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Primary chondrosarcoma of bone: a clinicopathological and oncological outcome analysis

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ABSTRACT

Chondrosarcomas are a heterogenous group of cartilaginous matrix-producing malignant neoplasms. They have varied morphological features and different clinical behaviors. The tumors usually range from benign low-grade tumors or intermediate atypical cartilaginous tumors (ACTs), to malignant, aggressive high-grade tumors. A multidisciplinary team at a tertiary sarcoma centre allows for optimal management of these patients. To determine the clinicopathological and survival outcomes of patients of chondrosarcoma, we report a total of 20 cases in the last 10 years at our institution. Our results have shown that chondrosarcomas occurred in different age groups and most common location was in axial skeleton. Most common histological type was conventional chondrosarcoma and majority were high grade tumors. All patients had undergone surgery and majority of the patients were alive after many years of treatment with an overall survival of 85%. Long term survival of patients is attributed to completeness of surgical resection.

Keywords: Chondrosarcoma, Mesenchymal, Conventional chondrosarcoma, Survival

INTRODUCTION

Chondrosarcoma, although relatively uncommon, constitutes a significant percentage of primary malignant osseous tumors. Following osteosarcoma, it is the second most common primary solid tumor of bone. Chondrosarcoma primarily affect adults with incidence rising with increasing age.[1] Survival rate of patients with chondrosarcoma is higher than that of osteosarcoma and Ewing's sarcoma with 5-year survival rate between 72-75% and 10 year survival rate of 69%.[2-4] The standard treatment for chondrosarcoma is surgical excision with adequate margins. This can be because chondrosarcomas are resistant to both chemotherapy and radiotherapy due to poor vascularity, high extracellular matrix and low percentage of dividing cells.[5] This study aims to analyse the demographic, clinical, pathological, and survival characteristics of chondrosarcoma cases at a tertiary care institute.

CASE SERIES

We report 20 cases of chondrosarcoma in the last 9 years with mean age of 45.2 years. Age at diagnosis ranged from 26 to 65 years. Out of 20 cases, 14 were male and 6 were female as shown in Table 1 and 2.

The 60% of cases were in extremities with lower extremity being most common, 30% in chest wall and 10% involving the pelvis. High grade tumor was more common constituting 60% of cases and low grade tumors 40% of cases. The most common pathological subtype of chondrosarcoma was conventional type (70%) and mesenchymal type (30%). The X-ray of local part was taken for all cases and majority showed features of lytic lesion with calcifications, cortical thickening and wide zone of transition. High grade lesions had permeative pattern, minimal or no calcifications, cortical breach on the X-ray.

Table 1: Overall clinicopathologic characteristics, treatment and outcome.

Variables	N	Percentage (%)
Age (in years)		
<45	10	50
>45	10	50
Gender		
Male	14	70
Female	6	30
Tumor site		
Upper limb	1	5
Lower limb	11	55
Chest wall	7	30
Pelvis	2	10
Tumor size		
<5 cm	Nil	Nil
5-10 cm	10	50
>10 cm	10	50
Tumor subtype		
Conventional	14	70
Mesenchymal	6	30
Tumor grade		
Low	8	40
High	12	60
Surgery		
Yes	20	100
No	Nil	Nil
Radiotherapy	Nil	Nil
Chemotherapy		
Palliative	3	15
Adjuvant	Nil	Nil
Distant metastases		
Yes	3	15
No	17	85
Survivor		
Alive	17	90
Dead	3	10
5-year, DFS*	-	85
5- year, OS**	-	85

*DFS-disease free survival, **OS-overall survival.

Table 2: Clinicopathological presentation, treatment management and outcome of 20 patients with chondrosarcoma.

Patients no.	Age (In years)	Gender	Tumor site	Tumor size (cm)	Pathological subtype	Grade of tumor	Surgery	Adjuvant radiation/chemotherapy/ palliative chemotherapy	Recurrence	Disease free survival after initial treatment
1	55	Female	Right iliac bone	10×10	Mesenchymal	High	Right hemipelvectomy	Nil	Lung metastasis	4 years (not alive)
2	35	Male	Right proximal humerus	10×10	Conventional	High	Shoulder girdle resection and CMP reconstruction	Palliative chemotherapy for distant recurrence	Lung metastasis	4 years (not alive)
3	55	Male	Left distal femur	10×10	Conventional	High	Distal femur resection with CMP reconstruction	Nil	Nil	8 years (alive)
4	60	Male	Right distal femur	12×12	Conventional	Low	Distal femur resection with CMP reconstruction	Nil	Nil	8 years (not alive)
5	58	Female	Left chest wall	11×11	Conventional	Low	Wide local excision	Nil	Nil	8 years (alive)
6	65	Female	Lt prox. femur	8×8	Conventional	Low	Proximal femur	Nil	Nil	6 years (alive)
7	46	Male	Right distal femur	7×7	Conventional	High	Distal femur resection with CMP reconstruction	Nil	Nil	6 years (alive)
8	65	Female	Left distal femur	12×12	Mesenchymal	High	Distal femur resection with CMP reconstruction	Palliative chemotherapy for distant recurrence	Yes	7 years (not alive)
9	50	Male	Left proximal femur	10×10	Conventional	Low	Proximal femur resection with CMP reconstruction	Nil	Nil	7 years (alive)
10	24	Male	Left proximal tibia	9×9	Conventional	High	Proximal tibial resection with CMP reconstruction	Nil	Nil	6 years (alive)
11	44	Male	Right proximal tibia	10×10	Conventional	Low	Proximal tibial resection with CMP reconstruction	Nil	Nil	6 years (alive)
12	34	Male	Left proximal tibia	13×13	Conventional	High	Proximal tibial resection with CMP reconstruction	Nil	Nil	6 years(alive)
13	58	Male	Left chest wall	8×8	Mesenchymal	High	Wide local excision	Nil	Nil	6 years (alive)
14	38	Male	Right 8th rib	7×7	Conventional	High	Wide excision with rib resection	Nil	Nil	5 years (alive)
15	27	Female	Rt prox femur	10×10	Conventional	High	Hip disarticulation	Nil	Nil	5 years (alive)
16	25	Male	Right chest wall	10×10	Mesenchymal	Low	Wide local excision	Nil	Nil	5 years (alive)
17	26	Female	Right chest wall	6×6	conventional	Low	Wide local excision	Nil	Nil	5 years (alive)
18	58	Male	Left chest wall	8×8	Mesenchymal	High	Wide local excision	Nil	Nil	6 years (alive)
19	44	Male	Left distal tibia	11×11	Conventional	High	Left below knee amputation	Nil	Nil	5 years (alive)
20	65	Male	Left chest wall	13×13	Mesenchymal	High	Wide local excision	Nil	Nil	5 years (alive)

All 20 cases had undergone surgery according to the site of tumor namely hemipelvectomy for pelvic tumors (2 cases), bone resection with custom made prosthesis reconstruction for extremity tumors (12), wide local excision of chest wall tumors (7 cases), unilateral hip disarticulation (1case) and below knee amputation for ankle tumor (1 case). After complete surgical resection (R0 resection), none of the patients received adjuvant radiotherapy or chemotherapy.

Out of 20 patients, 3 patients developed lung metastasis during follow-up with average disease-free interval of 3 years and all 3 had high grade chondrosarcomas. All three were given palliative chemotherapy and all 3 died due to progressive disease. One patient who had chondrosarcoma of proximal femur and underwent hip disarticulation developed local recurrence after a disease-free period of 5 years for which hemipelvectomy was done. The 5-year disease-free survival is 85%, and 5-year overall survival is 85%.

DISCUSSION

Chondrosarcoma is the 2nd most common primary osseous tumor behind osteosarcomas and makes up about 25% of total prevalence of bone tumors.[6] Chondrosarcoma occurs predominantly in patients aged 50-70 years.[7,8] In our study, 50% cases were in patients aged <45 years and 50 % in patients >45 years. The most common location of involvement chondrosarcoma includes bones of axial skeleton (pelvis, scapula, sternum, ribs) followed by proximal humerus.[9] In our study, 55% of tumors were found in lower limbs followed by chest wall (35%).

Most common subtype of chondrosarcoma in our study was conventional type. Primary or conventional chondrosarcoma arise in normal bone and is distinguished from rarer secondary tumors which occur in pre-existing enchondromas or osteochondroma.[9] Conventional chondrosarcomas account for 85-95% of chondrosarcomas and is subdivided into central, peripheral and periosteal. Non-conventional variants include clear cell, mesenchymal, dedifferentiated chondrosarcomas.[10] There are very limited treatment options for chondrosarcoma. Typically, chondrosarcomas are insensitive to chemotherapy and radiotherapy.[11] All the patients in our study had non metastatic disease at initial presentation and had undergone upfront surgery and none of them required adjuvant chemotherapy and radiation due complete resection. Zhan Wang et.al reported that surgical treatment of primary tumors would increase survival of metastatic chondrosarcomas.[12]

The most common site of distant metastasis in chondrosarcomas is the lung. Out of 20 patients, 3 of them developed lung metastasis during follow up period and received palliative chemotherapy but survived less than a year. All three had high grade chondrosarcoma (2 were conventional type and 1 was mesenchymal type). Conventional chondrosarcoma does not respond to chemotherapy.[13] Additionally chemotherapy was not associated with improved survival.[14,15]

Among the three most common primary sarcomas of the bone, chondrosarcoma has been shown to have the highest 5-year survival rate.[3] The 5-year survival of grade I chondrosarcomas is 83%. In our study the 5 year- disease free survival is 85% and overall survival 85%. The 5-year survival of grade II and III chondrosarcomas is of 53%.[16] The higher survival rate in high grade tumors in our case series is probably due to complete surgical resection with adequate negative margins.

CONCLUSION

This study has shown a variable incidence of chondrosarcoma in terms of age and tumor location. The continued role of surgical intervention over the years as the mainstay of treatment for both appendiceal and axial chondrosarcomas has been demonstrated in this study. Long -term survival of patients with chondrosarcoma depends on completeness of surgical resection which has been illustrated in our study. Also, survival rates vary significantly based on grade and metastases. Appropriate preoperative surgical planning enables completeness of surgical resection and good oncological outcomes.

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