

# Giant Primary Spinal bony extra-axial tumors: case series and literature review

## ABSTRACT

Aim – Aim of this study is to study cases of giant primary spinal bony tumours (maximum diameter- >5cm) operated in a single institute including presentation, imaging, management and follow up with **histopathological diagnosis.**

**Study – Case series with** retrospective analysis of data

Place and duration – single center study with data collected from Jan 2011 to Nov 2023 in Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow.

Methodology – This is retrospective analysis of primary spinal bony tumours who underwent surgery in a single institute from Jan 2011 up to Nov 2023 with minimum follow-up of 1 month. We assessed a total of 27 patients who underwent surgery for bony spinal tumors with size >5cm. Of these 10 cases were excluded as biopsy came out to be metastases. Five cases were excluded as they were recurrent cases and three cases lost to follow up. So, a total of 9 patient's data was tabulated and analyzed. We analyzed presentation, imaging, treatment and outcome of these patients with review of literature.

Results - Mean age of presentation for benign tumors was 24.5 years whereas it was 46 years for malignant tumors. Mean duration of presentation for benign lesion was 15.75 months whereas for malignant lesions it was 3.4 months. 5 cases out of 9 (55%) were in dorsal region, 1 case (11.11%) in cervicothoracic region and 3 cases (33.33%) were in sacral region. All patients presented with symptoms of back pain, 7 (77.77%) had motor symptoms, 5(55%) presented with sensory symptoms and 5(55%) with bladder bowel. Out of 4 benign lesion one underwent intralesional excision, rest underwent marginal en bloc excision. Out of 5 malignant lesion one underwent intralesional excision, rest underwent marginal en bloc excision. Out of 4 benign lesions two patients (50%) did not show any improvement at 1 month follow up. Out of 5 malignant lesion 1 patient (20%) did not show any improvement at 1 month follow up.

Conclusion- Giant primary bony spinal tumours are rare entities. Early diagnosis and surgical treatment are paramount for tumour control and functional improvement.

Keywords: Giant, Bony tumors, Benign, Malignant, Fixation

## Introduction

Primary spinal tumors are more frequently seen in young adults as compared to metastatic bone lesion. The most common benign vertebral column tumor in adults is vertebral hemangioma whereas the most common malignant vertebral column tumor is multiple myeloma and plasmacytoma followed by **chordoma. Most frequent presenting symptom of these patients is pain often worse at night and occurs** even on rest. (1). These tumors can lead

to radicular pain if nerve roots are irritated. An aggressive lesion is more likely to be accompanied by pathological fractures or significant spinal cord compression. MRI and Ct are standard imaging techniques required for diagnosis and defining the extent of lesion. Surgery is the mainstay of treatment in these cases. Few case series are published in literature giving details of the giant spinal bony extra-axial. This case series is our effort to showcase giant primary spinal bony tumors and their management along with review of literature.

## Case Series

This is a retrospective analysis of giant primary spinal bony tumors which underwent surgery in a single institute from Jan 2011 up to Nov 2023 with minimum follow-up of 1 month. Giant tumor was defined as tumor with maximum diameter > 5cm. We found a total of 27 patients who underwent surgery for bony spinal tumors with size >5cm. Of these 10 cases were excluded as biopsy came out to be metastases. Five cases were excluded as they were recurrent cases and three cases lost to follow up. So, a total of 9 patient's data was tabulated and analyzed.

A summary of the patient characteristics is summarized in table 1

S. n.	Age/ Sex	Presentation	Durati on	Size( Max Diame ter)	Level	Histopatholo gy	Treatment	Follow up(1 month)
1	23/f	Spastic quadriparesis(power 2/5) with left supraclavicular swelling	7months	12cm	Cervicothoracic junction	Aneurysmal bone cyst(benign)	Marginal en bloc plus posterior and anterior fixation	Reduction in spasticity and improvement in power (3/5)
2	24/f	Backpain, spastic paraplegia(0/5), sensory loss below D7 and urinary incontinence	36 months	10cm	Dorsal	Epithelioid osteoblastoma (benign)	Intralesional excision with posterior fixation	Nonimprovement in symptoms
3	32/m	Low back pain a/w difficulty in passing urine and stool	7 months	8cm	Sacral	chordoma (malignant)	Marginal en bloc	Improvement in pain and urinary symptoms
4	35/f	Low back pain with	12 months	10cm	Sacral	Giant cell tumor	Marginal en bloc	Improvement

		radiculopathy, lower limb weakness(3/5), sensory loss and bladder involvement	hs			(Benign)		radiculopathy
5	73/m	Upper backache, spastic paraplegia(0/5), sensory loss and bladder bowel incontinence	2 months	6cm	Dorsal spine	Plasmacytoma(malignant)	Marginal en bloc	No improvement in power sensation improved
6	49/m	Spastic quadriparesis(3/5) with posterior column involvement	5 months	7cm	Dorsal spine	Plasmacytoma(malignant)	Marginal en bloc	Reduction in spasticity no improvement in power
7	60/m	Middorsal back pain with spastic paraplegia(0/5) and urinary retention	2 months	7.5cm	Dorsal spine	Plasmacytoma(malignant)	Intralesional excision	Improvement in power(1/5) and urinary symptoms. Received RT at 4 months
8	16/m	Low back pain with low back swelling and urinary incontinence	8 months	15cm	sacral	Aneurysmal bone cyst(benign)	Marginal en bloc	Remained incontinent for urine
9	15/m	Upper back pain with spastic paraparesis (2/5) sensory loss and bladder bowel incontinence	1 month	8cm	Dorsal	Plasmacytoma(malignant)	Marginal en bloc	Improvement in power(3/5) and reduction in spasticity

Here are some of the illustrative images of the case of cervicothoracic ABC operated shown in figure 1,2 and 3.

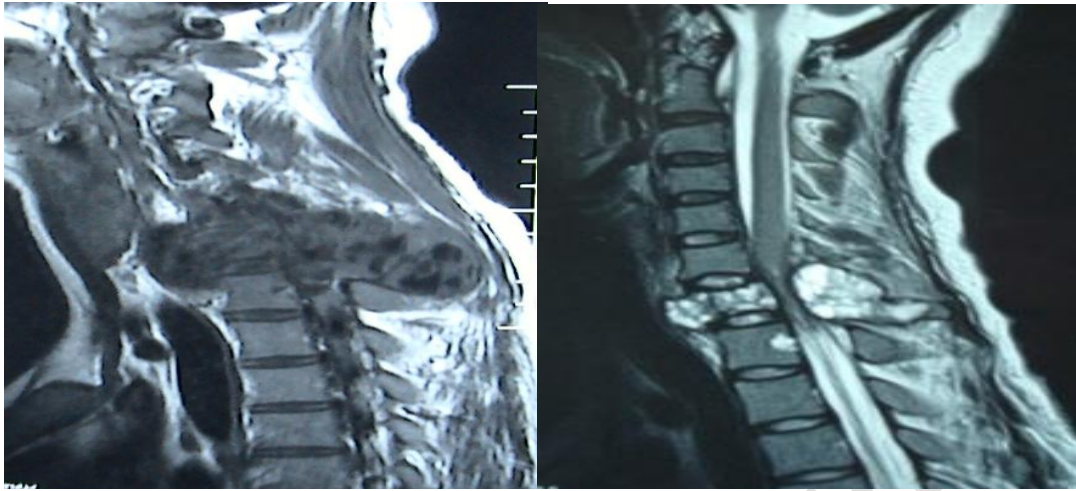


Fig 1 showing a large solid cystic bony tumor arising at cervicothoracic junction with cord compression at the level.

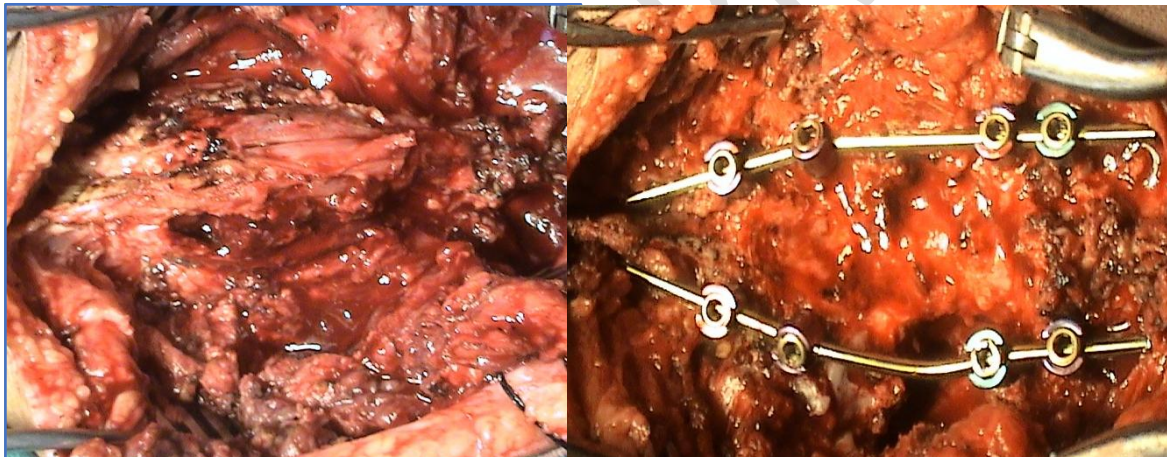


Fig 2 Image on the left showing red vascular lesion after exposure. Image on right showing excision of lesion with instrumentation.

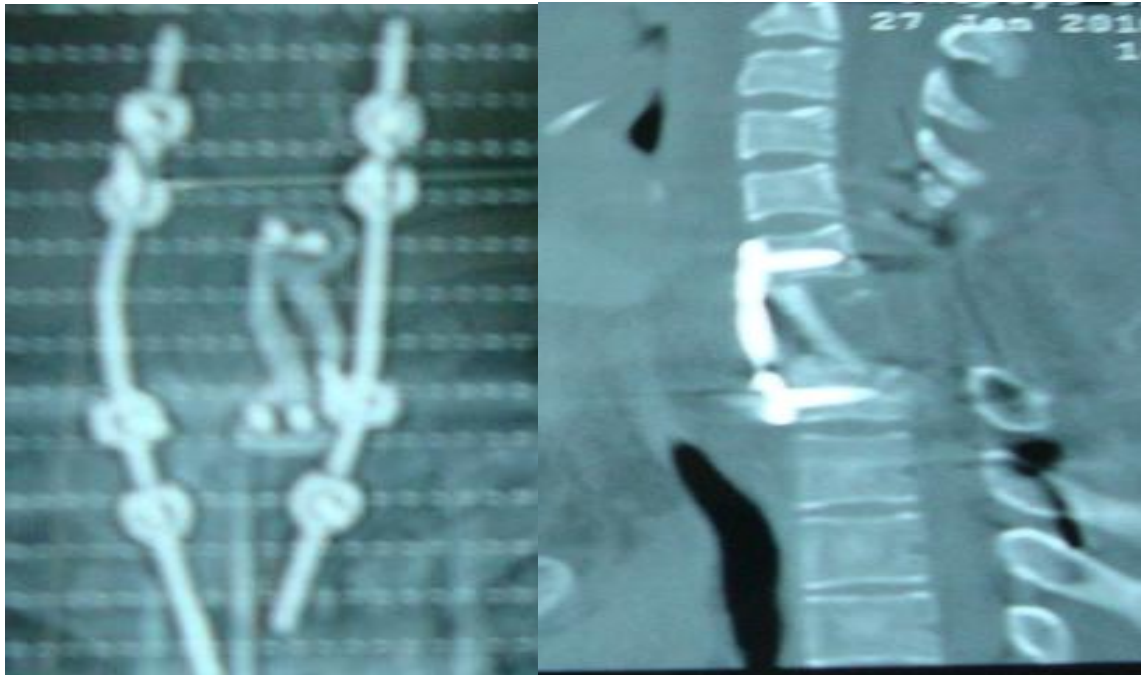


Fig 3 showing anterior and posterior instrumentantation following excision of lesion

## Discussion

We had a total of 9 operated cases of giant primary spinal bony tumors out of which 4 were benign and 5 were malignant. Mean age of presentation for benign tumors was 24.5 years whereas it was 46 years for malignant tumors. Mean duration of presentation for benign lesion was 15.75 months whereas for malignant lesions it was 3.4 months. 5 cases out of 9 (55%) were in dorsal region, 1 case (11.11%) in cervicothoracic region and 3 cases (33.33%) were in sacral region. All patients presented with symptoms of back pain, 7 (77.77%) had motor symptoms, 5(55%) presented with sensory symptoms and 5(55%) with bladder bowel symptoms. Out of 4 benign lesion one underwent intralesional excision, rest underwent marginal en bloc excision. Out of 5 malignant lesion one underwent intralesional excision, rest underwent marginal en bloc excision. None of the patient underwent wide or radical en bloc excision. None of the patients with benign lesion were advised adjuvant therapy, whereas one patient with malignant lesion who underwent subtotal excision underwent radiotherapy. Out of 4 benign lesions two patients (50%) did not show any improvement at 1 month follow up. Out of 5 malignant lesion 1 patient (20%) did not show any improvement at 1 month follow up.

**Primary spinal bony tumors** can present in a multitude of ways spanning from mechanical back pain to neurological deficits. Primary bony tumors can be primarily classified into benign and malignant lesions. Benign bony tumors generally occur in young adults and generally present with complaints of back pain increasing in intensity in nighttime. Malignant bony tumors generally occur in old age and presents with pain however the course of disease is short and generally have neurological deficit. In our series the mean age of presentation of patients with benign lesion was 24.5 years whereas it was 46 years for patients with malignant lesion. Mean duration of symptoms in patients with benign lesion was 15.75 months whereas its was 3.4 months in patients with malignant lesion. All patients presented

with history of back pain with 77% having motor symptoms and 55% had sensory loss and 55% had bladder bowel involvement. With advancement in imaging techniques and modalities diagnosis and treatment of these tumors have improved over time. MRI spine is imaging modality of choice for defining the extent of tumor as well as its relationship with neurovascular structures including spinal cord and nerve roots. CT spine is also used to better define the bony anatomy as well as destruction of bony structures. CT imaging is also important in planning for instrumentation in case of tumors associated with instability. The aim of surgery is to address mainly two important aspects, first is tumor burden and its compression over the neural structures, second is to address the instability caused by the destruction of bony elements by tumor. Cases with instability sometimes require multiple approach to safely resect the tumor as well as the instability. Out of nine cases two underwent spinal instrumentation following decompression of lesion. Sometimes there are extensive lesions which extend in multiple compartments sometimes along the myofascial planes and sometimes by infiltrating the myofascial planes. In those cases, sometimes the question arises if those cases can be competently handled by a neurosurgeon alone. In our experience it is better to take a multidisciplinary approach in handling such cases including pre-operative planning and post-operative surgical care. Extent of tumor resection can be designated as 'marginal', "wide," or "radical" en bloc resection. Any resection would be considered an "intralesional" resection when the tumor has been entered during the resection. In contrast, marginal en bloc resection involves removal of the tumor with dissection along the pseudocapsule but no entrance into the tumor. In wide en bloc resection, a continuous layer of surrounding healthy tissue is removed along with the tumor. Radical en bloc resection requires removal of the tumor along with the entire anatomic compartment of the tumor origin. However radical en bloc resection is generally not possible in spinal cases. In our series two cases underwent intralesional excision as pseudocapsule of tumour was adhered to neurovascular structures. Rest seven cases underwent marginal en bloc resection. However wide en bloc and radical en bloc resection was not possible in any of the case as the tumour pseudocapsule was in close proximity to neurovascular structures. The ability to accomplish marginal or wide en bloc resection of primary spinal tumors is largely based on tumor location and extension. Weinstein, Boriani, and Biagini devised a classification system (WBB staging system) for primary spinal tumours in which the vertebra is divided into 12 sectors numbered from 1 to 12 in clockwise order, with the spinal canal being the center. (16) In addition, the vertebra is divided into five layers ranging from the paravertebral extraosseous region to intradural involvement. Finally, the longitudinal extent of the tumour is defined by the number of spinal segments involved. Accordingly, neoplasms that are confined within the vertebral body or the posterior arch can be excised via marginal or wide en bloc resection. Moreover, tumours located eccentrically with unilateral pedicle or transverse process involvement (or both) and small paraspinous extension can be excised via marginal or wide en bloc resection with the sagittal resection technique. Conversely, tumours with extensive epidural involvement are not candidates for en bloc resection because the risk for neurological injury is high.

Most common benign primary spinal tumour is vertebral hemangioma and it commonly involves thoracic and lumbar spine. (2,3). We did not encounter any case of vertebral hemangioma in our study which is consistent with literature as they seldom cause clinical symptoms and are generally diagnosed as incidental finding. The lower thoracic spine is the most frequent site of symptomatic vertebral hemangiomas. (2,4,5). We encountered two cases of aneurysmal bone cyst which were first described by Jaffe and Lichtenstein (6) in 1942. Generally present in patients younger than 20 years with slight female preponderance, commonly involving thoracic spine. (7,8,9,10). In our series one case of ABC was male and one was female with age at presentation 16yrs and 23yrs respectively and site

sacral and cervicothoracic respectively. We also encountered one case of giant cell tumour. In literature GCTs account for 7-10% of all cases of primary spinal tumour (3,11). Patients with GCTs are most often seen initially in the third or fourth decade, and the sacrum is the most common site. Patients with sacral GCTs often have large tumours because of significant tumour growth before the development of neurological symptoms. Our case also presented in 3<sup>rd</sup> decade in sacral location with size of 10 cm (maximum diameter). We also had one case of osteoblastoma. Osteoblastoma typically presents in 2<sup>nd</sup> or 3<sup>rd</sup> decade of life and generally present with neurological compromise due to large size. None of the patients with benign lesion in our series underwent adjuvant therapy. Adjuvant radiation therapy has been shown to be beneficial to patients with GCTs and hemangioma. (17). In general response rate is worse with low grade tumours than with high grade tumours. The most common form of spinal radiation therapy is external- beam ionizing radiation. However, radiation therapy has its own pit falls the most severe of which is radiation toxicity to the spinal cord. Also, there is a possibility that exposure of surrounding tissues may lead to radiation-induced sarcoma especially in young patients with benign tumours. (18) In our series we had 5 malignant lesions in which four cases were plasmacytoma and one case of chordoma. Plasmacytoma is by definition a solitary lesion, whereas multiple myeloma is characterized by multiple lesions and considered a systemic neoplasm. This is the most common type of malignant spine tumour in the elderly, usually affecting men in their sixth or seventh decade of life. Bone pain affects almost 70% of patients and is the most common presenting symptom. (12). In our series we had cases of plasmacytoma presenting in 6<sup>th</sup> and 7<sup>th</sup> decade of life, however there was one case in which age of presentation was 16 years. One patient of plasmacytoma who underwent intralesional excision received radiotherapy. Plasmacytoma are radiosensitive and a good local control and long-term survival can be achieved with radiotherapy alone unless patients have spinal instability, neurological compromise, or intractable pain. (12,19-22). Chordomas are rare tumours that arise from embryonic notochordal remnants. They constitute 2% to 4% of all primary bone neoplasms. (13). In the spine the sacrum is the most common site of disease. The average age for sacrococcygeal chordomas is 56 years. Ideally wide or at a minimum marginal en bloc resection of spinal chordoma is the most critical factor in promoting tumour-free survival and decreasing local recurrence. (14,15). In our case age of presentation was 32 year with sacral location of tumour and marginal en bloc excision was done. Adjuvant radiotherapy is usually used for treatment of residual or recurrent chordoma. (15). Our study is a retrospective analysis of the data on giant primary bony spinal tumours, however larger prospective studies are required to devise proper guidelines for management of these tumours with emphasis on surgical management and adjuvant therapy.

## CONCLUSION

Giant primary spinal bony tumours are rare entities and can present in a multitude of way ranging from back pain to neurological compromise and spinal instability. With newer imaging techniques the diagnosis of these tumours at an early stage is possible. So, a patient with mechanical back pain increasing in night-time not improving with medication should warrant radiological imaging. Surgery with or without instrumentation remains one of the cornerstones of management. In cases of giant tumours, a multidisciplinary approach is often imperative. Role of adjuvant therapy in treatment of these tumours is much debated

and further studies are required to bring about a definitive guideline for their role in management of primary as well as residual/recurrent disease.

## **COMPETING INTERESTS**

Authors have declared that no competing interests exist in the study.

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