THE PUBLISHING HOUSE
OF THE ROMANIANACADEMY

Supplement 1/2015, 4th ISAA

GIANT SACRAL TUMOR WITH INTRAFORAMINAL EXTENSION AND INVASION IN THE LOWER PELVIC APERTURE

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Accepted November 14, 2015

Sacral tumors are rare and encompass numerous lesions. Primary malignant sacral tumors associate a poor prognosis. Aggressive surgical resection with surgical bed radiation and optional adjuvant chemotherapy is the mainstay of treatment, requiring even in advanced stages a multidisciplinary approach, involving neurosurgeons, vascular, colorectal, and plastic surgeons, oncologists and radiation oncologists. We present the case of an 59 years old male patient, diagnosed with a sacral tumor 6 years ago, admitted for L3 level cauda equina syndrome, sphincter dysfunction for 3 weeks, tailbone pain radiating posterior of both legs into his feet. The patient was diagnosed with giant sacral tumor with intraforaminal extension and lower pelvic aperture invasion. Anterior transabdominal and posterior sacral approaches were combined for partial tumor resection. Postoperative, complications were genital and buttock numbness, loss of bladder function, sexual function and bowel continence and deep vein thrombosis. There was no radiculopathy with excellent lower extremities strength. Operative bed radiation therapy was added. Most sacral tumors are diagnosed with extension in the lesser pelvis, invading neighboring structures, requiring aggressive surgical approach in an mixt interdisciplinary team. Total or subtotal sacrectomy should only be performed in experienced centers, in order to associate acceptable rates of morbidity and mortality.

*Keywords***:** sacral tumor, treatment, surgery, interdisciplinary

INTRODUCTION

The sacral pathology includes congenital lesions, inflammatory masses, neurogenic and osseous tumors, metastatic lesions, and others such as aneurysmal bone cysts and sarcomas. Primary malignant sacral tumors are rare[1]. They represent between 32 and 71% of all primary spinal tumors[2,3]. The majority of primary malignant tumors of the sacrum are low-grade malignancies like chordoma and chondrosarcoma, characterized by slow growth and long standing nonspecific initial symptoms[4]. In contrast, high-grade neoplasms like Ewing sarcoma, osteosarcoma may have a more aggressive and dramatic presentation[5]. In order to provide an effective treatment for these rare conditions, a multidisciplinary team must be selected, which includes oncologists, radiologists, radio-therapists, general surgeons and spine surgeons[6]. The fundamental step of the diagnostic process (preceded by proper clinical, morphological and functional studies, including laboratory tests) is the biopsy[7]. After establishing the pathological diagnosis, the team should plan a multimodular-, histotype- and patient specific treatment[6]. Aggressive surgical resection is the mainstay of treatment followed by radiation of the surgical bed and sometimes adjuvant chemotherapy[8]. The local recurrence rate after surgery is relatively high, due to the complex anatomy of the sacral region and the difficulty in achieving a complete resection, with tumor free margins[9].

MATERIAL AND METHODS

We present the case of a 59 years old male patient, admitted in *"Bagdasar Arseni"* Emergency Hospital in March 2015 for cauda equina syndrome with L3 level, Frankel C score, sphincter dysfunction for 3 weeks (with long-term urinary catheter), aching pain in his tailbone radiating down the posterior aspect of both legs into his foot. The patient was first diagnosed in 2009 when he presented to the hospital reporting low back pain. A MRI was performed at that time, showing specific signs for chondrosarcoma. The patient left the hospital refusing further investigations and treatment neglecting his disease for 6 years. At admission in our hospital, the patient reported weight loss, general weakness. He also suffered from type II diabetes with diabetic nephropathy, hypertension and stage III chronic renal disease.
Magnetic resonance imaging of the lumbar spine and sacrum compared with the initial examination showed a massive lombo-sacral tumor, of 82/82/135 mm, from L5 to S4, involving nerve roots and extending into the sacrum and neural foramen, with invasion in the lower pelvic aperture, with specific MRI signs for

chondrosarcoma (Figure 1). The imaging diagnosis techniques were completed by computerized tomography of the spinal cord and sacrum, with reconstruction, furthermore establishing the pelvic invasion, with an imprecise limit between the posterior rectal wall and the tumoral mass.

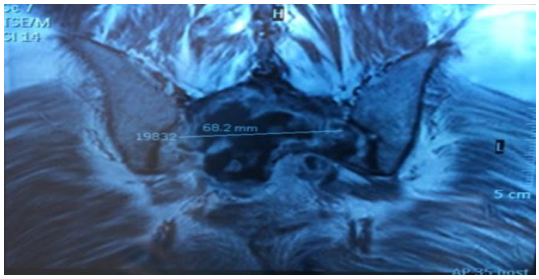

Figure 1 Imaging of the lumbar spine and sacrum

RESULTS AND DISCUSSIONS

Biological samples showed anemia and hypoalbuminemia. Clinical and laboratory investigations led to the diagnosis of giant sacral tumor with intraforaminal extension and invasion in the lower pelvic aperture. The patient was evaluated by a multidisciplinary team formed by a radiologist, an oncologist, a general surgeon, a neurosurgeon, a plastic surgeon, an orthopedic surgeon and a pathologist. Taking into account the pelvic invasion, a surgical intervention, in a mixt interdisciplinary team formed by a general surgeon and a neurosurgeon was indicated. The purpose was obtaining a histological diagnosis in order to be able to further conduct a specific oncological treatment and decompressing L5, S1, S2 roots, bilaterally. The neurosurgeon established no possibility of neurological recovery due to the extremely advanced stage. Anterior transabdominal and posterior sacral approaches have been combined to partially resect the tumor. Using an anterior approach, the general surgeon separated the tumor from the rectum. Then, the neurosurgeon performed a L5-S3 intralesional resection. The total operative time was 7 hours. Postoperative, as expected, the patient had genital and buttock numbness. He had lost bladder function and required self-catheterization. Sexual function was lost, as well as bowel continence. There was no radiculopathy and he had excellent strength in his lower extremities. His postoperative course was complicated by deep vein thrombosis, which was treated with oral anticoagulation therapy. As far as neo-adjuvant treatment in concerned, the patient underwent radiation treatment of the operative bed.

Clinical appearance and diagnosis

The clinical presentation of a sacral tumor is dependent on the anatomical localization of the lesion within the sacrum and influenced by the aggressiveness of the neoplasm[10]. The patient often report low back or buttock pain or may present with a visible sacral mass.

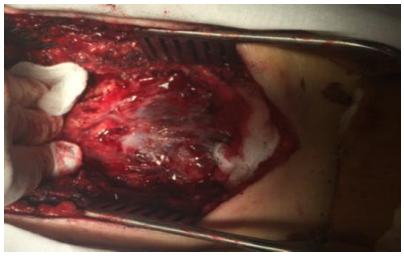

Figure 2 Posterior sacral approach

The patient may also present with neurological symptoms, but numbness, loss of sensation, decreased reflexes, sphincter dysfunction or motor deficit can be also the first sign[5]. A sacral tumor may determine the development of cauda equine syndrome requiring urgent surgical intervention[11]. The imaging diagnosis techniques performed on the sacral region include from plain radiography, with limited sensitivity, computerized tomography (CT) and magnetic resonance imaging (MRI) to scintigraphy and SPECT[9,12]. Some sacral tumor types have specific CT or MRI signs, the imaging process being the only appropriate method to describe the anatomic situation and the dimensions of the tumor, and to provide a definitive diagnosis. However, final diagnosis of primary sacral tumor can be made after an accurate histological examination, open biopsy representing the gold standard for the diagnosis of bone lesions, with 98% accuracy[13,14].

Non-surgical therapy Majority of primary sacral tumors, including chordoma and chondrosarcoma, are relatively resistant to the conventional radio- or chemotherapy, although radiotherapy can be used as an adjunctive treatment in case of intralesional surgical resection[5]. Chemotherapy is not effective in chondrosarcoma[15,16], but may be used in treating high-grade primary malignant sacral tumors, like Ewing sarcoma and osteosarcoma, followed by surgical intervention[5].

Surgical approach Resection of sacral tumors can be performed from anterior, posterior, lateral and combined surgical approaches, the decision in the use of one approach or another depending on the tumor localization and it's relationship with the surrounding anatomical structures[17], a combined anterior and posterior approach is considered the best for en bloc total and high sacrectomies[18].

Tumor resection In order to achieve a long-term oncological control in treating a malignant sacral tumor, en bloc resection with wide surgical margins is required[4,19]. An incomplete resection of the tumor leads to a fast local regrowth, while intralesional resections imply higher local recurrence rate and decreased survival[9,20]. En bloc total or partial sacrectomy is an uncommon surgical procedure, due to a relatively high number of challenging factors including the patient's age, preoperative general condition, the tumor's extension and its relationship with the surrounding anatomic structures [18]. The postoperative complications generated by the neurological level of the

resection include lower extremity sensory and motor deficit, as well as bowel or bladder incontinence and sexual dysfunction[21]. As far as the general surgeon's involvement is concerned, this is required in cases of rectal involvement by the sacrococcygeal tumor. This particular situation, although uncommon, demands a resection including the rectum, with colostomy before sacral resection[22]. In some cases, in order to prevent local recurrence and to achieve proper surgical margins, resection of the piriformis and gluteus muscles or sacroiliac joint is mandatory[1,23], achieving wide resection margins remaining the most important predictor of local recurrence and survival[9].

The likelihood of perioperative complications is high in sacral tumor surgery due to the prolonged operating time necessary for the complex surgical techniques often required in the surgical treatment of these patients with often results also in severe intraoperative bleeding. Other intraoperative complications include visceral and vascular perforation, unplanned nerve root resection. As far as postoperative complications are concerned, in early postoperative period the patient may develop different wound or surgical site infections, ranging from 25% to 50% in the literature. This may be caused by a various number of factors including the proximity to the perineal region, poor blood supply of the skin flap, neural atrophy[8].

Also neurological deficit, losing sphincter function, and sexual capacity are other possible postoperative complications, resulting from a planned or sometimes incidental resection of the sacral nerve roots[24,25].

CONCLUSIONS

The majority of primary sacrum tumors are low-grade malignant neoplasms that generate nonspecific symptoms thus being able to reach enormous dimensions prior to diagnosis. Due to the complex anatomy of the sacral region, the surgical treatment is a difficult and laborious one, often requiring not only multiple approaches but also multiple surgeons, of different specialties depending on the tumors expansion. Also, the postoperative complications are often significant, a complete resection with wide surgical margins being often impossible although necessary for achieving long term local oncologic control. Therefore, total or subtotal sacrectomy for the treatment of sacral tumors should only be performed in experienced centers, with the infrastructure to support management of this disease, in order to be associated with acceptable rates of morbidity and mortality.

Considering the rarity of the disease and the complexity of the management, further studies are required, conducted in multiple centers, which offer the possibility of interdisciplinary approach.

Acknowledgment: *"This paper was co-financed from the European Social Fund, through the Sectorial Operational Programme Human Resources Development 2007-2013, contract POSDRU/187/1.5/S/155463 "Supporting excellence in scientific interdisciplinary doctoral research in the economic, medical and social fields", coordinator The Bucharest University of Economic Studies".*

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