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Original Article

Chondrosarcoma: an analysis of clinical characteristics, treatment outcomes, and prognostic factors in a decade-long single center experience

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Abstract

Aim: Management of chondrosarcoma patients is associated with histologic subtypes and clinical behaviors. This study describes our institution's management and outcome experience for 12 chondrosarcoma patients with higher grades in rarer locations.
Materials and Methods: Pathologically diagnosed chondrosarcoma patients between January 2013 and January 2023 were retrospectively scanned. Patients under 18 years, duplicate records, second primary malignancy, index surgery at another hospital were excluded, and data of remaining 12 patients were statistically analyzed.
Results: Twelve patients (6 female, 6 male) had median age of 62 (21-94) years. Locations of primary tumors were femur (n=4), larynx (n=1), left gluteus (n=1), lung (n=1), periscapular soft tissue (n=1), tarsal bone (n=1), distal metacarps (n=1), scapula (n=1), and sternum (n=1). There were local relapses in three cases (chest wall, ipsilateral femur, acetabulum, and ipsilateral gluteal region) and distant metastasis in three cases (contralateral lung, lung, bilateral lung, and pleura) during the follow-up. The median tumor size was 5.65cm (1.5-21.5cm). Histologic subtypes were Chondrosarcoma (Grade I) (n=4), Chondrosarcoma (Grade II) (n=1), Chondrosarcoma (Grade III) (n=2), Dedifferentiated Chondrosarcoma (Grade III) (n=2), Extraskeletal Mesenchymal Chondrosarcoma (n=3). Surgical treatment was eight wide resections, one lobectomy, one laryngectomy, one amputation, and one curettage. Five patients received chemotherapy (2 neoadjuvant chemotherapy, 3 adjuvant chemotherapy), two patients received low dose (50.4Gy) (3DCRT; IMRT), and two patients received high dose (>60Gy) (IMRT) adjuvant radiation therapy. The median follow-up was 26.5 months (2-121 months). The median survival was 26.5 months (2-121 months). Median relapse-free survival was three months (0-96 months). The overall and relapse-free survivals at two years were 81.5% and 50%; at five years, 71.3% and 25%.
Conclusion: Our results demonstrate that surgery is the gold standard treatment, but well-managed adjuvant oncologic treatment provides undeniable advantage for overall survival in selected cases.

Keywords: Soft tissue sarcoma, chondrosarcoma, mesenchymal chondrosarcoma, dedifferentiated chondrosarcoma, radiotherapy, IMRT

INTRODUCTION

The term chondrosarcoma includes a group of malignant hyaline cartilage-producing bone tumors with multiple morphological subtypes. It typically affects the bones of the pelvis, shoulder, and ribs. It is also common in the knee, femur, and humerus. It is the second most common malignant tumor affecting bones, accounting for approximately 20% of all primary bone tumors. Chondrosarcomas can be classified into three grades based on the degree of malignancy, with the most severe form being grade III. Conventional chondrosarcoma is the most common subtype, with a rate of 85%. Other less common histologic subtypes are periosteal chondrosarcoma, mesenchymal chondrosarcoma, and clear cell chondrosarcoma (1). Apart from these morphological subtypes, the location of the tumor and histological grading are also used to exclude chondrosarcomas. Generally, the term central is used for chondrosarcomas arising from the medulla.

In contrast, the term peripheral is used for tumors arising from the fibrous cap (2). Histological grading is in four grades based

on cellular atypia, mitotic figures, and cellularity. Grade 1 and Grade 2 sarcomas constitute 85% of the cases, while Grade 3 and dedifferentiated sarcomas are seen in 15%.

Conventional chondrosarcomas may develop primary and secondary to a pre-existing cartilage neoplasm. Cases with Mafucci's syndrome or Ollier's disease usually develop secondary central chondrosarcoma from a pre-existing enchondroma; however, patients with osteochondroma develop secondary peripheral chondrosarcomas (3). Chondrosarcomas primarily affect the bones of the extremities. This site accounts for more than 40% of cases, and the most common site is the long bones of the legs, followed by the pelvic bones and the long bones of the arms (4). Pelvic bones are the most often sites of secondary tumors, followed by the scapula and proximal femur (5).

Furthermore, chondrosarcomas are not restricted to the skeletal system. Malignant chondroid tumors can also occur in soft tissue. In our case series, there are frequent locations, such as the pelvis and femur, rarer localizations, such as the sternum, and very rare localizations, such as the lung/thorax wall and larynx. Pathological findings for different localizations are similar and graded with the same criteria. The predictive difference between localizations in tumors with similar morphological features is mainly related to the possibility of excision with safe surgical margins (6).

This study describes our institution's management and outcome experience for 12 chondrosarcoma patients with various pathologic subtypes and rarer locations. We demonstrate the outcome of a group consisted of higher grades of chondrosarcoma than previously mentioned in the literature.

MATERIALS AND METHODS

We conducted a retrospective cross-sectional study of patients diagnosed with chondrosarcoma at our institution between January 2013 and January 2023. We collected data on patient demographics, tumor location, tumor size, histological grade, treatment modalities, and outcomes.

Patients under 18 years of age, duplicate recorded, having their index surgery at an outside hospital, with a second primary malignancy were excluded from the study. Institutional Medical records of the remaining 12 patients were revisited for statistical analysis. Written consent was obtained from the patients and their next of kin/family where necessary (for deceased patients).

Demographic and clinicopathologic data were statistically analyzed by the SPSS 25.0 (IBM Corporation, Armonk, New York, United States) program. Quantitative variables were expressed as mean ± (standard deviation) and median (minimum-maximum) in the tables. In contrast, categorical variables were shown as n (%). The overall survival was defined by the time recorded from the biopsy until the date of death (event) or the last known follow-up without death has occurred. The time of surgical intervention until the date of local or systemic relapse (event) was used to calculate the relapse-free survival. Due to the small size of our cohort, a statistically powered analysis was not possible.

This study follows the articles of the Helsinki Declaration. Aydın Adnan Menderes University Medical Faculty Non-Interventional Clinical Trials Committee has approved the study design with protocol number 2023/126.

RESULTS

We identified a total of 12 patients with chondrosarcoma diagnosis. The median age at diagnosis was 62 (21-94) years. Of the 12 patients, six were male, and six were female. The primary tumor location varied; the most common site was the femur in four patients, followed by the larynx, left gluteal region, lung, periscapular soft tissue, and tarsal bone, distal metacarpal bone, scapula, and sternum each in one patient. All twelve patients were non-metastatic at their initial presentation. However, three developed local recurrence (chest wall-case III; ipsilateral femur and acetabulum-case X; and ipsilateral gluteal region-case XII), and three developed distant metastasis (contralateral lung-case I; lung-case II; bilateral lung and pleura-case IV) during the follow-up. The median tumor size was 5.65cm (1.5-21.5cm). Histologically, four patients had Chondrosarcoma (Grade I) (33.3%), one patient had Chondrosarcoma (Grade II) (8.3%), two patients had Chondrosarcoma (Grade III) (16.7%), two patients had Dedifferentiated Chondrosarcoma (Grade III) (16.7%), and three patients had Extraskeletal Mesenchymal Chondrosarcoma (25.0%) (Table 1).

All patients underwent surgical tumor resection (8 wide resections, one lobectomy, one laryngectomy, one amputation, and one curettage) with or without adjuvant chemotherapy or radiation therapy. Four patients received chemotherapy (two neoadjuvant chemotherapy and three adjuvant chemotherapy). Two patients received low dose (50.4Gy) (3DCRT-Case I; IMRT-Case XII), and two patients received high dose (>60Gy) (IMRT-cases III and VI) adjuvant radiation therapy.

The median follow-up time was 26.5 months (2-121 months). Case IX had the shortest follow-up. She died due to a cerebrovascular event two months after the oncologic surgery. Case II has the most extended follow-up and is currently relapse-free. Four patients had local recurrence, and three patients developed distant metastasis. Two distant metastases were located in the lung, but case IV developed extensive bilateral metastases on the lungs and pleura. Case I developed lung metastases two years after the initial diagnosis and died due to pulmonary complications. Case II developed pulmonary metastasis nine years after the initial diagnosis and underwent lobectomy. He is currently relapse-free and under follow-up. All patients started systemic chemotherapy.

The median survival of patients was 26.5 months (2-121 months). Median relapse-free survival was three months (0-96 months). The overall survival rate at two years was 81.5%, and at five years was 71.3%. The relapse-free survival rate at two years was 50%, and at five years was 25%. Individual treatment outcomes of patients are available in Table 2.

Table 1. Sociodemographic characteristics of patients

	n	%
Sex		
Female	6	50.0%
Male	6	50.0%
Primary tumor location		
Femur	4	33.3%
Larynx	1	8.3%
Left gluteal region	1	8.3%
Lung	1	8.3%
Periscapular soft tissue	1	8.3%
Right feet III digit	1	8.3%
Right IVth digit distal metacarpal bone	1	8.3%
Scapula	1	8.3%
Sternum	1	8.3%
Chemotherapy		
Adjuvant	3	25.0%
Neoadjuvant	2	16.7%
None	7	58.3%
Surgery		
Other	4	33.3%
Wide resection	8	66.7%
Pathologic diagnosis		
Chondrosarcoma (Grade I)	4	33.3%
Chondrosarcoma (Grade II)	1	8.3%
Chondrosarcoma (Grade III)	2	16.7%
Dedifferentiated chondrosarcoma (Grade III)	2	16.7%
Extraskeletal mesenchymal chondrosarcoma	3	25.0%
Adjuvant radiotherapy		
No	8	66.7%
Yes	4	33.3%
Relapse		
None	9	75.0%
Yes	3	25.0%
Metastasis		
None	8	66.7%
Yes	4	33.3%
Mortality		
Alive	8	66.7%
Exitus	4	33.3%
	Mean (SD)	**Median (Min-Max)**
Age	56.58 (18.48)	62 (21-94)
Tumor dimension (cm)	6.78 (5.59)	5.65 (1.5-21.5)
Relapse free survival (m)	14.00 (27.49)	3 (0-96)
Overall survival (m)	39.92 (34.06)	26.5 (2-121)

SD: standard deviation

Table 2 Individual characteristics and outcomes of the twelve patients

Case	Age	Sex	Tumor dimension (cm)	Primary tumor location	Status at presentation	Chemotherapy	Radiotherapy	Pathologic diagnosis	Recurrence	Relapse free survival (months)	Overall survival (months)	Final status
I	62	Male	8	Thorax	Non-metastatic	Adjuvant	Adjuvant	Mesenchymal chondrosarcoma	Systemic metastasis lung	7	24	Ex
II	62	Male	4	Femur	Non-metastatic	Adjuvant	Adjuvant	Extraskeletal mesenchymal chondrosarcoma	Systemic metastasis lung	96	120	Alive
III	64	Male	15	Lung	Non-metastatic	Neoadjuvant	-	Chondrosarcoma (Grade I)	Local chest wall	19	52	Ex
IV	40	Female	5	Sternum	Non-metastatic	Adjuvant	-	Chondrosarcoma (Grade III)	Local lung, pleura	6	26	Ex
V	63	Female	2	Right IVth digit distal metacarpal bone	Nonmetastatic	-	-	Chondrosarcoma (Grade I)	None	0	12	Alive
VI	21	Female	21.5	Femur	Non-metastatic	-	Adjuvant	Chondrosarcoma (Grade III)	None	0	27	Alive
VII	46	Male	1.8	Right feet III digit	Non-metastatic			Chondrosarcoma (Grade I)	None	0	36	Alive
VIII	50	Female	7	Scapula	Non-metastatic	-	-	Chondrosarcoma (Grade II)	None	0	26	Alive
IX	94	Female	8.5	Femur	Non-metastatic	-	-	Dedifferentiated chondrosarcoma (Grade III)	None	0	2	Ex
X	67	Male	3.8	Femur	Non-metastatic	-	-	Dedifferentiated chondrosarcoma (Grade III)	Local ipsilateral femur-acetabulum	10	42	Alive
XI	69	Male	1.5	Larynx	Non-metastatic	-	-	Chondrosarcoma (Grade I)	None	0	15	Alive
XII	41	Male	6.3	Left gluteal region	Non-metastatic	Neoadjuvant	Adjuvant	Extraskeletal mesenchymal chondrosarcoma	Local ipsilateral gluteal region	30	84	Alive

DISCUSSION

Chondrosarcomas are rare tumors of mesenchymal origin. They define a diverse group of tumors with different morphological characteristics and distinct clinical behaviors.

Chondrosarcomas are not restricted to the skeletal system. Malignant tumors can also occur in soft tissue. In addition to locations such as the pelvis and femur, which are frequently seen, there are rare localizations such as the sternum and very rare localizations such as the lung/chest wall and larynx in our case series. Pathological findings for different localizations are similar and graded with the same criteria. The prognostic difference between localizations in tumors with similar morphological features is mainly related to the possibility of excision with safe surgical margins (6). Adverse prognostic factors circumscribe older ages, bigger tumor sizes, and locations without extremities for conventional chondrosarcomas (7).

Macroscopically, chondrosarcomas are large tumors, mostly more prominent than 5 cm (8). Our case series' median macroscopic tumor dimension was 5.65cm (1.5-21.5cm). Larger tumors in our case series were also associated with higher tumor grades, therefore, metastases (case I and III) or near-positive surgical margins (case VI). Chondrosarcomas have lobular bluish-gray or white sections consistent with hyaline cartilage. (Figure 1) There may be areas containing myxoid or cystic changes. Yellowish-white scale deposits with mineralization are often present. Erosion and destruction of soft tissues can be observed (9).

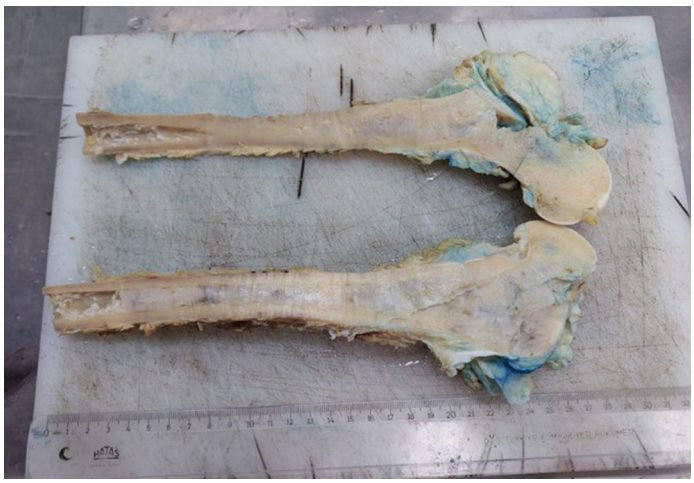

Figure 1. Macroscopic view of chondrosarcoma

Microscopically, on conventional chondrosarcomas, atypical chondroid cells are observed. The term atypical is used for irregular size and shape. Binucleated nuclei (Figure 2) and necrosis can be seen. Liquefaction of the cartilage matrix and myxoid changes in higher-grade chondrosarcomas are common (1). These tumors are categorized by their histologic grade: Grade 1 (Low grade) -atypical cartilaginous tumor- tumors usually very similar to normal cartilage or benign chondroma. The cellularity is not high, with chondroid cells having binucleated nuclei that are mostly not extended. There is no mitotic map. While the stroma cartilage area forms a large array, myxoid changes are usually not observed. (Figure 3). Grade 2 (Intermediate grade) tumors have higher cellularity, resulting in a decreased cartilage matrix. The stroma of the chondroid tissue can be more often myxoid. The nuclei are moderately sized. Mitotic activity is usually fewer than two on 10 High Power Fields. Chondrocytes may have giant hyperchromatic or vesicular nuclei. Chondroid cells with two or more nuclei are typical. (Figure 4). Cellularity is so high on grade 3 (high grade) tumors that the cartilage matrix is very sparse, and myxoid fields predominate. Chondroid cells are irregularly spindle-shaped.

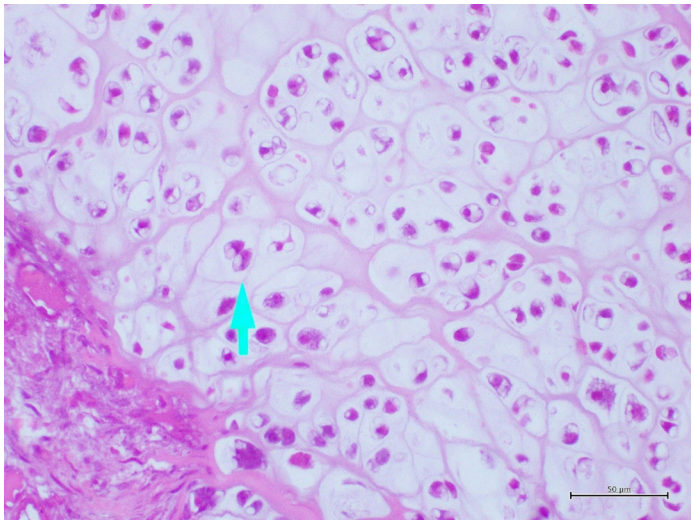

Figure 2. Typical binuclear cells

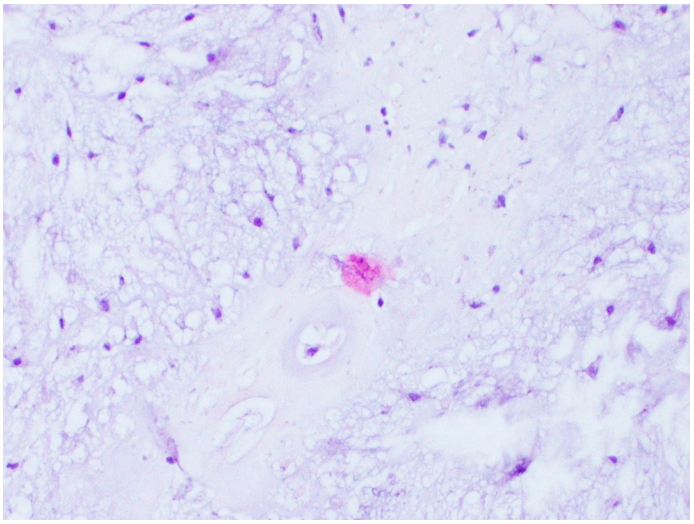

Figure 3. Grade 1 chondrosarcoma

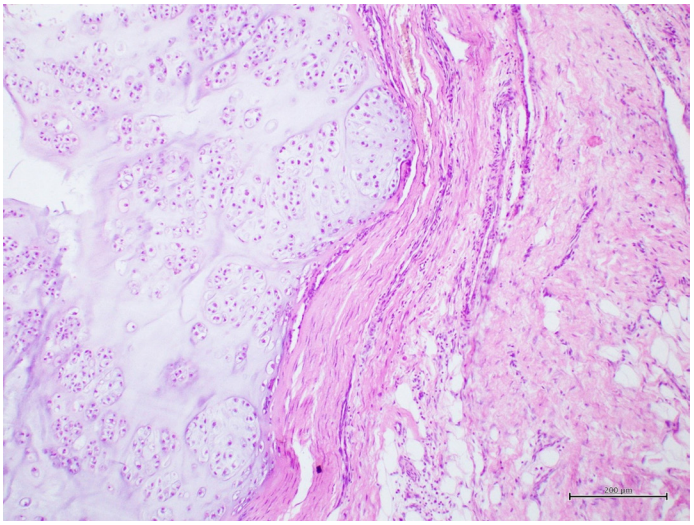

Figure 4. Grade 2 chondrosarcoma

Nuclei are pleomorphic and can be 7-8 times larger than average. Mitotic figures resembling necrotic areas are often (2) (Figure 5,6). Grade 1 and Grade 2 sarcomas constitute 85% of the cases, while Grade 3 and dedifferentiated sarcomas are seen in 15%. It is essential to thoroughly understand different histological subtypes since this information is beneficial in predicting the biological behavior of chondrosarcomas (10). While four of the 12 cases diagnosed in our center were grade I, only one was grade II. We had four grade III cases, two of which were in the dedifferentiated group. Our high-grade case rate was higher than the literature.

A fourth-grade group of chondrosarcomas, known as dedifferentiated chondrosarcomas, accounts for 10% of all chondrosarcomas. Dedifferentiation is seen in a small proportion of cases and is defined histologically by high-grade, usually spindle-shaped or pleomorphic tumors without a prominent cartilaginous stroma (Figure 7) (1). Dedifferentiated chondrosarcomas are commonly seen in sixth-decade or older adults, with equal distribution in both sexes and favor the femur as location (11,12). In our series, we have two cases (cases IX and

X) diagnosed with dedifferentiated chondrosarcoma, which align with the literature regarding the abovementioned characteristics.

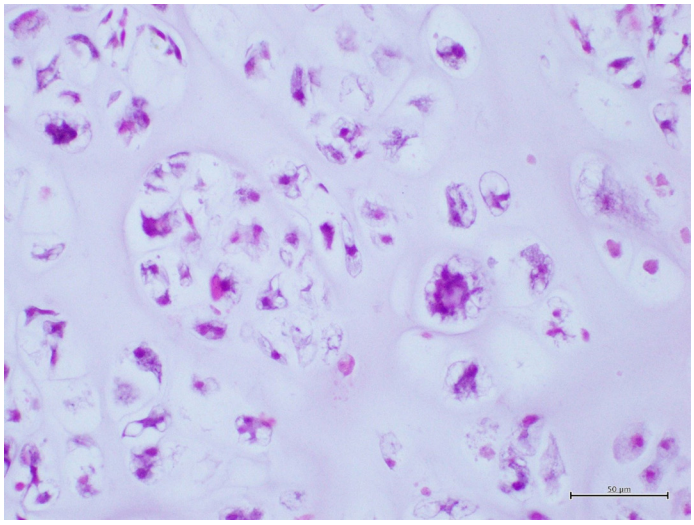

Figure 5. Grade 3 chondrosarcoma

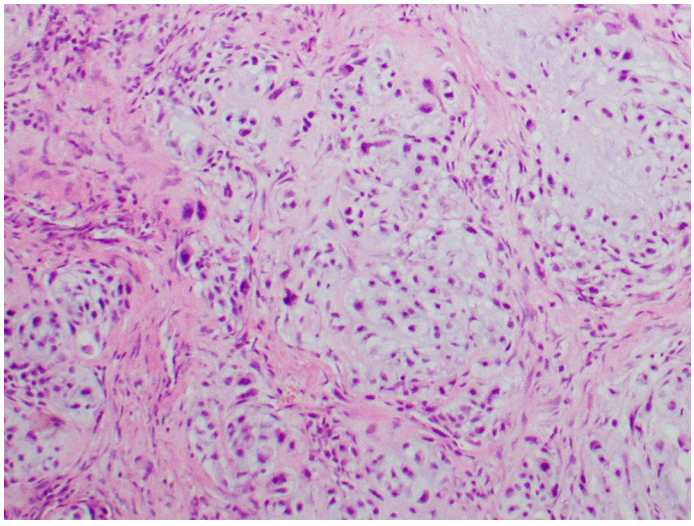

Figure 6. Grade 3 chondrosarcoma on thorax wall

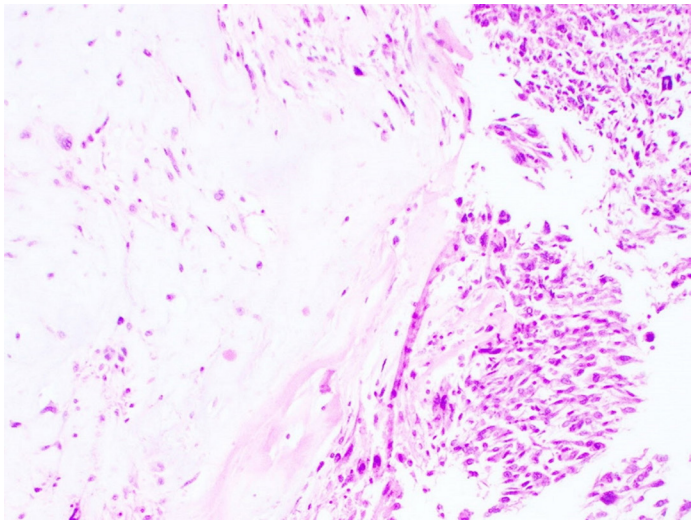

Figure 7. Dedifferentiated chondrosarcoma

In addition, we have three cases (cases I, II, and XII) diagnosed with mesenchymal chondrosarcoma, all of which are of extraskeletal origin. In mesenchymal chondrosarcoma, one of the subtypes other than the conventional group that constitutes most cases, a biphasic tumor structure with small-medium size, poorly differentiated round cells with increased nucleus/cell ratio, and well-differentiated hyaline cartilage areas with a characteristic staghorn vascular pattern is seen.

Immunohistochemical stains are not necessary in the majority of cases. Tumor cells strongly express S100 and D2-40 except in less differentiated areas (13). Pancytokeratin is negative. The p. Arg132His-IDH1 mutation-specific antibody is of limited utility because it recognizes only a small subset of IDH mutations in low-grade chondrosarcomas (14).

The histological features of chondroma, distinguished by higher cellularity, irregular cellularity, and the presence of binucleated cells, overlapped with those of low-grade chondrosarcoma. One of the most critical points is the growth pattern, which contains the lamellar bone that is already present.

There is an overlap between the morphological features of enchondroma and the morphological features of grade 1 chondrosarcoma. A distinction is made in grade 1 chondrosarcoma by increased cellularity, irregular formation of cells, and binucleated cells. One of the most important criteria is the growth pattern, in which the presence of entrapment of pre-existing lamellar bone and the absence of encasement indicate low-grade chondrosarcoma.

More than 20 percent of myxoid changes in the stroma also support the diagnosis of atypical chondroid tumor/grade 1 chondrosarcoma with an enchondroma. A thorough distinction in small biopsy specimens can be difficult, and clinicopathologic correlation is critical. Lesion location and patient age are essential if these criteria are used (15).

Based on morphology alone, it is impossible to distinguish osteochondroma from secondary peripheral atypical chondroid tumor/grade 1 chondrosarcoma. Cystic degeneration, nodular formation, or necrosis have not been shown to indicate progression. The distinction should be made in a multidisciplinary setting where the strength of the cartilage cap is most important. In grade 2, secondary peripheral chondrosarcoma, mitosis, and small pleomorphism can be seen, neither of which is present in ordinary peripheral atypical chondroid tumor/grade 1 chondrosarcoma (16).

Skull base chondrosarcomas should be differentiated from chordomas, including the chondroid subtype. Grade 3 chondrosarcoma is similar to chondroblastic osteosarcoma. Periosteal osteosarcoma is included in the differential diagnosis. Peripheral chondrosarcomas are distinguished from central chondrosarcomas by their location on the bony surface.

The most common point mutations in chondrosarcoma occur in the genes IDH1 and isocitrate dehydrogenase 1 and 2 (IDH1-

IDH2). Mutations in the IDH1 and IDH2 genes have been described in various hematopoietic malignancies and half of the conventional chondrosarcoma cases. These two mutations are also associated with cases with low overall survival. The IDH mutations can also be used in the differential diagnosis of high-grade/dedifferentiated chondrosarcomas and undifferentiated pleomorphic osteosarcoma or chondroblasts osteosarcomas (14).

Other often mutated genes in chondrosarcomas include TP53, MDM2, and CDK4 (15). Additionally, many genes in normal biologic pathways were overexpressed in chondrosarcomas. For example, AKT regulates cell survival, proliferation, growth, and glycogen metabolism (16).

Next-generation sequencing was applied to the samples from two patients in our case series. In our case with lung-chest wall tumor, variants were detected in FANCD2 (pathogenic), KMT2C (pathogenic and possibly pathogenic), AKT1, and FGFR (unclear) genes. In our case of chondrosarcoma located in the sternum, possible pathogenic variants were found in the KIT, TSC2, and KMT2C genes.

Although these genes are not the most frequently mutated, some are being investigated for targeted therapy in various tumors. For example, AKT1 inhibitors targeting the Phosphoinositide 3-kinase pathway are in clinical trials for treating chondrosarcoma (17). The KMT2C gene, which we found a pathogenic variant, is being investigated for immunotherapy efficacy in lung small-cell carcinoma (18). New studies involving larger case groups should support existing data.

Outcomes of chondrosarcoma patients depend on grade, stage, and surgical margins because higher grade shows (10,19,20) decreased patient survival. Based on a recent meta-analysis, survival with different histologic grades was as follows: 5-year survival for chondrosarcomas grade 1 was 82% to 99%, 2-63% to 92%, and 3-0% to 77% (6). The median survival of patients in our series was 26.5 months (2-121 months). The overall survival rate at two years was 81.5%, and at five years was 71.3%. In this context, our results concord with the literature (7,20,21). Furthermore, ten-year survival rates according to the grades were reported as 80-99% for Grade I, 58-86% for Grade II, and 0-55% for Grade III (6,20,22).

The relapse-free survival rate at two years was 50%, and at five years was 25%. Fromm et al. reported their five-year relapse-free survival as 75%, with 24% relapses in the first year. Histologically, 43% of their patients were Grade I, and 47% were Grade II, so their series were mostly low-grade tumors, contrasting with ours (23). Our relapse-free survival rates were also concordant with the literature (6,20). Studies in the last 30 years relating to treating and managing chondrosarcoma have recently reached stability with no more promising survival improvement (24).

Management of patients with chondrosarcoma is closely associated with histologic subtypes; therefore, it is clinical behavior (10). Despite the continuous development of novel therapeutic approaches, findings still imply surgical treatment's clinical and practical importance in the long term (25). Complete wide surgical excision of the tumoral mass (including intralesional curettage and cryotherapy) (26) as much as possible is favored. Thus, the primary aim is to achieve clear surgical margins.

Chondrosarcomas are known for their radioresistance and chemoresistance due to the extracellular matrix, low percentage of dividing cells, and poor vascularity (27). Using (neoadjuvant and adjuvant) chemotherapy and radiotherapy may provide additional benefits in select cases such as mesenchymal and dedifferentiated chondrosarcomas. Moreover, as Catanzano et al. advocated, irradiation should be strongly considered in patients whose tumors are located at surgically-hard-to-reach points (such as the base of the skull, spinal cord, and pelvis) to obtain negative surgical margins or will cause incompatible morbidity (28). A retrospective series from Princess Margaret Hospital underlines that prescribing postoperative low doses (40-50Gy) of radiotherapy improves local control (29). In our case series, four patients (cases I, III, VI, and XII) received adjuvant radiotherapy due to tumor dimensions, pleural invasion, and near-positive surgical margins. Radiation therapy doses were 50.4Gy in two, while total doses for the other two were |>60Gy. One patient was treated with 3D conformal radiotherapy (3DRT), and three others were with intensity-modulated radiotherapy (IMRT).

Technical advances in radiation oncology have provided us with tumoral dose escalation for better late side effect control. This results in more successful management in selected high-risk chondrosarcoma patients with tumors in surgically challenging locations or for unplanned positive margins. Furthermore, advanced radiotherapy techniques (IMRT, proton beam radiotherapy, heavy ion radiotherapy, and stereotactic radiotherapy) enable overall survival benefits (28).

Limitations

Although our cases' follow-up results are promising, this study has some limitations. This is a small case series; therefore, we could not perform a powered statistical analysis. In future studies with a bigger sample size and better insights into the estimation of overall survival and relapse-free survival rates, the relationship between pathologic and genetic risk factors could be elucidated. Furthermore, the retrospective design of this study precludes the impact of adjuvant therapies on index oncologic surgery. More extensive controlled trials are needed to understand treatment modalities' associations better and enable stronger extrapolations for clinical outcomes.

CONCLUSION

Chondrosarcoma is a rare malignant tumor that primarily affects bones and typically presents in middle-aged patients. Surgical resection is the mainstay of treatment, with adjuvant chemotherapy or radiation therapy used in some instances. The long-term prognosis is generally favorable, with a five-year survival rate of 71.3%.

Conflict of Interests
The authors declare that there is no conflict of interest in the study.

Financial Disclosure
The authors declare that they have received no financial support for the study.

Ethical Approval
Aydın Adnan Menderes Univesity Medical Faculty Non-Interventional Clinical Trials Committee has approved the study design with protocol number 2023/126.

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