



Spinal Aneurysmal Bone Cysts – Case Series

Asen Cekov¹, Ivan Tarev¹, Anastas Kanev¹

¹ Neurosurgery Clinic, Acibadem City Clinic, Tokuda University Hospital, Sofia, Bulgaria

Corresponding author: Ivan Tarev, Acibadem City Clinic Tokuda Hospital, Sofia, Bulgaria; Email: tarevmd@gmail.com

Received: 21 February 2024 ♦ **Accepted:** 7 May 2024 ♦ **Published:** 30 June 2024

Citation: Cekov A, Tarev I, Kanev A. Spinal aneurysmal bone cysts – case series. *Folia Med (Plovdiv)* 2024;66(3):402-408. doi: 10.3897/folmed.66.e121395.

Abstract

Aneurysmal bone cysts are benign neoplastic lesions that are extensively vascularized and can occasionally exhibit destructive and expanding activity. They typically affect young people, with a small predilection toward females, and most frequently arise during the second decade of life. Aneurysmal bone cysts account for about 1% of all bone tumors and usually develop in long tubular bones, with the spine being much less commonly affected.

We present here three cases of aneurysmal bone cysts in the spinal cord that were surgically treated at the Department of Neurosurgery between 2009 and 2022. These patients were admitted to the clinic with symptoms of spinal cord or vascular compression. The volume of operative intervention was total or subtotal resection of the formation. No postoperative neurological deficits were observed, and no early complications were recorded after the interventions. All patients were discharged from the department with clinical improvement. One underwent reoperation after 4 years because of a cyst recurrence. The diagnosis of aneurysmal bone cyst was confirmed by histological examination in all cases.

Keywords

aneurysmal bone cyst, operative treatment, results

INTRODUCTION

Spinal aneurysmal bone cysts (ABC) are benign, expansive lesions located under a thin subperiosteal bone layer that, when infiltrating the body, arches, and articular processes of the vertebrae, can cause compression of the nerve structures in the vertebral canal. The most common symptoms are pain, deformity and neurological dysfunction. The goal of surgical treatment is decompression of the neurovascular elements, excision of the tumor mass and correction of a possible vertebral dislocation, with subsequent stabilization if necessary.

Clinical case 1

A matter of interest is the case of an 11-year-old girl who underwent emergency surgery at another clinic due to suspected malignant destruction of Th5-Th6 vertebrae and was referred for oncological treatment. A highly vascularized bone tumor

protruding into the vertebral canal was found intraoperatively, and a three-level, wide decompressive laminectomy was performed. Due to imaging data of a recurrent destructive neoprocess involving the fifth and sixth thoracic vertebrae (Fig. 1) and symptoms of local soreness and weakness in the lower limbs, the girl was admitted to the clinic for surgical treatment.

After resection of the tumor tissue that was infiltrating the bony structures and compressing the myelon, stabilization was performed in two stages. During the first stage, the posterior segment was stabilized with sublaminar hooks, which do not limit the growth of the spine, and at the second stage, employing a transthoracic approach, a titanium mesh filled with bone fragments was implanted in place of the missing parts of the vertebral bodies (Fig. 2). The histological examination showed blood-filled cystic spaces with a cellular giant cell-rich wall, and the diagnosis of aneurysmal bone cyst was established (Fig. 3). Fourteen years later, the girl is in excellent condition, without spinal deformities or neurological deficits, and an active athlete.

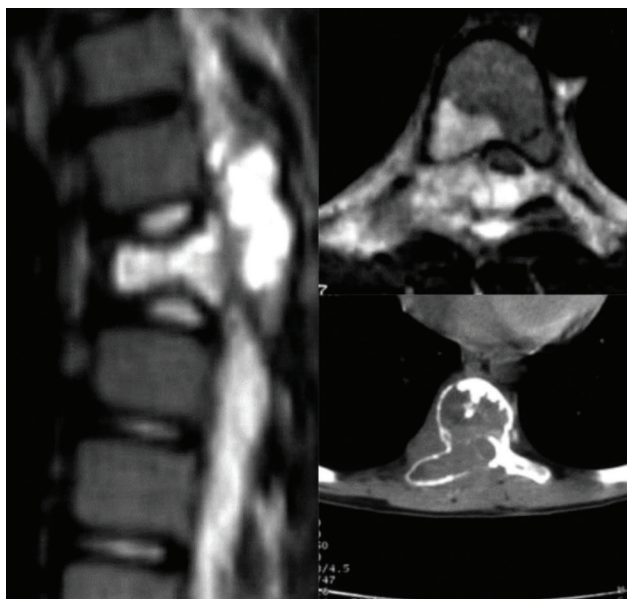


Figure 1. Preoperative imaging studies of a girl with an aneurysmal bone cyst at the Th5-Th6 level showing a cystic lesion that affects the body, pedicle, and arch of the vertebrae.

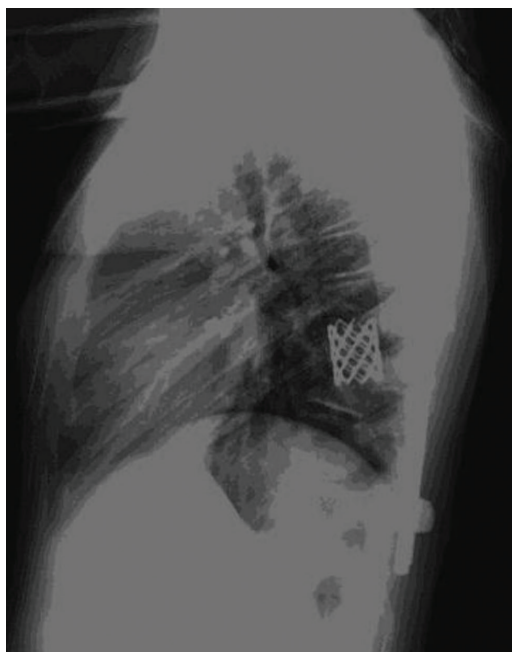


Figure 2. Radiograph of a girl with an aneurysmal bone cyst at the Th5-Th6 level after surgical treatment.

Clinical case 2

An 11-year-old boy was brought to the neurosurgery clinic because he was unable to bend over and was having trouble walking due to excruciating lower back pain. The neurological examination revealed discrete weakness and hypoesthesia of the lower extremities. The MRI study showed evidence of a lesion covering the arch and body of the L1 vertebra that was compressing the myelon. During the surgical interven-

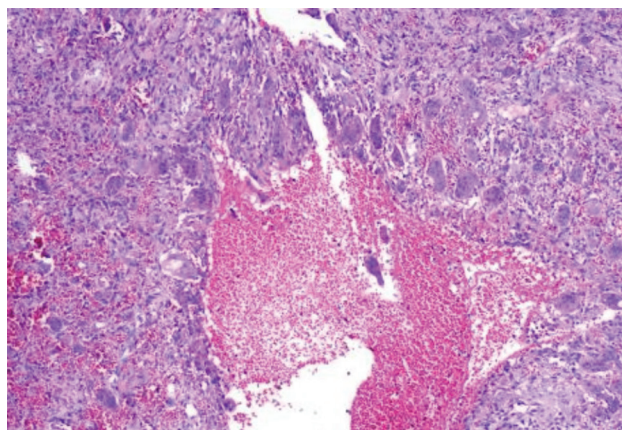


Figure 3. Hematoxylin and eosin (H&E): blood-filled cystic space with a cellular giant cell-rich wall.

tion, a highly vascularized paraspinal tumor, entering the vertebral canal and compressing the nerve structures, was encountered. A total resection of the lesion was performed under optical magnification and tissue material was sent for histological verification. Following surgery, the patient's overall and clinical somatic condition improved, and the postoperative pain syndrome decreased. Histological examination showed a benign tumor composed of blood-filled, giant cell-rich walled spaces and osteoclast-like giant multinucleated cells with regular round nuclei, without atypia (Figs 4, 5). The diagnosis of aneurysmal bone cysts was established. During the follow-up, regular check-ups over a period of 3 years were performed, and the boy manifested no significant symptoms.

During the fourth postoperative year, the patient came to our institution with newly emerging pain in the thoracolumbar area and difficulty walking, accompanied by episodic headaches. An MRI study of the spine was performed, showing evidence of a recurrent cyst in the L1 vertebra (Fig. 6).

The boy underwent a reoperation, during which we performed a partial hemilaminectomy of the L1 vertebra on the left and curettage of a bone cyst, affecting the pedicle and body of the same side. The patient was discharged from the clinic on the third postoperative day with significant improvement in symptoms and no evidence of a residual lesion. He is being monitored regularly at follow-up examinations and has no significant symptoms. Control MRI studies show no data for recurrence of the lesion (Fig. 7).

Clinical case 3

A 76-year-old man was admitted to the clinic with a two-month history of severe headaches, recently accompanied by vertigo and swallowing disorders. A few years ago, he experienced an episode of a similar severe headache and difficulty swallowing. The imaging study showed data of ischemic disorders in the left posterolateral segment of the medulla oblongata, a cystic lesion of the left interarticular segment of the C2 vertebra, and a fused fracture of the dens

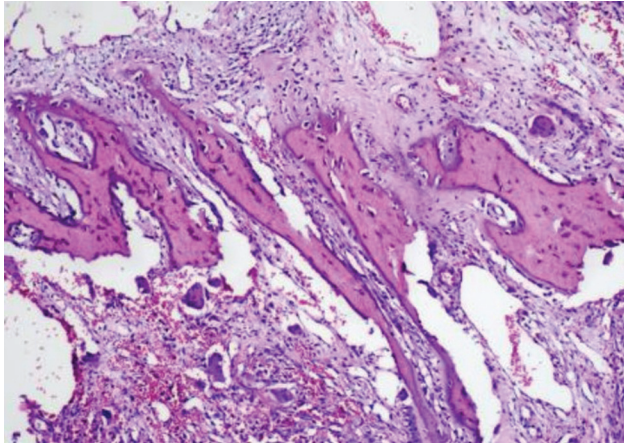


Figure 4. Trabecular bone in the cyst wall and giant osteoclast-like cells (H&E).

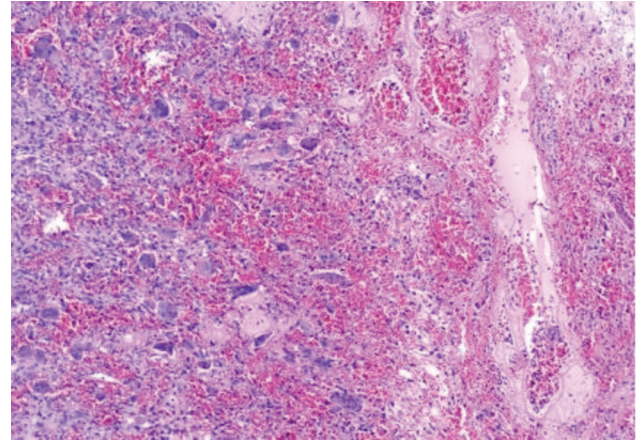


Figure 5. Blood-filled cystic spaces with a cellular giant cell-rich wall (H&E).

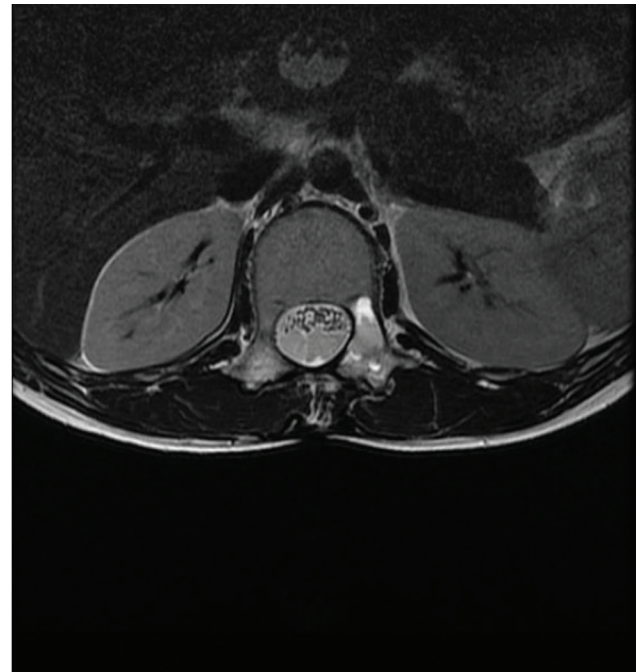


Figure 6. MRI of a boy with an ABC showing a cystic lesion affecting the left pedicle and the vertebral arch of the L1 vertebra.

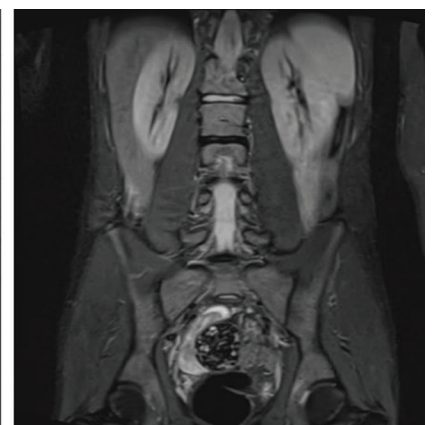
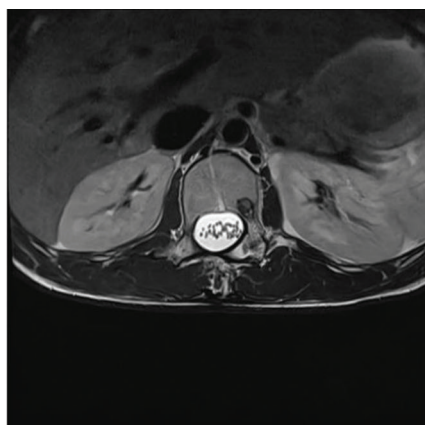


Figure 7. MRI of a boy with an ABC 3 years after the second operation, showing no recurrence of the lesion.

axis with anterior translocation of C1, causing compression of the myelon and the left vertebral artery. Intraoperatively, we found a severely compressed dura matter by the arch of the C1 vertebra due to its anterior translocation. The left interarticular segment of the C2 vertebra was visibly swollen and deformed. After performing a C2 laminotomy, a tumor formation with a central hemorrhagic part and a peripheral solid capsule was visualized. We performed a laminectomy of the C1 vertebra and resection of the C2 vertebra lesion, demonstrating apparently healthy margins. Good decompression of the myelon and vascular structures was achieved. The patient was discharged on the third postoperative day with solid improvement in neurological status and general somatic condition. Histological examination showed numerous giant cells in connective tissue that line large sinusoidal blood-containing spaces and blood-filled cystic spaces with cellular giant cell-rich walls (Figs 8, 9). The diagnosis of aneurysmal bone cyst was established.

DISCUSSION

ABC stands for aneurysmal bone cyst, where “aneurysmal” denotes the existence of an enlargement and “cyst” denotes

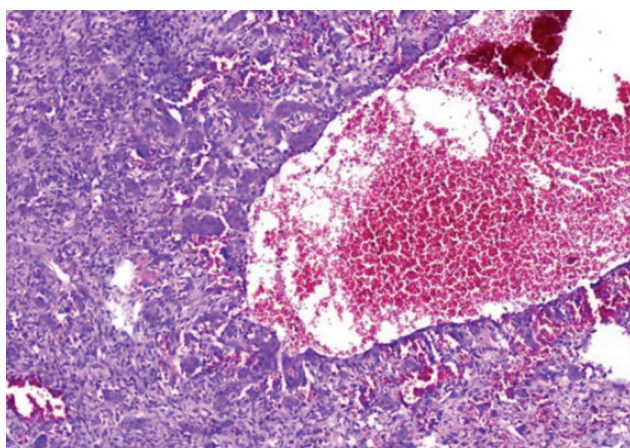


Figure 8. Numerous giant cells in connective tissue that line large sinusoidal blood containing space (H&E).

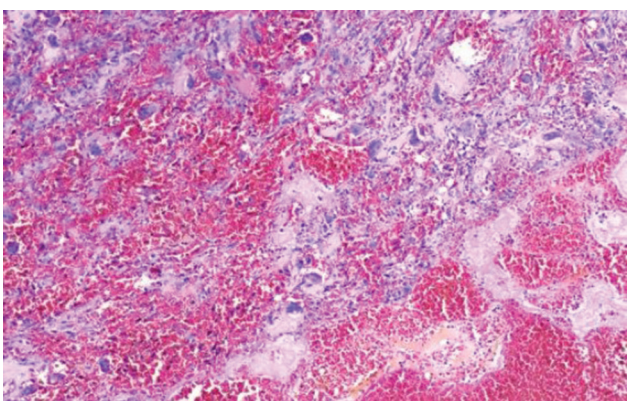


Figure 9. Blood-filled cystic spaces with a cellular giant cell-rich wall (H&E).

the presence of cavities filled with fluid. But modern studies prove that it is not about “aneurysm” and “cyst”, but about a tumor with an unclear etiology and pathogenesis.

^[1] The first description of an ABC was made by Van Arsdale (1893), and their name was given by Jaffe and Lichtenstein (1942). The incidence of ABCs is about 1% of all bone tumors, with 75% of patients being under 20 years of age.

^[2] The spine is involved in 3%–20% of all cases. ABCs are predominantly found in children and young adults and are most often diagnosed in the second decade of life, with a male-to-female ratio of 1/1.16.^[3]

Aneurysmal bone cysts present as benign, expansive lesions involving the subcortical-subperiosteal layers of the bone, changing its shape but rarely extending beyond the periosteal border. Removal of the bony cortex reveals honeycomb-like structures filled with blood. Aneurysmal bone cysts are made up of cavernous vascular spaces, covered in most cases with endothelium, but other structures characteristic of a vascular wall are not found. The vascular spaces are surrounded by fibrous septa. In some cases, the stroma fills more than half of the volume of the tumor mass, with the cystic part occupying a more limited volume.

ABCs can develop in any skeletal bone, but their most common localization is in the long tubular bones, followed by the spinal vertebrae, and less often in the flat bones of the skeleton. In the case of vertebral localization, the dorsal elements - pedicles, arches, and processes, are predisposed areas, but the body of the vertebra can also be affected, with a high risk of pathological fracture and neurological disorders of varying severity. The lumbar region is most commonly affected, followed by the thoracic and cervical segments.

Various pathophysiological theories for the occurrence of ABCs have been discussed. One of them suggests that an ABC is a response to vascular disorders, leading to increased intraosseous pressure, which causes bone expansion and destruction. According to other theories, an ABC's traumatic genesis is followed by an aberrant reparative process.^[4] DiCaprio et al. demonstrated familial inheritance.^[5]

The goal of current research on ABCs is to clarify the chromosomal translocations of the USP6 gene, which is thought to be in charge of the neoplastic process.^[6] USP6 is a ubiquitin-specific protease whose gene is localized on the short arm of chromosome 17 (17p13.2). USP6 plays an important role in the regulation of protein degradation, angiogenesis, and the inflammatory response. It is important to note that rearrangements in the USP6 gene are not unique to ABCs. They are also described in other benign entities, for example, nodular fasciitis, myositis ossificans, and fibro-osseous pseudotumors of the fingers, called by some authors USP6-related neoplasms.^[7] The USP6 assay is not universally available and is not routinely used in the diagnosis of ABC.

Spinal aneurysmal bone cysts are characterized by pain of varying intensity and symptoms of involvement of neural structures, with the development of neurodeficiency (e.g., paresthesias, hypesthesias, and, less often, movement

disorders). The clinical symptoms of cysts with vertebral localization develop with the characteristics of slowly evolving spinal cord compression, and the clinical manifestations depend on the level and degree of bone destruction and compression of the myelon and nerve structures (scoliosis, paresis, and radiculopathies). The disease's onset can be acute, resulting in severe neurological deficits and a pathological fracture. The rapid development of clinical manifestations in some patients usually leads to severe deficits and irreversibility of neurological symptoms. These patients only have a chance if they receive prompt and adequate surgical treatment.^[8]

On radiographs, aneurysmal bone cysts present as lytic, expansive, and lobulated lesions with a distinct sclerotic border. Computed tomography (CT) and magnetic resonance imaging (MRI) are the leading diagnostic imaging methods in spinal localization. These studies lead to visualization of an osteolytic lesion with expansive growth and the presence of multiple fibrous septa and fluid-fluid levels. In addition, they are indispensable in the assessment of the risk of a pathological fracture and support the preoperative planning of surgical interventions.

The 2020 WHO classification of bone tumors defines two nosological groups: aneurysmal bone cysts (ABCs) and ABC-like changes, as an evolution of other bone neoplasms. Bone tumors leading to ABC-like changes are mainly benign, e.g., chondroblastoma, fibrous dysplasia, giant cell tumors, and osteoblastoma, but they could also be malignant, e.g., osteosarcoma.^[9] Of particular importance is the differentiation of ABC from telangiectatic osteosarcoma, which is malignant in nature and can mimic ABC in all aspects. Confirmation of the diagnosis by biopsy with subsequent histological verification is mandatory for adequate treatment.

Many therapeutic options have been applied over the years in the treatment of ABCs, from aggressive interventions to radiation therapy and simple observation, as well as the minimally invasive techniques that have become widely used in recent years. Sclerotherapy with different sclerotic agents is a minimally invasive intervention that requires repeated procedures and could lead to good results. Some medical publications recommend sclerotherapy as a treatment method for ABCs with spinal localization.^[10] Another minimally invasive method in the treatment of ABCs is percutaneous thermoablation.^[11] Cases of spontaneous healing have also been reported.^[12] A conservative behavior may be preferred in some cases with small lesions and with a location that carries a low risk of pathologic fractures.^[13] Promising results are observed by using the relatively new therapy with the monoclonal antibody denosumab, inhibiting osteoclast activation and, respectively, bone resorption and remodeling.^[14]

Therapeutic approaches to spinal aneurysmal bone cysts are complicated by the risk of spinal instability and neurological complications.^[15] Compromising the stability and protective functions of the spine puts the spinal cord and cauda equina at risk of injury, with subsequent neurodeficiency and impaired quality of life, even death. These cir-

cumstances make the treatment of vertebral ABCs complex and require a multimodal approach. Treatment of ABCs generally includes selective arterial embolization, surgical resection of the lesion, and radiotherapy for residual tumor mass.^[16] It has been established that incomplete removal of the tumor mass does not necessarily lead to recurrence, therefore, radiotherapy is justified only in cases of recurrence. Feigenberg et al. believe that locally applied radiotherapy (20–36 Gy) is sufficient to solve the problem of local recurrence.^[17] With the leading role of radiotherapy in the treatment, the unfavorable outcome is a severe deformity of the spine, which is why the main guarantee for a good outcome remains surgery.^[18]

The standard treatment is operative treatment - maximal resection of the tumor mass, total if possible, with decompression of the nerve structures, and possible one- or two-stage spinal stabilization. Total excision, preferably en bloc, provides the highest curing rate^[19], while subtotal is associated with a high relapse rate. In total resection, the entire cyst wall should be removed, along with any abnormal spongy tissue and bony surfaces lined by fragile, hypervascularized membranes.^[20] The main operative approach is laminectomy, total or partial, which, with involvement of the vertebral body, remains insufficient and may require corpectomy and stabilization. Some authors recommend the implantation of titanium mesh filled with allogeneic bone marrow and vertebral body plaque^[21], while others recommend vertebral body fusion in young people to be performed with a bone autograft.^[8]

CONCLUSION

Spinal aneurysmal bone cysts are benign lesions that can cause various neurological disorders, depending on their size and location. There are various therapeutic options, but the standard treatment is operative - decompression, total resection and, if necessary, stabilization of the affected segment. The need for circumferential stabilization requires the intervention to be carried out in two stages. Radiotherapy is applicable to patients with relapses. Selective arterial embolization is a valuable method that reduces intraoperative bleeding.

Acknowledgements

The authors have no support to report.

Funding

The authors have no funding to report.

Competing Interests

The authors have declared that no competing interests exist.

REFERENCES

- Restrepo R, Zahrah Z, Pelaez L, et al. Update on aneurysmal bone cyst: pathophysiology, histology, imaging and treatment. *Pediatr Radiol* 2022; 52(9):1601–14.
- Rossi G, Rimondi E, Bartalena T, et al. Selective arterial embolization of 36 aneurysmal bone cysts of the skeleton with N-2-butylcyanoacrylate. *Skeletal Radiol* 2010; 39(2):161–7.
- Leithner A, Windhager R, Lang S, et al. Aneurysmal bone cyst: a population based epidemiologic study and literature review. *Clin Orthop Relat Res* (1976-2007) 1999; 363:176–9.
- Biesecker JL, Marcove RC, Huvos AG, et al. Aneurysmal bone cysts. A clinicopathologic study of 66 cases. *Cancer* 1970; 26:615–25.
- Di Caprio M, Murphy M, Camp R. Aneurysmal bone cyst of the spine with familial incidence. *Spine* 2000; 25(12):1589–92.
- Panoutsakopoulos G, Pandis N, Kyriazoglou I, et al. Recurrent t(16;17)(Q22;P13) in aneurysmal bone cysts. *Genes Chromosomes Cancer* 1999; 26:265–6.
- Oliveira AM, Perez-Atayde AR, Dal Cin P, et al. Aneurysmal bone cyst variant translocations upregulate USP6 transcription by promoter swapping with the ZNF9, COL1A1, TRAP150, and OMD genes. *Oncogene* 2005; 24:3419–26.
- Kanamiya T, Asakawa Y, Naito M, et al. Pathological fracture through a C6 aneurysmal bone cyst. Case report. *J Neurosurg* 2001; 94(2 suppl.):302–4.
- Sasaki H, Nagano S, Shimada H, et al. Diagnosing and discriminating between primary and secondary aneurysmal bone cysts. *Oncol Lett* 2017; 13:2290–6.
- Liu X, Han SB, Si G, et al. Percutaneous albumin/doxycycline injection versus open surgery for aneurysmal bone cysts in the mobile spine. *Eur Spine J* 2019; 28:1529–36.
- Park HY, Yang SK, Sheppard WL, et al. Current management of aneurysmal bone cysts. *Curr Rev Musculoskelet Med* 2016; 9:435–44.
- Malghem J, Maldague B, Esselinckx W, et al. Spontaneous healing of aneurysmal bone cysts. A report of three cases. *J Bone Joint Surg Br* 1989; 71:645–50.
- Cottalorda J, Chotel F, Kohler R, et al. Aneurysmal bone cysts of the pelvis in children: a multicenter study and literature review. *J Pediatr Orthop* 2005; 25:471–5.
- Lipplaa A, Dijkstra S, Gelderblom H. Challenges of denosumab in giant cell tumor of bone, and other giant cell-rich tumors of bone. *Curr Opin Oncol* 2019; 31:329–35.
- Desai SB, O'Brien C, Shaikh R, et al. Multidisciplinary management of spinal aneurysmal bone cysts: A single-center experience. *Interv Neuroradiol* 2019; 25(5):564–9.
- Palmisciano P, Hunter M, Lokesh N, et al. Aneurysmal bone cyst of the spine in adult patients: A systematic review and comparison of primary vs secondary lesions. *J Clin Neurosci* 2022; 100:15–22.
- Feigenberg S, Marcus R, Zlotecki R, et al. Megavoltage radiotherapy for aneurysmal bone cysts. *Int J Radiat Oncol Biol Phys* 2001; 49(5):1243–47.
- Boriani S, De Iure F, Campanacci L, et al. Aneurysmal bone cyst of the mobile spine: report of 41 cases. *Spine* 2001; 26(1):27–35.
- Ozaki T, Halm H, Hillmann A, et al. Aneurysmal bone cysts of the spine. *Arch Orthop Trauma Surg* 1999; 119:159–62.
- Gupta VK, Gupta SK, Khosla VK, et al. Aneurysmal bone cysts of the spine. *Surg Neurol* 1994; 42:428–32.
- Kinoshita A, Kataoka K, Taneda M. Multilevel vertebral body replacement with a titanium mesh spacer for aneurysmal bone cyst: technical note. *Minim-Invasive Neurosurg* 1999; 42(3):156–58.

Аневризматические костные кисты позвоночника – серия клинических случаев

Асен Цеков¹, Иван Тарев¹, Анастас Канев¹

¹ Клиника нейрохирургии, Аджибадем Сити Клиник, Университетская больница „Токуда“, София, Болгария

Адрес для корреспонденции: Иван Тарев, Аджибадем Сити Клиник, Больница „Токуда“, София, Болгария; Email: tarevmd@gmail.com

Дата получения: 21 февраля 2024 ♦ Дата приемки: 7 мая 2024 ♦ Дата публикации: 30 июня 2024

Образец цитирования: Cekov A, Tarev I, Kanev A. Spinal aneurysmal bone cysts – case series. Folia Med (Plovdiv) 2024;66(3):402-408. doi: 10.3897/folmed.66.e121395.

Резюме

Аневризмальные костные кисты – это доброкачественные неопластические образования, которые обильно васкуляризованы и иногда могут проявлять деструктивную и расширяющуюся активность. Они обычно поражают молодых людей, с меньшей частотой проявления у женщин, и чаще всего возникают во втором десятилетии жизни. Аневризмальные костные кисты составляют около 1 % всех опухолей костей и обычно развиваются в длинных трубчатых костях, при этом позвоночник поражается гораздо реже.

Мы представляем здесь три случая аневризмальных костных кист спинного мозга, которые были хирургически пролечены в отделении нейрохирургии в период с 2009 по 2022 год. Эти пациенты поступили в клинику с симптомами компрессии спинного мозга или сосудов. Объем оперативного вмешательства составил полную или субтотальную резекцию образования. Послеоперационного неврологического дефицита не наблюдалось, и ранних осложнений после вмешательств не зафиксировано. Все пациенты были выписаны из отделения с клиническим улучшением. Один перенёс повторную операцию через 4 года из-за рецидива кисты. Диагноз аневризматической костной кисты во всех случаях был подтверждён гистологическим исследованием.

Ключевые слова

аневризматическая костная киста, оперативное лечение, результаты
