Abstract Background: Aneurysmal bone cysts, first described by Jaffe and Lichtenstein in 1942, are benign lesions that may easily be mistaken for a malignant tumor both radiographically and pathologically. These diagnostic problems are due to their rapid growth, extensive destruction of bone, wide extraosseous tumor masses, and marked cellular exuberance. The differential diagnosis of aneurysmal bone cysts including giant cell tumor, calcified solitary bone cysts, low-grade osteosarcoma, and telangiectatic osteosarcoma becomes even more complicated when the lesion arises at sites other than the long bones and presents with extensive extraosseous tumor masses. The latter cases – especially when they occur as sacral or presacral tumors – present challenges with respect to successful treatment, which should combine surgical removal of the entire lesion following oncological criteria to prevent recurrences and osteosynthesis to guarantee the biomechanical stability of the spinal-pelvic junction. Here we report on the clinical case of a female patient with an aneurysmal bone cyst of the sacrum and extensive extraosseous tumor masses. The current report documents the diagnostic and surgical challenge of a gigantic aneurysmal bone cyst of the sacrum and its successful management.

Keywords Aneurysmal bone cyst · Sacral tumors · Pelvic surgery · Bone tumor

Introduction

Aneurysmal bone cysts are benign although locally destructive lesions that have frequently been reported to have a high risk of local recurrence [10, 13, 14, 19, 20, 22]. Rates of local recurrence after primary treatment – other than en bloc excision – have been reported ranging from 12% (4 of 34 patients after curettage) [10] to 31.5% (32 of 105 patients irrespective of the therapeutic regime) [19], or even as high as 59% in a series of 44 patients treated by curettage alone, with or without bone grafting (26 of 44 patients) [14]. The definitive etiology of aneurysmal bone cysts still remains to be defined. Jaffe’s original consideration of the possibility of a hemorrhagic ‘blow-out’ engrafted on a pre-existing lesion [11] was used by Biesecker and colleagues [2] as the basis for their hypothesis. They suggested that aneurysmal bone cysts are arteriovenous malformations associated with other primary bone lesions (32% according to Biesecker et al. [2]; 40% according to Mirra [15]) that might lead to elevated vascular pressures. Dr. Mirra has beautifully summarized these theoretical considerations regarding the nature of aneurysmal bone cyst as well as the corresponding consequences for their appropriate treatment: ‘...ABC is not a neoplasm in any valid sense, but most likely an anomalous, reparative process stubbornly resistant to elimination by simple curettage alone.’ This does not preclude the problem and the necessity that a possibly associated lesion, e.g., osteosarcoma or chondroblastoma, has to be identified and treated appropriately.

Here we report on the clinical case of a female patient with an aneurysmal bone cyst of the sacrum and extensive extraosseous tumor masses. The current report documents the diagnostic and surgical challenge of a gigantic aneurys-
Case Report

A 17-year-old woman who recognized an increase in her abdominal girth presented to a local general surgeon because of moderate abdominal pain, which had been unrelieved over a period of 4 weeks. In addition, she complained about intermittent weakness of her right leg. Ultrasound examination revealed a cystic-solid tumor extending from the abdominal wall to the sacrum. Subsequent MRI demonstrated a 16×12×14 cm extraosseous tumor with expansive growth that filled the small pelvis and had dislocated the right pelvic vessels and the ureter to the left, as seen also on the intravenous pyelogram. Complete lytic destruction of the massa lateralis of S1 and S2 as well as partial erosion of L5 was noted but was ascribed to expansive growth of a soft-tissue tumor. The patient was then admitted to our clinic in November 1999 with the working diagnosis of a malignant retroperitoneal tumor, thought to be arising from the right ovary.

CT was performed to delineate the osseous destruction. This study showed that the tumor was accompanied by extensive lysis of the right sacrum compromising the biomechanical integrity of the spinal-pelvic junction and extending to the right neuroforamina and sacral nerve roots, respectively. CT as well as MRI demonstrated internal septation with some faint traces of mineral within the tumor but without a calcified rim at the periphery (Fig. 1). Based on these radiological and clinical data, our preoperative diagnosis was an aneurysmal bone cyst of the sacrum. When classified according to the radiographic portion of Enneking’s staging system for benign tumors [8], the present case represents an invasive stage 3 lesion characterized by an ill-defined border, incomplete to missing reactive bone margins, and cortical destruction with soft-tissue extensions.

To confirm the diagnosis histologically, an open biopsy was performed through an explorative median laparotomy. Histology and macroscopy presented the characteristic signs of an aneurysmal bone cyst — in this case arising from the sacrum — with extensive cavernomatous spaces and huge extraosseous tumor masses. The walls were composed of fibrous tissue with thin strands of osteoid and woven bone partially covered by active osteoblasts. Formation of cartilage with a faint chondroid matrix is found in some

Fig. 1 A Plain X-ray and intravenous pyelogram present a lytic destruction within the right sacrum with an extensive soft-tissue shadow and dislocation of the right ureter to the left. B MRI demonstrated a huge extraosseous tumor with internal septation. The size of the tumor is 16×12×14 cm, and it fills the small pelvis and dislocates the right pelvic vessels to the left. C Axial CT shows complete lytic destruction of the right massa lateralis of the sacrum as well as faint traces of mineral that represent the slivers of woven bone shown in histology (Fig. 2). D The axial slice demonstrates a well-perfused tumor that — when compared with B — indeed leads to nearly complete occlusion of the inner pelvic ring.