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Case Report

# A Giant Sacrococcygeal Chordoma: Case Report and Literature Review

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**ABSTRACT**

**Introduction:** Chordomas are rare low-grade malignant lesions that are mostly seen in the spine. They constitute about 1% of intracranial tumors and 3-4% of all primary bone tumors. The principal spinal location is usually the sacrococcygeal and infrequently the sphenooccipital and cervical areas. **Case Presentation:** We present a 74-year-old man with a huge sacrococcygeal mass extending to both buttocks. Computerized tomography (CT-scan) and magnetic resonance imaging (MRI) as well as computerized tomographic angiography (CTA) evaluations were suggestive to chordoma. Immunohistochemical analysis confirmed the diagnosis after the patient was successfully operated on. All aggravating symptoms resolved after the operation. **Conclusion:** The management of giant sacrococcygeal chordomas can be very challenging but with all-inclusive treatment, complete cure is achievable although recurrence can occur. In our case, surgery alone was curative. Preoperative CTA aided us in achieving total tumor resection.

## INTRODUCTION

Chordomas constitutes about 1% of intracranial tumors and 3-4% of all primary bone tumors (1). These lesions are mostly seen in the spine although case have reported in other parts of the body. In the spine, 50-60% chordomas are seen in sacrococcygeal regions while 35% and 15% are seen in sphenooccipital and cervical regions respectively (2,3). Magnetic resonance imaging (MRI) is the gold standard imaging modality for assessing the degree of invasion of the tumor into soft tissue. However, plain X-ray and computerized tomography (CT-scan) is still very valuable in diagnosing these lesions (3). The standard treatment modalities for these lesions includes surgical resection, radiation therapy as well as chemotherapy. Even with all inclusive treatment, recurrence is common (4-6). We present a case of giant sacrococcygeal chordoma which we total resected with the aid of pre-operative CTA without further complications.

## CASE REPORT

We present a 74-year-old man with a huge sacrococcygeal mass extending to both buttocks. He noticed the mass 10 years ago but apparently did not seek remedy partly because of financial constrain and partly because the mass did not hinder his day to day activities. Over the years, the mass increased in size and aggravated into pelvic pain that radiates to both legs but more to right beg with urinary and fecal incontinence a month prior to presentation at our facility. He also experiences slight numbness in both legs. He could walk with support. Systemic evaluation was unremarkable. On examination we saw a huge sacrococcygeal mass extending to both buttocks measuring 10 x 12 x 10cm (Figure 1a). Neuromuscular examination revealed normal limb muscle strength, muscle tone as well as reflexes. General systemic examination was also unremarkable.

Initial CT-scan revealed a sacrococcygeal mass measuring 15 x 15cm (Figure 1b). A follow-up MRI also revealed a mass at the same site measuring 17 x 17 x 15 cm. On T1 imaging, the lesion had lower to intermediate homogeneous signal intensity with areas of focal high signal intensity. On T2 imaging, the lesion varied from homogeneous to heterogeneous with high signal intensity. The lesion had some septation and pseudocapsule (Figure 2, A-D). The lesion was enhanced on contrast agent with high homogeneous to heterogeneous signal intensities. There was lumbar degeneration

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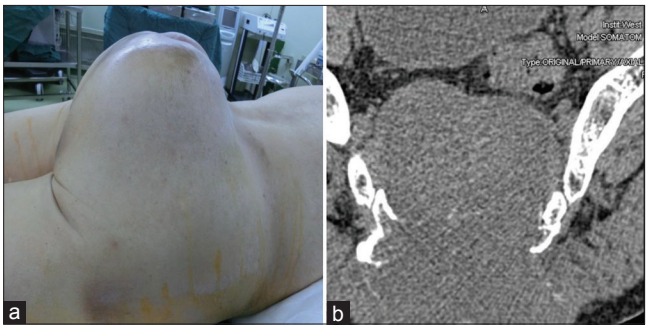

**Figure 1.** a is an image of the tumor during our assessment pre-operatively while b; is a pre-operative CT-Scan

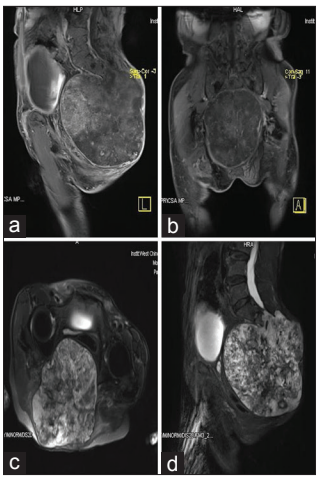

**Figure 2.** a-d are pre-operative MRI showing a huge tumor at the sacrococcygeal area

with some degree of osteoporosis. L3-4, L4-5, as well as L5-S1 disc were bulging forward with dural sac compression (Figure 3a). CTA however, showed normal vertebral architecture (Figure 3b). All routine laboratory investigations as well as chest x-ray and ECG were essentially normal.

We used posterior approach to resect this tumor. Intra-operatively, saw a huge sacrococcygeal mass which bulges into the skin. It measured about 17cm*17*15cm in size (Figure 4, a & b). The lesion had septations and each one was solid, tough with a capsule. The capsules were completely separated. The blood supply of the lesions was extremely abundant. The was ongoing internal necrosis in tumor. The necrotic regions were seen as jelly-like red and black. Also, the lesion was adhered tightly to the nerve roots of the sacral plexus. Furthermore, the tumor invaded and destroyed the sacrococcygeal bones. The bones in the inferior segment of the sacrococcygeal were eroded and destroyed along the border of the capsule. The tumor was completely resected en-block with maximum preservation of the pelvic fascia. The rectum and bladder were not invaded by the tumor. Post-operative MRI revealed total tumor removal (Figure 3, c &d) Immunohistochemical staining revealed positivity of pan cytokeratin (PKC) and S- 100 which consistent with chordoma with ICD-O encoding: 3 (Figure 5). The patient recovered remarkable after the operation. He was discharged home ten days later. Two years follow-up reveal no recurrence of the lesion and the all neurological defects restored.

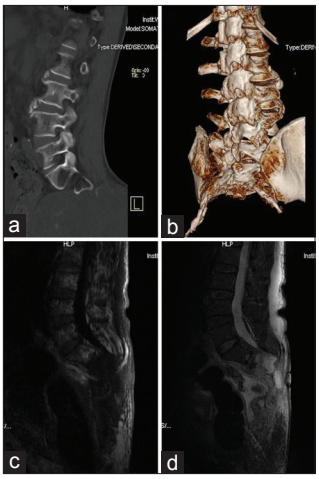

**Figure 3.** a; is a pre-operative MRI showing L3-4, L4-5, as well as L5-S1 disc bulging forward with dural sac compression. b; is a pre-operative CTA showing the lumber vertebrae and other surrounding vasculature. C & D are post-operative MRI showing total resection of the tumor

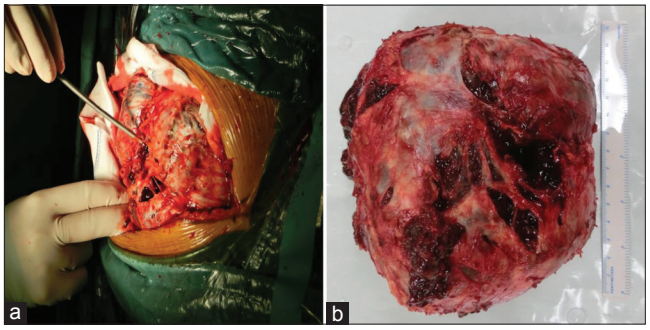

**Figure 4.** a; is an image showing the tumor after exposure intra-operatively while b; is an image of the resected lesion

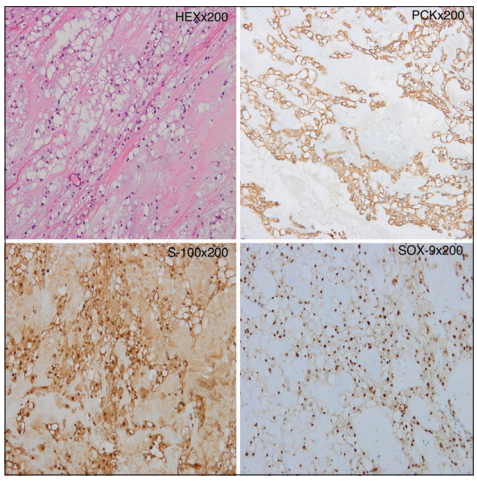


**Figure 5.** Is immunohistochemical images showing HEX, PCK, S-100, and SOX-9

## DISCUSSION

Sacrococcygeal chordomas are seen in about 50-60% of all patients presenting with chordoma while 35% and 15% are seen in sphenooccipital and cervical regions respectively (2,3). This lesion usually arises from the em-

bryonic notochordal remnant. The notochord constitutes part of the tissues from which the vertebrae is formed during embryonic development. The notochord typically vanishes after the emergence of the vertebrae but in rare cases, some tissue remains which may later develop into chordomas in late adulthood (2,3). Most sacral chordomas are seen in the elderly in their 50-70s with males predominately affected than females (2). Our case is also a male with advance age.

The most cardinal clinical presentation of sacrococcygeal chordomas is urinary retention, rectal sphincter incompetence, as well as radicular pain. These symptoms were the coordinal presentation in our case which necessitated him to seek remedy. It is evidence that as the tumor continues to flourish and invades on the sacral foramina, nerve root neuropathies occur resulting in the above symptoms. Studies have established that distant metastasis arises in 3-4.8% of the patients, most often associated with the lungs, liver, bone, skin, as well as lymph nodes (1,7). These neurologic signs and systems as well as distance metastasis are indicators of radical disease (2). Our patient presented with above signs and systems but had no distance metastasis.

The gold standard radiological modality in evaluating these lesions is usually both CT-scan and MRI most especially in patients with nonspecific symptoms (8). On MRI these lesions are depicted with high signal intensity on T2-weighted imaging because pathologically they are depicted with vacuolated cells as well as the production of abundant intra- and extracellular mucin (8-10). Furthermore, the associated prolonged T2 observed in these lesions is as a result from a property these lesions share with the nucleus pulposus of the intervertebral disc (10,11). These lesions are also depicted with high signal intensity on T1-weighted sequences because of focal areas of hemorrhage and high protein content of the myxoid and mucinous collections (8-10,12). Nevertheless, the presence of hemosiderin these lesions often make them have low signal intensity on T2-weighted imaging. These peculiar MRI characteristics are seldomly seen in spinal chordomas however, they are often observed in about 72% of patients (2,8-10). We advocate preoperative CTA in all case of giant sacrococcygeal chordoma because is able to tell the extend of invasion of the sacral bones and vertebrae.

The most efficient treatment modality for lesion located the sacrococcygeal region is surgery (2). It is normally challenging to achieve total resection of lesion during the initial operation because the anatomical location of these lesions hinders the aptitude of total resection (2,13). With very meticulous operation, we achieved total resection of the lesions in our patient (Figure). On the other hand, primary or adjuvant radiotherapy is an optional treatment modality although controversial because of minimal response and radio-resistance. Radiotherapy can use in circumstances like local tumor control, inoperable lesions, contaminated surgical margins, as well as elderly debilitating patients (5,14). These lesions seldom metastasize to distant sites however, Akyol et al. notice metastasis in their case 2 years after radiation therapy (2). Chemotherapy medications such as anthracyclines, cisplatin, and alkylating agents have not proven to effective in advanced lesions (6,15). All-inclusive treatment with intratumoral chemotherapy and surgical has proven to effective the local control of these lesions (16). Furthermore, molecular targeted therapies such as imatinib, an inhibitor of platelet-derived growth factor receptor-$\beta$ and c-KIT, has proven to effective in patients with advanced lesions (2,17). Nevertheless, the epidermal growth factor receptor (EGFR) antagonists has also proven to effective in patients with lesions refractory to imatinib (2).

On the microscopy, the lesions are frequently lobulated by septums of the connective tissue.4 On histological examinations, the tumor cells are structured in masses inside a myxoid stroma and the cytoplasm is mostly pale with multitudes of vacuoles making them appear as the so called "physaliphorous cells." Immunohistochemical studies usually reveal cells positivity with antibodies against vimentin, S-100, as well as epithelial membrane antigen (1,2,7,18). In our case, Immunohistochemical staining revealed positivity of pan cytokeratin (PKC) and S- 100.

## CONCLUSION

We advocate preoperative CTA in all case of giant sacrococcygeal chordoma because is able to tell the extend of invasion of the sacral bones and vertebrae. Preoperative CTA aided us in achieving total tumor resection. The management of giant sacrococcygeal chordomas can be very challenging but with all-inclusive treatment, complete cure is achievable although recurrence can occur. In our case, surgery alone was curative.

## ETHICS APPROVAL AND CONSENT TO PARTICIPATE

The ethical committee of West China Hospital fully approved our case study. The patient and his relatives were informed about our intention to involve him in a case study and he/they agreed to partake in the study. He/they signed the consent form before the operation was carried out according to all surgical protocols and consent was also obtained for the case details and accompanying images to be published.

## AUTHOR CONTRIBUTIONS

All authors contributed toward data analysis, drafting and critically revising the paper and agree to be accountable for all aspects of the work. All the authors have seen and approved the final manuscript for submission.

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