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# Definitive Sclerosing Intensity Modulated Radiotherapy in A Child with Recurrent Aneurysmal Bone Cyst- A Clinical Case from our Practice

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**ABSTRACT**

Aneurysmal bone cysts (ABCs) are rarely diagnosed aggressive benign bone tumors. They are mainly diagnosed in children and adolescents in the long bones of the limbs, but in 12 %-30% are localized in the spine. Since the value of external beam radiotherapy (EBRT) for ABC has not been well defined, we present our observations and treatment outcome after one year from the definitive intensity modulated radiotherapy (IMRT) up to 30 Gy. for recurrent spine ABC in adolescent age. We present in dynamics the MR images of ABC after 3 months, after 6 months and after 1 year of the radiation treatment, analyzing the achieved tumor reducing and sclerosing effect. Our observations lead to the conclusion, that IMRT is an effective sclerosing treatment method for relapsed or inoperable disease, achieving not only a symptomatic pain-relieving effect, but also significant reduction of the bone tumor lesion.

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## Introduction

Aneurysmal bone cysts (ABC) are aggressive benign lesions [1]. Most cases have been identified in the first two decades of life; it is rare to find them after the age of 30 [2]. ABC is an expansive, lytic pseudo tumoral bone lesion composed of blood-filled spaces separated by connective tissue septa formed by reactive bone tissue, fibroblasts, and osteoclast-type giant cells [3]. The hemodynamic changes caused by the fistula lead to vasodilation and bone destruction, resulting in the formation of ABC [4]. Treatments have ranged from simple curettage, with or without bone graft, injection of a fibrosing agent, complete resection surgery, radiotherapy (RT) and selective arterial embolization [5]. We present our treatment results by means of the MRI study performed after three months, after six months and after one year after the completion of intensity modulated radiation therapy (IMRT) up to 30 Gy due to recurrent spinal ABC in adolescence.

## Clinical Case

Since September 2022, a 13-year-old boy presented with slowly progressive low back pain that spread to the posterior surface of the right leg. The pain was hardly affected by conservative treatment. After CT and MRI imaging studies and consultation with a neurosurgeon, the child was admitted for surgical treatment. From Spine **MRI/ November 2022**- Evidence of a tumor process on the right involving the body and right pedicle of the fifth lumbar vertebra (L5), exerting antero-lateral compression on the dural sac and on the nerve root at that level, with imaging features of aneurysmal bone cyst (ABC) (Figure 1).

**Intraoperative**

Percutaneous vertebroplasty was performed through the left pedicle at L5. A hemilaminectomy was performed at L5 and partial at L4. An extradural tumor formation was encountered from the body of L5 with ventral compression on the dural sac, concentrically involving the right root. The mass was soft, profusely bleeding, with the presence of multiple intradural hemorrhagic cysts and fibrous stroma at the periphery, with macroscopic features of a primary bone tumor of the body of L5. The tumor was subtotally resected, achieving wide decompression of the neural structures. Vertebroplasty of the bony tumor bed was performed.

**Pathohistological result/ 08.12.2022**- Fresh hemorrhages predominate in the examined material, followed by cystic spaces without clearly formed vascular walls. In their periphery, proliferation of mesenchymal cells and clustering of osteoclast-type giant cells can be seen. Single bone lamellae are found. The lesion is partially encapsulated. The diagnosis of ABC was confirmed. In January 2023, a follow-up MRI of the spine was performed, which established a recurrence of the disease with tumor size advancing -7.5/6/5.1 cm with compression on the spinal canal.

**Spine MRI /March 2023**- The inhomogeneous mostly high-signal T1 multicystic mass involving the body of L5 on the right with the corresponding peduncle, arch and transverse process persisted. The dimensions in anterior-posterior dimension are about 6.1 cm, transverse 7.8 cm and caudal-cranial 5.1, the advancement being at the expense of the transverse dimension / towards the body medially and to the left. The stenosis of the neuroforamen on the right persists as the corticalis is not disturbed. Conclusion - MRI

data for an advancing aneurysmal bone cyst (Figure 2). Considering the benign nature of the lesion and its volume, in April 2023, we performed performed intensity modulated radiotherapy (IMRT) using the VMAT method up to a total dose of 30 Gy (Figure 3). During RT, non-steroidal analgesic drugs were required, as the pain syndrome did not resolve until the RT end. 3 months after the completion of IMRT, we performed a MRI (Figure 4), which showed a significant reverse reduction of the aneurysmal bone cyst against the background of complete anesthesia without the use of pain medication.

**Spine MRI after 6 months from IMRT**-The structure of the formation is heterogeneous with confluent hyperintense areas in T2 and STIR, hypointense in T1, as well as reticular osteosclerotic areas. No detectable cortical erosion. Osteosclerotic material was noted intraspongeally, presented as low signal in all dimensions Conclusion- No data on significant dynamics of the mediated bone lesion (Figure. 5).

**Spine MRI after 1 year from IMRT**-The monitored heterogeneous volumetric hormation is presented without dynamics in its axial dimensions 4.3 / 7.9 cm with transitional 4.3 / 8.4 cm with some advancement of the cystic component. The formation is non-dynamic in distribution and involves the body of the fifth vertebra, swelling its right pedicle, lamina and facet joint. Causes stenosis of the corresponding recess and neuroforamen. The structure of the formation is markedly heterogeneous with cystic areas as well as reticular osteosclerotic areas. No detectable cortical erosion. No dynamics of the residual cyst with current dimensions of 2.1 cm projecting to the erector spinae muscle. The intervertebral disc at the L 4/5 level is slightly elongated, with a right eccentric disc protrusion foraminal homolaterally - no dynamics. Level L5/S1- no significant disc pathology, free neuroforamina. The body of the fifth lumbar vertebra is presented wedge-shaped down n dorsal aspect with deformation to the right. Osteosclerotic material intraspongiously, presented as low signal in all dimensions. Built lumbar lordosis, without evidence of spondylolisthesis. Conclusion- No data on significant dynamics of the mediated bone lesion (Figure 6).

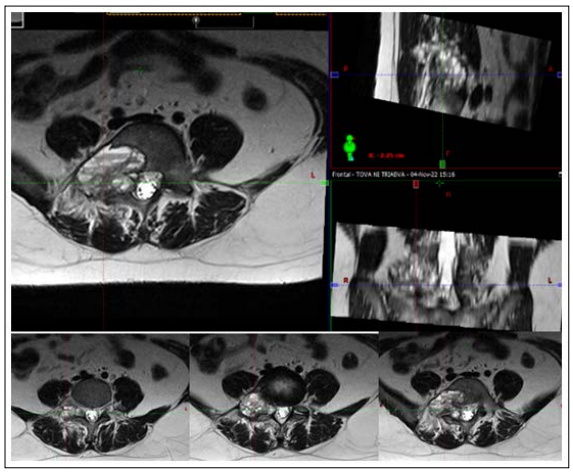

**Figure 1:** Spine MRI/ November 2022

Evidence of a tumor process on the right involving the body and right pedicle of the fifth lumbar vertebra (L5), exerting antero-lateral compression on the dural sac and on the nerve root at that level, with imaging features of aneurysmal bone cyst

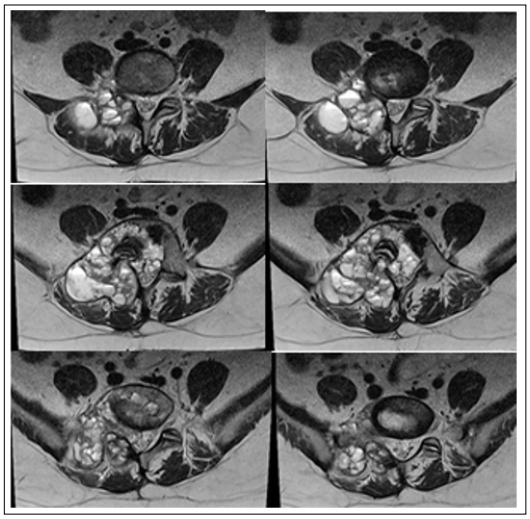

**Figure 2:** Spine MRI /March 2023

The inhomogeneous mostly high-signal T1 multicystic mass involving the body of L5 on the right with the corresponding peduncle, arch and transverse process persisted. The dimensions in anterior-posterior dimension are about 6.1 cm, transverse 7.8 cm and

caudal-cranial 5.1, the advancement being at the expense of the transverse dimension / towards the body medially and to the left. The stenosis of the neuroforamen on the right persists as the corticalis is not disturbed.

**Conclusion**

MRI data for an advancing aneurysmal bone cyst.

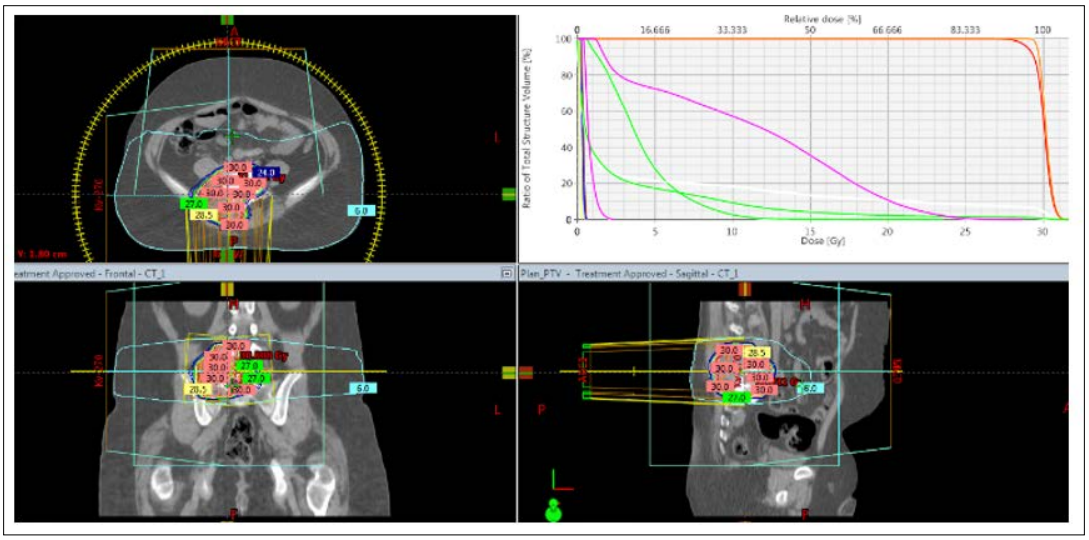


**Figure 3:** Intensity Modulated Radiotherapy (IMRT) using the VMAT method up to a total tumor dose 30 Gy with a daily dose 2 Gy

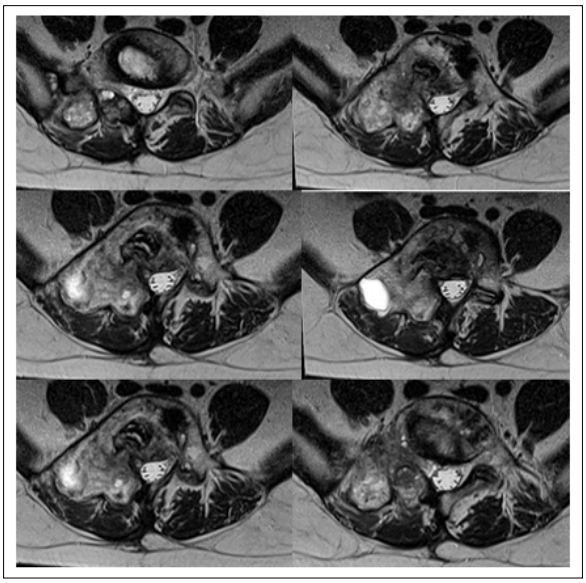

**Figure 4:** The Follow-Up MRI 3 Months after the Completion of IMRT

The tracked heterogeneous volume formation is presented with a significant reverse development and reduction of a large part of the cystic bone components with a lack of sediment among them in the form of levels. The bone structure is heterogeneous, with confluent T2 and STIR hyperintense areas, without cortical erosion. Residual cyst lateral to the bulging bony structure of L5 measuring 1.5/1.9 cm.

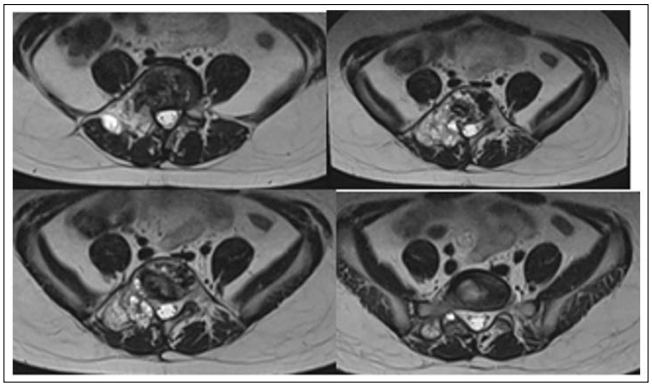

**Figure 5:** The Follow-Up MRI 6 Months after the Completion of IMRT/ November 2023

The structure of the formation is heterogeneous with confluent hyperintense areas in T2 and STIR, hypointense in T1, as well as reticular osteosclerotic areas. No detectable cortical erosion. Osteosclerotic material was noted intraspongeally, presented as low signal in all dimensions. Conclusion- No data on significant dynamics of the mediated bone lesion.

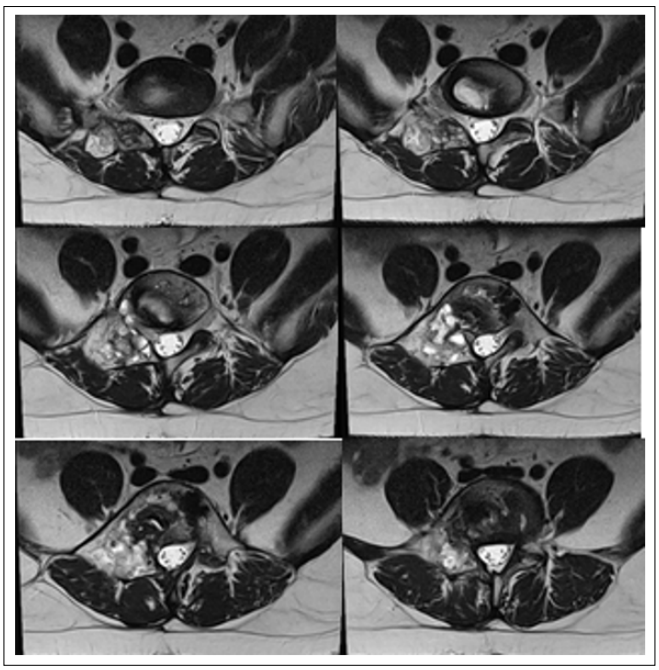

**Figure 6:** Spine MRI after 1 Year from IMRT

The monitored heterogeneous volumetric hormation is presented without dynamics in its axial dimensions 4.3 / 7.9 cm with transitional 4.3 / 8.4 cm with some advancement of the cystic component. The formation is non-dynamic in distribution and involves the body of the fifth vertebra, swelling its right pedicle, lamina and facet joint. Causes stenosis of the corresponding recess and neuroforamen. The structure of the formation is markedly heterogeneous with cystic areas as well as reticular osteosclerotic areas. No detectable cortical erosion. Conclusion- No data on significant dynamics of the mediated bone lesion.

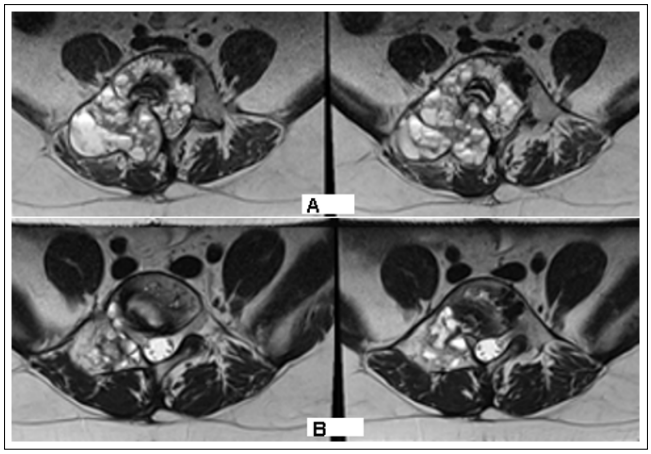


**Figure 7:** A/ Comparison of MRI Before IMRT/ a and after 1 Year from IMRT/B

The monitored heterogeneous volumetric hormation is presented with dynamics in its axial dimensions from 6.,1/7.8 cm up to 4.3 / 7.8 cm with some advancement of the cystic component.

**Discussion**

ABC is currently classified as a benign osteoclastic giant cell-rich tumor on the 2020 WHO Classification of Tumors of Bone [6]. It is a highly vascularized lesion composed of cavities of blood content, separated by connective tissue septa, and is surrounded by a layer of cortical bone, with the potential ability to expand [7]. ABC are usually expansile blood-filled cavities within the bone lined by proliferative fibroblasts, giant cells, and trabecular bone [8]. The spine ABC represent 15% of primary bone tumors [9]. Because there are few reported aggressive case reports of spinal ABC, diagnostic and treatment algorithms remain controversial and variable [10]. Common presenting symptoms of ABC are ill-defined somatic pain, stiffness, and swelling with associated bone destruction and pathologic fracture [11]. Our 13-year-old patient presented with slowly progressive low back pain radiating to the posterior surface of the right leg. The pain was almost unaffected by medication, which required a number of imaging studies to clarify the pain syndrome. On MRI presence of multiloculated fluid-fluid filled cysts is characteristic of ABC. Still, it can be confused with other types of lesions such as osteosarcoma or giant cell tumors, so biopsy of the lesion is necessary to obtain a definitive diagnosis [12]. In the presented patient, the aneurysmal bone cyst was diagnosed in September 2022, and after subtotal surgery, progression was reported with a typical MRI image including the tumor with sharp contours and a characteristic multicystic septate structure with formed liquid-liquid levels. ABC causes ventral and lateral stenosis of the spinal canal with dural compression and obliterates the corresponding root canal (Figure 2). The treatment of ABCs continues to be a matter of controversy, due to the lack of defined guidelines. Multiple options have been described, to be used alone or in combination, including curettage with or without bone grafting, complete tumor resection, selective preoperative embolization, radiotherapy (RT), chemotherapy, and intralesional injections [13-17]. Radiotherapy has been used historically to treat ABCs primarily, as adjuvant therapy in cases of recurrence, and inoperable ABC lesions [11]. Feigenberg et al. advocated RT to nine patients with recurrent (4 patients) or inoperable ABC (5 patients). The dose prescribed ranged from 20-60 Gy [18]. Radiotherapy is an option currently reserved for patients at high risk of not withstanding surgery or for those who are resistant to surgical treatment, even more so considering the potential risks of post-radiation myelopathy or sarcomatous transformation [19].

There are many hypotheses regarding the growth of the aneurysmal bone cyst and its malignancy after radiation therapy. Malignant bone lesion development at the site of a previously treated bone cyst without a history of radiotherapy is a rare clinic event with only five patients reported in the literature [20-23]. Although the role for malignant transformation has not been firmly clarified, it may be comparable with that a malignant lesion occurring within an area of bone infarction [24-26]. The unbalance in the remodelling process between progenitor cells and the scaffold which provides three-dimensional support was correlated with spontaneous malignant transformation of the progenitor cells [27]. Considering the benign nature of the lesion and its volume, we performed intensity modulated radiotherapy (IMRT) using the VMAT method up to a total dose of 30 Gy (Figure 3 ).The follow-up MRI 3 months after the completion of IMRT in juli 2023, presented with a significant reverse development and reduction of a large part of the cystic bone components with a lack of sediment among them in the form of levels. The bone structure is heterogeneous, with confluent T2 and STIR hyperintense areas, without cortical erosion (Figure 4). Comparative analysis of the post-radiation MRI at the 6th month and at the first year after the radiation completion up to a total tumor dose 30 Gy reported a lack of dynamism in lesion sizes. The structure of the ABC is markedly heterogeneous with cystic areas as well as reticular osteosclerotic areas. No detectable cortical erosion (Figure 5 and Figure 6). Figure 7 presents the significant reduction of the bone lesion and its ossification one year after completion of radiotherapy compared to the pre-radiation status of the recurrent lesion. Given the data on malignant transformation of ABC, in the presence of pain symptoms in childhood after radiotherapy, re-irradiation of the residual lesion is not applied. It is known that radiation treatment in childhood is carried out with lower total doses compared to patients in adulthood and old age, precisely because of the increased risk of the appearance of a secondary malignant disease many years after irradiation even with small radiation doses.

## Conclusion

Aneurysmal bone cysts (ABCs) are rarely diagnosed aggressive benign bone tumors in childhood. Radiotherapy is an option currently reserved as adjuvant therapy in cases of recurrence, and inoperable ABC lesions. With regard to the observed MRI changes in the ABC after 3 months from the completion of RT, we report a significant reduction of the bone lesion accompanied by sclerosing effect.. After 6 months and after one year, the MR finding is stationary and without significant dynamics. In case of a new-onset neurological pain syndrome, do not repeat radiotherapy in childhood, as there is a risk of secondary oncogenesis. The pain is controlled after consultation and recommendations from a pediatric neurologist and a pediatric orthopedist.

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