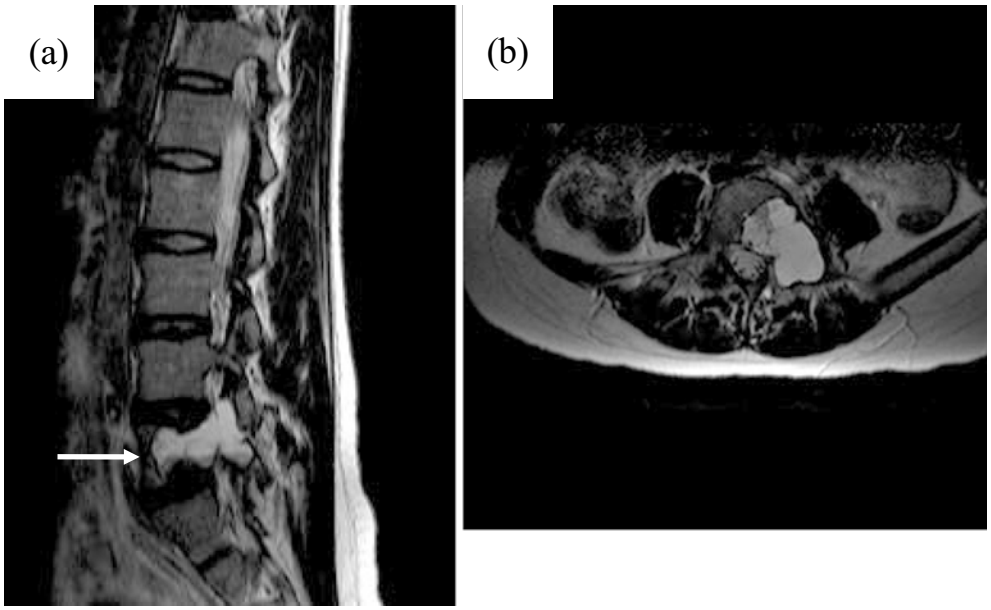

Figure 2. MRI of the lumbar spine showing characteristic findings of aneurysmal bone cysts.
T2-weighted sagittal images and T2-weighted axial image centered on the L5 pedicle revealing a cystic lesion involving the left L5 vertebral arch, pedicle, and vertebral body with fluid-fluid levels (arrow).

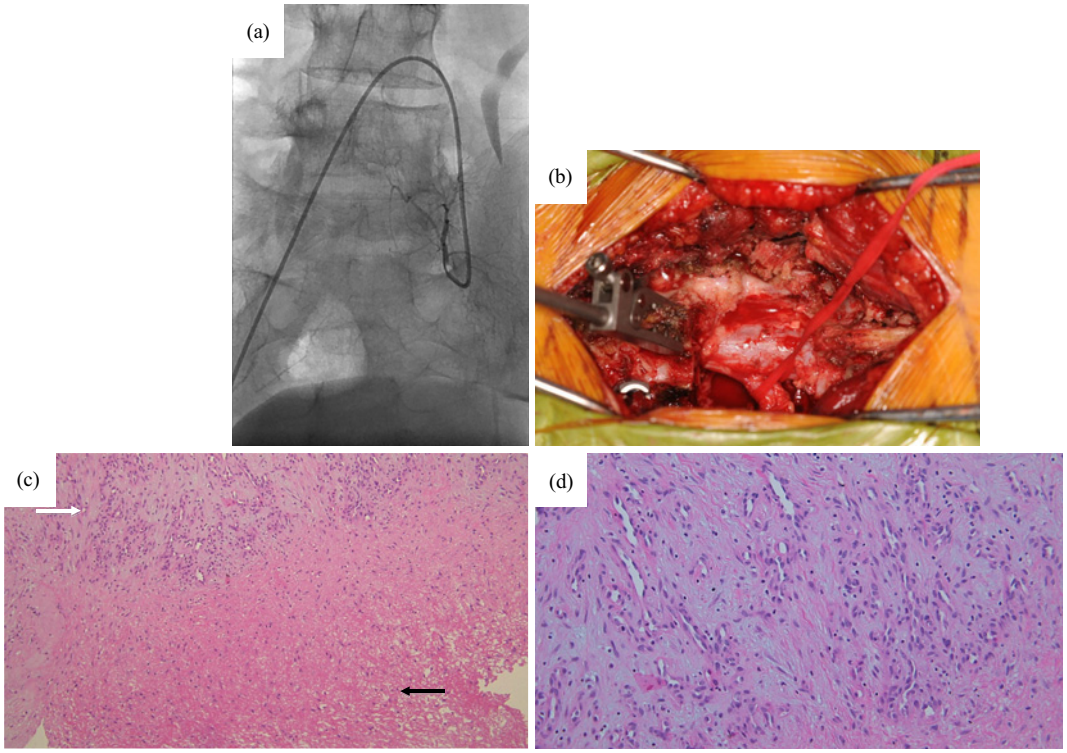

Figure 3. Preoperative embolization and Pathology photograph.
(a) Embolization of the bilateral L4 lumbar arteries and left iliolumbar artery was performed. (b) Intraoperative photograph after resection of the tumor. The left L5 nerve root was preserved, and the cage was inserted cephalad to the L5 root and then expanded. (c) In a low-power view, fibrous connective tissue associated with capillary proliferation (white arrow) and fibrin deposition (black arrow) are observed. These findings are consistent with the diagnosis of an aneurysmal bone cyst. (d) In a high-power view, there is fibrous connective tissue associated with capillary proliferation. No malignancy is observed.

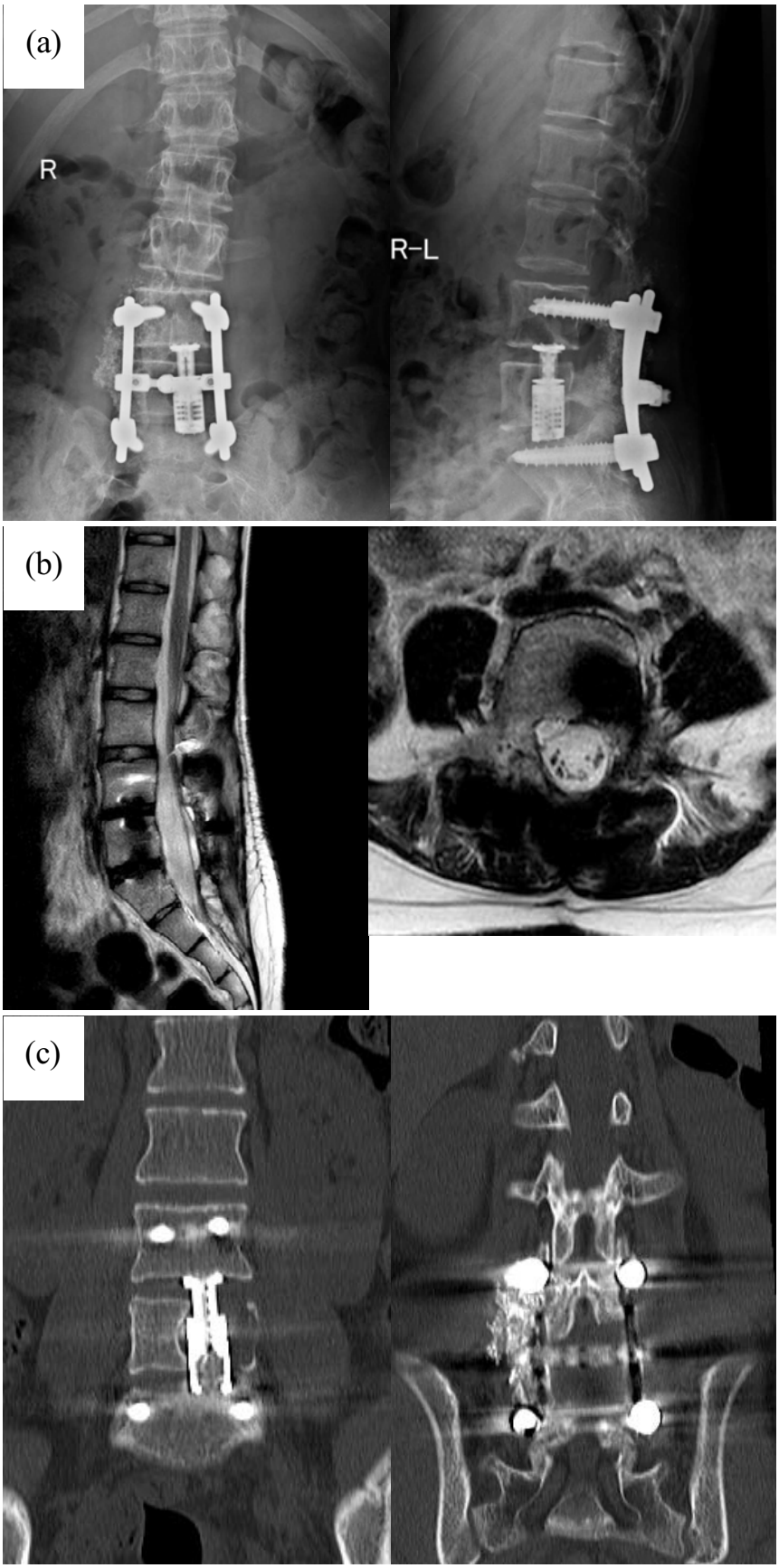

Figure 4. Postoperative X-ray and MRI.
(a) Postoperative X-ray of the lumbar spine (b) Postoperative MRI two years after surgery with no signs of recurrence. (c) Postoperative CT two years after surgery showing interbody fusion between L4 and S1 within the expandable cage and by the right posterolateral fusion mass.

column of a patient with Turner syndrome who was treated with preoperative embolization, surgical curettage, and posterior fixation using an expandable cage.

Conflicts of Interest: The authors declare that there are no relevant conflicts of interest.

Author Contributions: Tadashi Nukaga, and Akihiko Hiyama wrote and prepared the manuscript, and all of the authors participated in the study design. All authors have read, reviewed, and approved the article.

References

1. Zileli M, Isik HS, Ogut FE, et al. Aneurysmal bone cysts of the spine. Eur Spine J. 2013;22(3):593-601.
2. Hay MC, Paterson D, Taylor TK. Aneurysmal bone cysts of the spine. J Bone Joint Surg Br. 1978;60-B(3):406-11.
3. Papagelopoulos PJ, Currier BL, Shaughnessy WJ, et al. Aneurysmal bone cyst of the spine. Management and outcome. Spine. 1998;23(5):621-8.
4. Zenonos G, Jamil O, Governale LS, et al. Surgical treatment for primary spinal aneurysmal bone cysts: experience from Children's Hospital Boston. J Neurosurg Pediatr. 2012;9(3):305-15.
5. Amendola L, Simonetti L, Simoes CE, et al. Aneurysmal bone cyst of the mobile spine: the therapeutic role of embolization. Eur Spine J. 2013;22(3):533-41.
6. Boriani S, De Iure F, Campanacci L, et al. Aneurysmal bone cyst of the mobile spine: report on 41 cases. Spine. 2001;26(1):27-35.
7. Gladden ML, Jr., Gillingham BL, Hennrikus W, et al. Aneurysmal bone cyst of the first cervical vertebrae in a child treated with percutaneous intralesional injection of calcitonin and methylprednisolone. A case report. Spine. 2000;25(4):527-30; discussion 531.
8. Bondy CA, Turner Syndrome Study Group. Care of girls and women with Turner syndrome: a guideline of the Turner Syndrome Study Group. J Clin Mol Endocrinol. 2007;92(1):10-25.
9. Schoemaker MJ, Swerdlow AJ, Higgins CD, et al. Cancer incidence in women with Turner syndrome in Great Britain: a national cohort study. Lancet Oncol. 2008;9(3):239-46.
10. Sasani M, Ozer AF. Single-stage posterior corpectomy and expandable cage placement for treatment of thoracic or lumbar burst fractures. Spine. 2009;34(1):E33-40.