



Multidisciplinary Management of Spinal Chordoma: Single Institution Study

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Abstract

Introduction: Chordoma are rare, slowly growing, locally aggressive neoplasms of bone that arise from embryonic remnants of the notochord. This work aimed to evaluate the treatment results of a series of chordoma patients in our institute. More also, we try to clarify the importance of approaching such patients through a multidisciplinary team to maximize the oncologic and functional outcomes and to improve the patient's quality of life.

Patients and Methods: This study was conducted at SECI, surgical department during a period of 6 years duration. Patients were treated through multi-disciplinary team specialists of our cancer institute. Surgery and postoperative radiation therapy was offered to all patients.

Results: Results involves 8 patients with an average follow-up interval of 38.5 months. We had four cases of local recurrence which was salvaged with re-surgery and/or adjuvant radiotherapy. We had no postoperative motor neurologic deficit, with an excellent sphincter control in all patients. The 3-year survival rate in this study is 87.5%.

Conclusions: Multi-disciplinary treatment approach for chordoma is fundamental to improve overall outcomes. Surgery continues to be the primary modality in the management of chordoma. Rates of local recurrence, as well as survival, appear to be dependent on the achievement of negative surgical margins. Patients who relapse locally may have a poor prognosis but both radiation and surgery can be used as salvage therapy.

Introduction:

Chordoma is a rare primary bone neoplasm which arises from embryonic remnants of the notochord [1]; so it has an axial skeleton predilection. Chordomas are divided pathologically into conventional, chondroid and dedifferentiated types. Conventional chordomas are the most common and large vacuolated cells (physaliferous cells) are a characteristic of this tumor. Despite its low metastatic potential; distant metastasis to lung, bone, soft tissue, lymph node, and liver has often been reported in large set of patients 3. Spinal chordoma is often diagnosed late because of indolent and vague symptoms and a location deep in the pelvis. Therefore, it's usually large at the time of diagnosis and they may involve adjacent neurovascular structures and nearby vital organs in the pelvis 2. Bulky tumors adjacent to critical structures frequently preclude margin-negative resections. Because of that and because of its poor sensitivity to conventional radiotherapy and chemotherapy, chordoma almost always represent a surgical challenge [2, 4].

Surgery continues to be the primary modality in the management of chordomas. Rates of local recurrence, as well as survival, appear to be dependent on the achievement of negative surgical margins, with recurrence rates on the order of 70% in cases where negative margins are not achieved [5].

Resection should aim to be extensive and complete because, if not, recurrence is certain. More also, in the sacral region, preservation of S2 roots (at least in one side) is essential to avoid sphincter incontinence.

This work aimed to evaluate the treatment results of a series of chordoma patients in our institute. More also, we try to clarify the importance of approaching such patients through a multidisciplinary team to maximize the oncologic and functional outcomes and to improve the patient's quality of life

Material and Methods:

We approached those patients through our standard multidisciplinary team which included radio-diagnosis specialist, surgical oncologist, neurosurgeon,

clinical oncologist and rehabilitation specialists including pain management.

This study reviews outcomes of management of chordoma patients in our institute (South Egypt Cancer Institute) during the period of 2009-2015. Data of the records of chordoma patients were collected for demographic stratification, presentation, tumor characteristics and treatment method implemented. Data collection also included the multimodality treatment used, surgical approach, pathology reports with special concern to surgical margins and postoperative complications. The follow-up data was collected including the duration till the last visit or death, functional outcome, time of relapse and long-term morbidity.

Results:

Between 2009 and 2015, eight patients with chordoma disease were treated in South Egypt Cancer Institute, three males and five females. Their age ranged from 20 to 68 years old. Seven out of the eight patients were presented to our center primarily without previous surgical attempts. The last patient presented with a relapsing disease after previous surgical resection elsewhere.

Presentation:

Low back pain was the main presenting symptom in our cases. Radicular pain was present in 2 cases. Obstructed labor was the first presentation in one case. Incontinence was the presenting symptom in one case and reported in medical history of a second case as short-timed self-limited experience. Other symptoms included dysuria and dyschezia. Abdominal mass with no mechanical intestinal obstruction was the presentation in the lumbar chordoma case. He complained of pain more intense at right flank and lower back that radiate to the knees. The patient also experienced temporary short-timed incontinence for urine and stool resolved in few days. Another case presented to us with local recurrence detected in PET-CT imaging performed during follow-up after previous surgical resection of sacral chordoma 9 months before. The duration of these symptoms ranged from 2 months to 2 years with average of 8.5 months.

Anatomical Site and Size of the Tumor:

Sacral chordoma was recorded in six cases, in three of them S2 was involved while in another three its upper limit reached S3. In one case, we recorded a chordoma in the lumbar region, which extends opposite D12 and L1, 2 and 3 vertebrae with sizable intra-spinal and retroperitoneal extension. Perineal and pararectal mass lesions were the site of relapse in the last case. Mean tumor size was 11.6 cm (ranging between 8 and 20 cm).

Preoperative Diagnosis:

The preoperative diagnosis was based mainly on MRI imaging in conjunction with CT when needed. Pre-operative core needle biopsy was recorded in 5

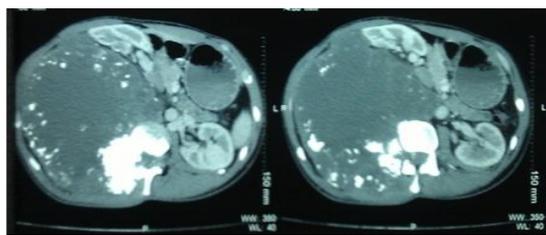
cases. In one patient diagnosis was through PET-CT screening during his follow up, that showed local relapse through increased uptake in the para-rectal area.

Surgical Approach:

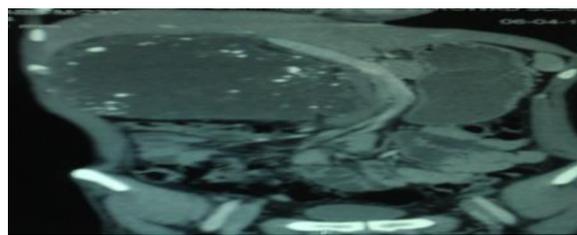
For surgical treatment, either a combined anterior and posterior approach (in 2 cases) or posterior approach (in 6 cases) was used. The surgical plan depended on the site and size of the tumor. In the combined antero-posterior approach, the surgical margin was wide in 1 case and intra-lesional in the 2nd case. When the posterior approach was carried out in other six cases, we had a wide margin in three patients and marginal resection on three cases.

In case of lumbar chordoma, MRI of spine and MSCT-Pelviabdomen revealed large retroperitoneal mass lesion measuring 15×18×20cm extending downward opposite the level of the 12th dorsal vertebra down to 3rd lumbar vertebra. The lesion was destructing the right lamina and pedicle of D12 and L1 vertebrae with intra-spinal extension causing compression upon the conus medullaris. The mass lesion was also shifting the big vasculature structures to the left side of abdomen and right kidney was shifted para-umbilical in position (Figure 1). Patient experienced temporary short-timed incontinence for urine and stool resolved in few days. EMG showed picture of lumbo-sacral radiculopathy with affection of cauda equina. Tru-cut needle biopsy was taken from the mass and the histo-pathologic examination reported chordoma nature of the lesion. This patient presented to our institute after being treated as an inoperable case in another institute by local radiotherapy, where he received 3000GY/10ttt. A two-staged surgical intervention was used in this case. Anterior approach was implemented first where surgical excision of the main bulk of the mass was done abdominally with a residual in the form of intra-spinal extension. Moreover, we encountered diaphragmatic infiltration which was resected and repaired over an inter-costal tube. The patient had smooth post-operative convalescence with removal of inter-costal tube. The 2nd stage was operated by neurosurgeon through a posterior approach where the residual intra-spinal extension of the tumor was resected. Final post-operative pathology report confirmed the diagnosis of chordoma with intra-lesional margin of resection.

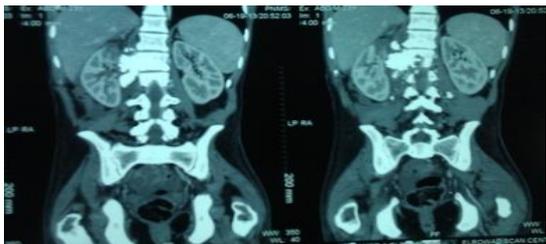
The other seven cases were sacrococcygeal chordomas mainly opposite the 3rd, 4th and 5th sacral vertebrae. One case was a young lady 20 years old presented with low back pain in her last 1.5 months of her pregnancy which was attributed to pregnancy till date of delivery when she faced an obstructed labor. Sacrococcygeal mass lesion opposite S2, 3, 4 & 5 and coccyx was discovered. One month after cesarean section delivery, she had a posterior approach technique with complete surgical resection of the lesion and preservation of S2, 3 and 4 nerve roots. Final post-operative pathology report confirmed the diagnosis of chordoma with wide margin of resection. The patient received adjuvant radiation therapy and she has no evidence of disease till her last follow-up visit.



A

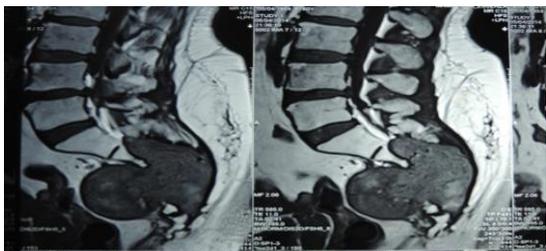


B



C

Figure 1. A. MSCT-PA of a case of male patient 25 years with lumbar chordoma showing large retroperitoneal mass lesion measuring 15×18×20cm destructing the right lamina and pedicle of D12 and L1 vertebrae with intraspinal extension. The mass lesion is shifting the big vasculature structures to the left side of abdomen and right kidney. B. The mass is seen compressing the right lobe liver stretching both the portal vein and the IVC. C. MSCT-PA showing residual mass lesion with intraspinal extension the first session (anterior approach) before the second session (posterior approach).



A



B



C



D

Figure 2. A. Sagittal Plane of magnetic resonance image of the for a case of female patient 56 years; showing large destructive mass lesion measuring 9×9×6cm infiltrating the sacrum. B. Intra-operative view of the mass operated through posterior approach. C. Intraoperative view showing preservation of sacral nerve roots. S1 nerve root is hooked by vascular tape. D. Late post-operative view of the patient showing the scar of the posterior approach and marking for radiotherapy planning field.

Table 1: Patient's data.

No.	Age	Gender	Symptoms	Duration of symptoms	Site	Size (cm)	Presentation	Surgical approach	Margins	TCNB	Additional Procedures	Radiotherapy	Saved nerve roots	Complications	Recurrence/ metastases	Time to relapse in months	Follow-up time in months	Outcome	Site of relapse
1	25	M	Abdominal pain with distension. Back pain with transient limping	6 months	lumbar with intraspinal extension	15*18*20 cm	PP	Combined (anterior then posterior)	Intra-lesional	Yes	Resection of infiltrated part of diaphragm with repair	Preop.	lumbar roots	-	Recurrence Metastasis	35	38	DOD	Local, Liver & Lung
2	56	F	Low back pain with radiculopathy	2 years	S2,3,4,5	9*9*6 cm	PP	Posterior	Wide	Yes	-	Preop.	S2,3,4	-	No	-	30	NED	
3	46	M	Back pain with incontinence to defecation	6 months	S3,4,5	9*4*5 cm	PP	Posterior	Wide	Yes	Colostomy	Postop.	S2,3 in one side	-	Recurrence ; operated.	63	80	NED	Local
4	24	F	Dysuria, dyschezia, Back pain	2 months	S3,4,5	9*7*5 cm	PP	Posterior	Marginal	No	-	Postop.	S2,3	Sphincteric weakness Wound dehiscence	Recurrence	9	40	AWD	Local; multiple spine levels
5	26	F	Dysuria, dyschezia with incontinence Back pain	8 month	S2,3,4,5	10*6*3 cm	PP	Combined anterior & posterior	Marginal	No	Covering colostomy	Preop. and postop.	S2,3	Wound dehiscence Urinary Incontinence	Recurrence	24	66	AWD	Local
6	68	F	Back pain with radicular pain	1 year	S3,4,5	18*11*15	PP	Posterior	Wide	Yes	-	Postop.	S2,3,4	-	No	-	14	NED	
γ	20	F	Back Pain Dyschezia Obstructed Labor	15 months	S2,3,4,5 and coccyx	10*8*9 cm	PP	Posterior	Wide	Yes	-	Postop.	S2,3,4	-	No	-	27	NED	
^	48	M	Local recurrence after previous resection 9 months ago		Perineal and para-rectal	3*2.5cm 5*3.5cm	SP	Posterior	Marginal	No	-	Chemotherapy Additional Post-op Radiotherapy	-	-	No	-	13	NED	

M: male; F: female; NED: no evidence of disease, DOD: dead of disease; AWD: alive with disease. PP: Primary presentation. SP: Secondary presentation. S: Sacral.

Table 2: Association of surgical approach with outcome

Approach	Number of patients	Recurrence	Outcome
Combined antero-posterior	2	2 (100%)	1 Alive (50%) 1 Dead (50%) at 38 mo.
Posterior	6	2 (33%)	6 Alive (100%)

Table 3: Association of surgical margins with recurrence

Margin	Number of patients	Recurrence	Time to recurrence
Wide	4	1 (25%)	63 mo
Marginal	3	2 (66%)	16.5 mo (9-24)
Intra-lesional	1	1 (100%)	24 mo.
Total	8	4 (50%)	32.7 mo. (9-63)

Table 4: Association of tumour size with outcome

Size	Approach	Surgical margin	Recurrence	Outcome
> 10 cm	Posterior (2)	1 wide 1 marginal	2 (50%)	3 alive – 1 AWD – 2 NED
	Antero-posterior (2)	1 wide 1 intra-lesional		1 dead – 1 DOD
< 10 cm	Posterior (4)	2 wide 2 marginal	2 (50%)	4 alive – 3 NED – 1 AWD

DOD: dead of disease; DOC: dead of complication; AWD: alive with disease; NED: no evidence of disease.

Table 5: Association of tumor site with outcome

Site	Surgical margin	Recurrence	Outcome
Lumbar (1)	Intra-lesional	1 (100%)	1 DOD
S2 (3)	2 Wide	1 (33%)	1 NED
	1 Marginal		1 AWD
Below S2 (4)	2 Wide	2 (50%)	3 NED
	2 Marginal		1 AWD

DOD: dead of disease; DOC: dead of complication; AWD: alive with disease; NED: no evidence of disease.

Table 6: Disease status at final follow-up.

Status of disease	No. of Patients
Alive without evidence of disease (NED)	5
Alive with evidence of disease (AWD)	2
Death due to disease (DOD)	1

The only case presented to our department with previous surgical exploration was discovered during PET-CT imaging in post-operative follow-up after 9 months. The patient was completely asymptomatic and the image showed para-rectal and perineal mass lesions. These lesions were approached posteriorly with marginal resection and he received additional radiation therapy. The other five cases were approached posteriorly in four cases (Figure 2) and combined posterior and anterior approach was used in one case. Pathology reports showed wide margin in three cases and marginal resection in two cases. Three of them developed local recurrence.

One interesting thing about case no. 3 (Table 1) is that the patient was subjected to surgical resection of the mass along with sacrectomy (S3, 4 and 5) through combined anterior and posterior approach. He received adjuvant radiotherapy with smooth follow-up for five years when the patient developed recurrent intra-pelvic para-rectal mass. Re-exploration through anterior approach was done with successful resection of the recurrent mass; confirmed to be recurrent chordoma in the final pathology report. The patient is free of disease till date of the study.

As regards sacrectomy, three cases underwent resection of S2, 3, 4 and 5 with the coccyx, while the other three cases underwent resection of S3, 4 and 5 with the coccyx. Sacral nerve roots S2, 3 and 4 were preserved in three sacrococcygeal chordoma cases. S2 and 3 sacral nerve roots were preserved in two cases on both sides and preserved in one case on one side only.

The operative time in this study ranged from 3.5 hours–6 hours with an average of 4.9 hours and blood loss ranged from 500 mL–3,700 mL with an average of 2400 mL. The mean blood transfusion needed during the procedure was 2.5 units of packed RBCs (1 – 5 units) and 1.8 units of fresh frozen plasma (0 – 4 units).

Complications:

Urinary incontinence and fecal incontinence (excluding the patient who had a preoperative colostomy due to anal incontinence) occurred in one patient (12.5%). Drop foot occurred in one patient (12.5%) who improved by physiotherapy and medical treatment. Early wound infection occurred in two patients (25%). Wound dehiscence and gapping which required surgical debridement occurred in two cases (25%), one case underwent excision through posterior approach and the other through the combined approach.

Radiation Therapy:

Most of the patients received radiotherapy. Neo-adjuvant radiotherapy was used in three out of the eight patients. One of these three patients received also palliative radiotherapy and chemotherapy after relapse while the other five patients received only adjuvant radiotherapy.

Outcome:

Overall, Four patients (50%) had relapses after a mean duration of 32.7 months (9 - 63). Among those, two patients were approached through the combined

antero-posterior technique. One case had inadequate margins (intra-lesional) and second case had marginal resection. The other two cases were operated by the posterior approach with marginal resection in one case and wide resection in the other.

We had 6 cases of sacral resections and one case of para-rectal and perineal resection for a recurrence. One of these cases had an anal incontinent with a diverting stoma at presentation due to S2 involvement that leads to incompetent anal sphincter. As regard the functional outcome, all cases showed good sphincter control in the immediate postoperative period apart from one case (12.5%) that had a urinary incontinence and a diverting colostomy due to an iatrogenic rectal injury that had been closed thereafter.

For treatment of relapses, one patient had local recurrence in conjunction with liver and pulmonary metastasis. He received chemotherapy and pain relieve regimen and died in 38th month of follow up. Another case had recurrence at multiple dorsal and lumbar vertebrae, and she received palliative radiotherapy and pain relief regimen. She is still alive with the disease after 40 months of follow up. In the third case relapse occurred after 5 years of follow up and he underwent re-resection and alive with no evidence of disease. The last case had a local recurrence in 24th months of follow up. She received local radiotherapy and still alive with disease for 66 months of follow up.

No peri-operative mortality was recorded in this study, we had only one mortality due to wide spread metastasis and local relapse in the 38th month of follow-up.

At their last follow-up, five patients had no evidence of disease (NED), two patients were alive with disease (AWD). The mean follow-up interval was 38.5 months (range, 13 – 80 months). The 3-year survival rate in this study is 87.5% (seven alive and one dead) at a mean follow-up of 38.5 months (Table VI).

Surveillance:

Clinical follow-up was conducted regularly by computed tomography or magnetic resonance imaging of the pelvis was done every 3-6 months.

Discussion:

Chordoma has unique characteristics that make it a challenging disease in many aspects. It's a rare (4% of malignant bone tumors), slow-growing, locally aggressive neoplasm of bone which arises from embryonic remnants of the notochord [6]. The deep locations within the pelvis and the mobile part of the spine which account for 65% of the site incidences and because of indolent and vague symptoms, chordomas are often diagnosed late [1,2,7]. The tumors occur twice as frequently in men as in women and are uncommon under the age of 40 years [7]. However, in this study the female incidence represent almost double the male incidence, and 50% of the cases are below the age of 40 years.

Spinal chordomas are usually large at the time of diagnosis and they may involve adjacent neurovascular

structures and vital organs [2]. The true extent of the disease is therefore often not appreciated until late in the course of the malignancy. The mean time interval between the onset of symptoms and presentation was 8.5 months ranging between 2-24 months in our records; it has previously been reported as 24 to 44.4 months in literature [2,7,10].

Spinal chordoma patients commonly presented with poorly localized lumbar or gluteal pain. This is owed to pressure of the tumor mass on the pre-sacral fascia and anterior soft tissues within the rigid pelvis. Tumor infiltration into the vertebral foramina and pressure on adjacent sacral nerve roots may also result in urinary incontinence, bowel dysfunction and lower limb neurological compromise. Approximately one third of patients have radicular pain involving the lower limbs at presentation due to compression of the ipsi-lateral sacral nerve or ilio-lumbar trunk [8]. Low back pain was the main presenting symptom in our cases. Incontinence was the presenting symptom in one case and reported in medical history of a second case as short-timed self-limited experience. We record unusual complaints among patients in this series of cases. Obstructed labor was the first presentation in one case. Huge abdominal mass that occupy most of the abdomen was recorded in the lumbar chordoma case. He complained of pain more intense at right flank and lower back that radiate to the knees. In another case the presentation was by local recurrence that had been detected in PET-CT imaging performed during 9th month of follow-up after previous surgical resection of sacral chordoma.

The relative rarity of the disease also accounts for why most previous clinical studies have been based on small series collected over a long period of time with highly variable treatment and outcomes. So that the cumulative experiences of a multidisciplinary team are essential to afford the best care and achieve good functional and oncologic outcomes to chordoma patients.

Diagnosis of chordoma is based on imaging and histopathology. Many authors have reported that MRI is more useful than CT in determining tumor infiltration in chordoma patients [13,23,24]. Owing to the excellent soft-tissue resolution and multi-planar capacity, MRI is now considered the best imaging modality to assess tumor location, size, morphology and relation to adjacent structures. The risk of malignancy can be estimated based on following radiologic features: heterogeneity of the mass, irregular or infiltrative borders, variable enhancement after gadolinium administration, rapid growth. Biological behavior also correlates with diffusion restriction. T1-weighted images are useful for determining tumor infiltration into bone and that T2-weighted images are useful for determining tumor infiltration into soft tissue [24]. Diffusion-weighted imaging can distinguish benign and malignant lesions, using quantitative ADC measurements [25]. During this study, MRI was the cornerstone in the preoperative work up and in the surgical planning in conjunction with CT when needed. The extent of soft tissue infiltration is assessed best using MRI with T2-weighted images whereas bone

infiltration is usually examined using CT and MRI with T1 weighted images.

Less invasive methods for pathologic diagnosis are much favorable. This includes fine needle aspiration (FNA) or image-guided core needle biopsy. Open biopsy or trans-rectal and trans-vaginal biopsies increase the risk of local recurrence rate and metastasis as well as decreases overall survival rate [8]. In our series, the preoperative diagnosis was based on core needle biopsy in 5 cases. Two patients underwent surgery based only on clinical and radiographic findings and in another case diagnosis based on PET-CT screening during his follow up, that showed local relapse through increased uptake in the para-rectal area.

Because of chordoma has poor sensitivity to radiotherapy and chemotherapy, surgery continues to be the primary modality in the management of chordomas [2,4,7,8].

The deep locations of spinal chordoma, especially in the sacral region and its complex anatomy and its close relationship with major vessels, sacroiliac joints, sacral nerve roots and pelvic viscera like the rectum. This makes it hard to achieve wide surgical margins during surgery and increases peri-operative morbidity and mortality. Resection of the tumor with wide margin represents a major surgical challenge that may require sacrificing important nerve roots, mechanical elements that contribute to the stability of the pelvis and even viscera, causing deficits in mechanical, neurological or visceral functions. Therefore, it is essential that the patient be informed of all these risks and accepts them before treatment [15,17]. More important it's essential to approach those patients through a multi-disciplinary specialized team to optimize the treatment results and to minimize the morbidity.

Through their small sized series of cases and over long time, authors trying to predict factors that influence the survival and local recurrence in chordoma patients.

Different studies have shown that the single most important predictor of survival and local recurrence was the margin obtained during resection [8,10,12,16].

The dismal prognosis of chordoma patients treated with traditional surgical debulking with insufficient surgical margin (with or without adjuvant radiotherapy) has been illustrated in a population-based study of 51 cases with extended follow-up in which only one long term survivor was identified [11]. In a series of 52 chordoma patients, all (100%) of patients treated with radiation alone, palliative therapy, or intra-lesional intra-capsular excision had local recurrence within 17–20 months. In contrast, only 20% of patients had local recurrence after en bloc resection with appropriate margins at 56–94 months [5]. Similarly, in another study where aggressive surgical approaches were used to achieve total wide resection in up to 70% of patients, resulting in long-term control in 50% of cases [9].

In a study of 39 patients whom underwent surgical resection for sacral chordoma, 4 out of 23 patients (17%) with wide excision margins developed local recurrence while 13 out of 16 patients (81%) with intra-lesional or marginal resections developed recurrence

[8]. Also in another study, localized disease recurrence was reported in 28% of patients with en bloc sacrectomy but in 64% of patients in which the tumor had been breached intra-operatively [13].

In this work, we manage to obtain wide resection margin in 4 cases, marginal resection in another 3 cases (Table III). In the last case (Intra-lesional Resection), we had to operate in 2 sessions, the 1st one was anterior approach through which the main intra-abdominal and retro-peritoneal bulk of the tumor was removed. The 2nd was posterior approach through which the inter-spinal resection achieved. The local recurrence rate was 50%.

The anatomical location of the chordoma within the sacrum is an important prognostic variable with studies showing greater cephalad tumor extension leading to increased risk of disease recurrence and reduced survival [20,22]. In one of these studies; local disease recurrence occurred in 30% of patients with tumors at or caudal to the S3 vertebral level but in 50% of patients with tumors cephalad to S3 and this difference was statistically significant [22]. In this work, apart from lumbar chordoma case, three out of seven patients had their disease up to S2 level and the remaining ones where at and below S3 sacral vertebra (Table V).

Chordoma size is another important prognostic variable with larger primary tumors being associated with increased risk of disease recurrence and reduced survival [8,14]. There is however no uniform consensus on the diameter of the tumor above which prognosis significantly worse. In a study that followed 17 patients after sacrectomy, the local recurrence rate was 46% with tumors greater than 10 cm and 17% in tumors less than 10 cm [21]. Other studies have shown increased risk disease recurrence and reduced survival in tumors greater than 5-10 cm in diameter [8,10,14].

The mean tumor diameter in this work was 11.6 cm (ranging between 8 and 20 cm). In another studies, a mean diameter of 9 cm was reported in a series of 52 patients and 7.8 cm in a series of 12 patients [2,10]. In contrast to most literature, the rate of tumor recurrence was similar (50%) in our study whether the maximum tumor diameter was above or below 10 cm. However, 100% of our patients were alive at the last follow-up if the tumor diameter was 10 cm or below, and 75% were alive if it was more than 10 cm initially (Table IV). Smaller tumor size seems a favorable factor with regard to recurrence and survival, but a statistical analysis was not possible because of the limited number of patients.

Extirpation of spinal chordoma is possible through posterior or combined antero-posterior approach. Multiple authors are trying to propose an algorithm for the surgical approach of pre-sacral tumors. The algorithms are similar as regard the size, pelvic side wall and visceral involvement, but differ in regards of the sacral piece involved S2, S3 or S4 [7,18,19]. The posterior approach has the advantages of being single stage procedure with shorter operating time but the viscera, organs and great vessels within the pelvis are at increased risk of injury during osteotomy. In the contrast, the combined antero-posterior approach enables the visceral organs to be dissected away from

the tumor and protected during the osteotomy but is associated with the additional morbidity of the laparotomy incision. One may assume that the approach should be individualized for each case and depends on the experience available.

In addition to the technical challenges obtaining negative margins, the surgical management of chordoma may result in poor functional outcomes. We approached sacrectomy through a combined team of onco-surgeon and neuro-surgeon and the functional results reported were good in the immediate postoperative period apart from one case (12.5%) that had a urinary incontinence and a diverting colostomy due to an iatrogenic rectal injury that had been closed thereafter.

Recurrent tumors are generally more challenging for surgical interventions and clearly have worse overall outcomes [9].

Local recurrence has been associated with a higher risk of distant metastasis. Several authors report the most common site of recurrence in gluteus muscles (gluteus maximus and piriformis); therefore, for complete resection of the tumor, it is necessary to achieve a wide posterior margin including gluteus muscles. However, no local recurrence has been observed in the anterior region. This is thought to be due to the presence of a firm pre-sacral fascia that runs from the anterior longitudinal ligament to the sacrum which avoids anterior invasion. For this reason, extensive margins in the anterior region are not necessary [27].

In this series, local recurrence rate was 100% after intra-lesional resection margin and 66% after marginal resection margin (Table III). Local recurrence was reported in one of four cases (25%) of wide margin of resection, recurrence occurred in 63th months of follow-up and unusually it's located intra-pelvic despite that a posterior approach was implemented during resection. Recurrence resection was conducted via anterior trans-abdominal approach and has no evidence of disease at last follow-up.

A recurrence rate ranges from 33% to 70% and five-year survival as high as 86% has been reported in literatures [1,3,8]. The mean follow-up in this series was 38.5 months (range, 13 – 80 months); the 3-year survival rate estimated to be 87.5% and the recurrence rate was 50% after a mean duration of 32.7 months (9 - 63). No peri-operative mortality was recorded in our study, however we had only one mortality, (Died of Disease) due to wide spread metastasis and local relapse in the 38th month of follow-up. The rate of metastasis is between 20% and 43% in literatures [1,3,28].

The value of radiotherapy in chordoma as primary or adjuvant treatment has been debated. Whereas some investigators have reported little effect with radiotherapy, others have described improved local control and prolonged disease free survival after radiotherapy as the sole modality or as adjuvant treatment [2,8].

In this series of cases, neo-adjuvant radiation was received on palliative base as patients were received to us from other centers considered them inoperable and

received radiation there on palliative base. The adjuvant radiation was received in patients with marginal or positive margin on the hope rather than on the belief to improve local control.

Given the rarity of the chordoma tumor, the value of radiation therapy for its treatment needs to be addressed in future prospective multi-institutional studies. In a study by Imai, 95 patients with medically un-resectable sacral chordoma were treated with carbon ion radiotherapy, and the overall survival rate and local control rate at 5 years was 86 % and 88 % respectively [29]. This treatment may offer a promising alternative to surgery. Moreover, chordoma are not sensitive to chemotherapy, although, some chemotherapeutic agents are used in recurrent disease or in un-resectable tumors, such as imatinib mesylate or in combination with cisplatin [3].

In conclusion, multi-disciplinary treatment approach for chordoma is fundamental to improve overall outcomes. Surgery continues to be the primary modality in the management of chordoma. Rates of local recurrence, as well as survival, appear to be dependent on the achievement of negative surgical margins. Patients who relapse locally may have a poor prognosis but both radiation and surgery can be used as salvage therapy.

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