

# A Giant Recurrent Aneurysmal Bone Cyst of the Sacrum: Discussion of Total Resectability in a Pediatric Case

## Sakrumda Tekrarlayan Büyük Anevrizmal Kemik Kisti: Pediatrik Bir Vakada Total Çıkarılabilirliğinin Tartışılması

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**ABSTRACT** Aneurysmal bone cyst (ABC) is a locally aggressive tumor-like lesion of the bone. Sacral location is rare. Currently recommended treatment for a benign ABC lesion is complete intralesional curettage with or without selective arterial embolization and attempt to salvage unilateral sacral 2-5 roots. A 14-year-old girl presented with severe pain and a large mass in her lower lumbosacral region. Imaging studies demonstrated a destructive lesion, arising from the second sacral vertebral level, extending posteriorly and bilaterally into the gluteal muscles, and anteriorly into the pelvis. Open biopsy established the diagnosis of ABC. Marginal extra-capsular resection via a combined anterior and posterior approach, and postoperative radiation therapy were performed. The patient was pain-free, and no evidence of local recurrence was detected at the 84-month follow up postoperatively. Marginal extra-capsular resection and adjuvant radiotherapy are thought to be effective for the treatment of this large ABC of the sacrum.

**Key Words:** Bone cysts, aneurysmal; sacrum

**ÖZET** Anevrizmal kemik kisti (AKK) kemiğin tümör benzeri, lokal olarak agresif seyreden bir lezyonudur. Sakral bölgede yerleşimi nadirdir. Günümüzde iyi huylu bir AKK için önerilen tedavi selektif arteriyel embolizasyon ve tek taraflı 2-5 sakral kökün kurtarılmaya çalışılması ile birlikte ya da tek başına komple intralezyonel küretajdır. Ondört yaşında bayan hasta alt lumbosakral bölgede geniş bir kitle ve şiddetli ağrı şikayeti ile başvurdu. Görüntüleme yöntemleri ile ikinci sakral vertebra seviyesinden çıkan, arkada her iki tarafta gluteal kaslara önde de pelvise kadar uzanan destrüktif bir lezyon tespit edildi. Açık biyopsi ile AKK tanısı kondu. Marjinal ekstra-kapsüler rezeksiyon ve operasyon sonrası radyasyon tedavisi uygulandı. Operasyon sonrası 84 aylık takipte hastanın ağrısı yoktu ve lokal rekürens ile ilgili herhangi bir bulgu saptanmadı. Sakrumda yerleşmiş geniş AKK tedavisinde önden ve arkadan birlikte ekstra-kapsüler rezeksiyon ve adjuvan radyoterapinin efektif olduğu düşünülmektedir.

**Anahtar Kelimeler:** Kemik kistleri, anevrizmal; sakrum

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**A**neurysmal bone cyst (ABC) is an aggressive tumor like lesion of the bone. It may arise from any bone and constitutes 6% of all bone tumors.<sup>1-4</sup> Spinal involvement has only been reported in case reports or short series in the literature.<sup>2,4-6</sup> Sacral location is even rarer.<sup>2,7,8</sup> Despite the benign histopathology of ABC, its clinical course is locally aggressive, and the recurrence rate is high.<sup>1,2</sup> Currently recommended treatment for spinal ABC is complete intralesional curettage with or without selective arterial embolization and attempt to salvage unilateral sacral 2-5 roots.<sup>5,8</sup> The authors herein present a very rare, giant, recurrent ABC, originating from

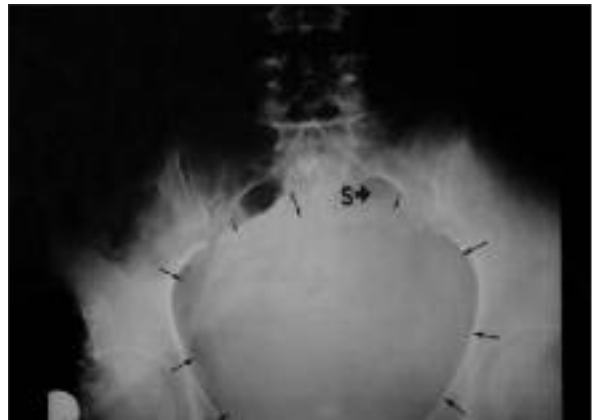
the second sacral segment, which was treated successfully by a combined anterior and posterior surgery and an additional adjuvant radiotherapy.

## CASE REPORT

The family of the patient gave informed consent for their child to be presented. A 14-year-old girl presented with buttock pain and a mass over the sacrum. In spite of the persisting symptoms following an intralesional excision performed in another center one and a half years ago, she received no further treatment. At the time of her first visit to our institution, the patient was not able to sit and lie in supine position because of the large mass. Physical examination revealed a poorly localized, tender and soft mass of 30 x 30 x 20 cm, over the sacrum, extending bilaterally and posteriorly (Figure 1). The anus was displaced, and the overlying skin was indurated and warm. Bowel and urinary functions were normal and lower extremity neurologic functions were intact, but she had saddle type anesthesia. Plain radiographs showed a large and ill defined lytic lesion, originating from the second sacral vertebra, distally (Figure 2). The first sacral foramens and bilateral sacroiliac joints were spared. Computed tomography and magnetic resonance imaging confirmed the location of the tumor, and demonstrated its anterior and posterior extension. The tumor with fluid levels and multiple septa formation, compressed the bladder and the uterus, and was shown to adhere to the sigmoid colon (Figures 3, 4). Open biopsy established the di-

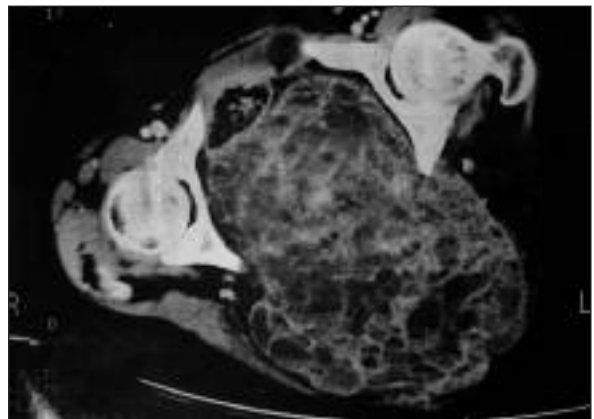


**FIGURE 1:** Preoperative photograph showing the size of the mass.



**FIGURE 2:** X-ray showing a large, expansile mass, arising from second sacral vertebra.

S; first sacral foramen.



**FIGURE 3:** CT image of the mass with multiple septa formation and fluid levels.

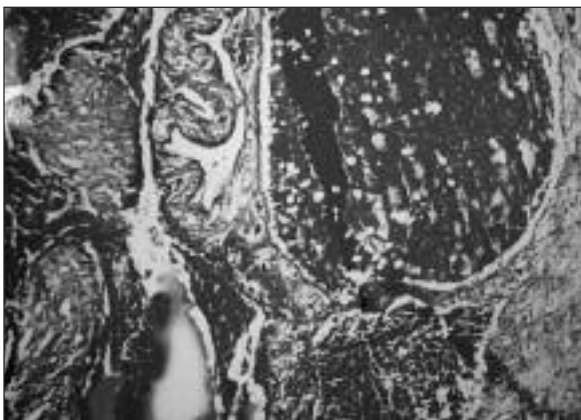
agnosis of ABC (Figure 5). Resection of the tumor through a combined anterior and posterior approach was performed. No local recurrence was encountered at 84 months of follow-up (Figure 6).

## SURGICAL TECHNIQUE

Following the necessary preoperative measures, surgery was performed through an extended Phannestiel approach. The rectus abdominis muscles were dissected, and the lateral abdominal wall muscles were incised parallel to the skin incision. After opening the visceral peritoneum and retracting the small bowel, the anterior side of the lesion was fully identified, along with the aorta and the inferior vena cava. Then, internal iliac arteries and veins were ligated bilaterally. The capsule of the tumor was very thin, therefore, to prevent un-



**FIGURE 4:** Anterior and posterior extension of the mass in magnetic resonance images.



**FIGURE 5:** Histopathological appearance of the tumor.

controlled hemorrhage in case of perforation, two small incisions into the lacunae of the lesion were performed, and the blood was aspirated. After the evacuation, the lesion shrank, and it became easier to detach it from surroundings. The adhesions to the sigmoid colon, the inner wall of the pelvis, the uterus, the tubes and the ovaries were freed from

the lesion, without perforating the capsule. After the lesion was fully detached from the viscera, anterior cortical osteotomy below the first sacral foramen was performed with the help of fluoroscopy. A temporary colostomy was performed and the wound was closed. Thereafter, the patient was placed in prone position, and a midline longitudinal incision from the level of the fifth lumbar vertebra to the anus was performed. The gluteal muscles were marginally detached from the lesion. The S1 and S2 roots were identified. More distal roots including the distal part of S2 roots were buried within the tumor mass, and found to lose continuity. Therefore, the dural sac distal to the first sacral roots, was ligated and transected. A posterior sacral osteotomy was performed at the second sacral vertebral level, and it was connected to the anterior osteotomy. The mass was then totally mobile and free, and it was removed totally. To prevent posterior displacement of the pelvic viscera, a polypropylene mesh graft was sutured between the



**FIGURE 6:** No evidence of local recurrence in MR images at the last follow-up.

gluteal muscles. Six samples were taken from different areas, for the assessment of the surgical margins. The duration of the operation was six and half an hours. A total of 17 units of whole blood, 13 units intraoperatively and 4 units postoperatively was transfused. The patient was mobilized on the third, and discharged on the eighth postoperative day. There were no early or late surgery-related complications. Four weeks postoperatively, the patient was given 30 cGy of radiotherapy. The colostomy was closed at the postoperative third month. At the postoperative 84 months of follow up, there was no clinical or radiological (including X-ray, CT, and MRI) evidence of local recurrence (Figure 6) and moreover, no dysfunction of the bowel or bladder was noted. The area of saddle type anesthesia which was present preoperatively got smaller but was still continuing.

## DISCUSSION

Sacral tumors are often diagnosed late. The mass is difficult to notice on general musculoskeletal examination, because it tends to expand anteriorly into the pelvic cavity. In addition, symptoms are usually nonspecific and develop insidiously in months to years. Although, they are often readily palpable on rectal examination, they are usually discovered during imaging studies. The tumors, once identified, are usually large and extensively invade the sacral elements.

ABCs are benign, rapidly growing, destructive tumors of bone. Sacral location is exceedingly rare.<sup>2,7,8</sup> Huvos<sup>4</sup> reported 1% of sacral involvement in his series of 394 ABC cases. Campanacci<sup>2</sup> in his large series of ABC, reported only two, located in the sacrum.

Preoperative embolization is recommended in hypervascular tumors such as ABC to reduce tumor size and the risk of intraoperative hemorrhage.<sup>5,8,9</sup> Boriani et al.<sup>5</sup> going further, stated that, embolization should be the first choice for spinal ABC. However, embolization may not always be appropriate for all sacral tumors, therefore, bilateral ligation of the internal iliac vasculature and luncular evacuation are recommended for very large and hypervascular tumors.<sup>9,10</sup> Preoperative selec-

tive embolization should be the first choice in our case, following histopathologic confirmation of the diagnosis, but unfortunately it was not available in our center at that time.

Recurrence rates after intralesional resection are as high as 30-60%, leaving marginal extra-capsular resection the only way of treatment to reduce the risk of recurrence.<sup>8,11</sup> Since, the sacrum does not fit well to the compartmental concept, and comprise neural elements, a marginal extra-capsular resection is not usually possible, and complex surgery is required to provide tumor-free tissue.<sup>11-14</sup> When marginal extra-capsular resection is not possible, a complete, intralesional excision without sacrificing any neurological structures, combined with adjuvant therapy is recommended.<sup>8,11</sup> However, the possibility of local recurrence, resulting from inadequate resection, even after embolization and radiotherapy still exists. Subtotal tumor debulking and preservation of neural structures is found more appropriate in patients with metastatic disease to preserve quality of life.<sup>15</sup> On the other hand, radical resection of the tumor with sacrifice of the neural elements below the L5 nerve root has been advocated for aggressive benign or potentially curable malignant tumors involving the sacrum.<sup>15</sup>

Sacral resection can be performed through an anterior, a posterior, or a combined approach depending on tumor location, size and type. Posterior approach is preferred when the tumor is located at a distal sacral segment, or, the posterior part of the proximal sacrum.<sup>8,11-13</sup> Instead, a combined anterior and posterior procedure is indicated, when there is a large presacral mass involving the S1 segment, or a total resection of the sacrum is required.<sup>8,11,13</sup> Vascular control is also improved with this approach.<sup>15</sup> We found combined approach very effective for the marginal extra-capsular resection in this case, though it is a demanding procedure, with high perioperative mortality and morbidity.

An important consideration is the involvement of the sacral nerves, which control bowel and bladder function. Autonomic and somatic nerve supplies to the rectum, anal canal and urinary blad-

der travel through the sacral nerve roots S2 to S4. Gunterberg and coworkers<sup>16,17</sup> and recently, Todd et al.<sup>11</sup> showed that unilateral loss of sacral nerves did not affect overall bowel and bladder function, whereas, bilateral sacral nerve resections below S2 caused bowel and bladder dysfunction, in accordance with the previous literature. Marginal extra-capsular resection must be assessed carefully from the standpoint of neurologic deficit versus local tumoral control in the sacropelvic area, where recurrence may initiate a long series of inadequate procedures that will result in pelvic obstruction, gynecologic problems, spontaneous bleeding and death. With this in mind, and also taking into account the preoperative findings and intraoperative condition of S2 and more distal roots, we performed ligation and transection of the dural sac distal to the second sacral roots.

Radiotherapy is an effective adjuvant with favorable results in the treatment of ABC.<sup>2,17,18</sup> It must be kept at the minimum effective dose (usually 30-50 cGy recommended) in order to decrease the risk of radiation-induced sarcoma.<sup>18</sup> Accordingly, 30 cGy adjuvant radiotherapy was given to our patient. S1 and S2 roots should absolutely be preserved bilaterally. Although it was reported that unilateral loss of sacral nerves did not affect overall bowel and bladder function,<sup>11</sup> bilateral nerve resections below S2 did not cause any bowel or bladder dysfunction in our case.

In conclusion, marginal extra-capsular resection through a combined anterior and posterior surgery, and adjuvant radiotherapy provided a successful outcome in this recurrent, huge ABC of the sacrum.

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