Management in Low-Grade Chondrosarcoma

An ESUN Editorial

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Editor's Note: This editorial is part of an ongoing series of Op Ed pieces written by one of the members of the Board of Directors of the Connective Tissue Oncology Society (CTOS). Dr. Randall is a member of the CTOS board. These editorials are intended to address important and controversial issues in the field. The "Questions, Comments & Counterpoint" column allows readers to express their opinions in response to these Op Ed pieces. Click here to send in an opinion.

Coordinating Editor's Note: There are many controversies in sarcoma. I summarized a number of them in my CTOS Op Ed Controversies in Sarcoma in the April 2009 issue of ESUN. One of the most controversial subjects in musculoskeletal tumor surgery is how to manage low grade chondrosarcomas. I felt that this subject warranted extra time and space to discuss this very difficult problem of how to manage low grade chondrosarcomas, and the center of this controversy is based on the pathology. It is the grading of this disease that makes it so challenging and the final diagnosis is dependent on the clinical, radiologic and pathologic findings in order to make that final diagnosis which has been so well presented here in this extended Op Ed piece by Drs. Novais and Randall. Doug Letson, MD, Coordinating Editor of the CTOS Op Ed series.

What is chondrosarcoma?

Chondrosarcoma is a malignant cancer whose tumor cells produce a pure hyaline cartilage that results in abnormal bone and/or cartilage growth. The term chondrosarcoma is used to define a heterogeneous group of lesions with diverse morphologic features and clinical behavior.

Cartilage is a type of dense connective tissue. It is composed of cells called chondrocytes which...
Who gets Chondrosarcoma? Is it common?

Chondrosarcoma is a rare disease, with an estimated incidence of 1 in 200,000 per year (1). It is the third most common primary bone cancer and it is estimated that they accounted for 3.6% of the annual incidence of all primary bone malignancies in the USA in 2006, after multiple myeloma and osteogenic sarcoma (2). Chondrosarcomas represented 9.2% of the malignant tumors in patients at one single institution and approximately 86% of these were primary chondrosarcoma. (3). Classically, chondrosarcoma have been described with a slight preference for male patients (4). However in a recent analysis of the SEER Database (Surveillance, Epidemiology and End Results) a higher proportion of appendicular chondrosarcomas was found in women (60.9%) than in men (46.7%) and a lower proportion of axial chondrosarcomas was found in women (30.3%) than in men (40.4%). The age distribution of patients with chondrosarcoma shows a gradual age-related increase, with the peak incidence occurring during the sixth and seventh decades of life. In an analysis of the National Cancer Data Base (NCDB) of the American College of Surgeons over 70% of reported cases were in patients aged 40 years and older.

Chondrosarcoma is not contagious. It cannot be passed on to another person by exposure to a chondrosarcoma patient. Although specialists are not yet certain what causes chondrosarcoma, there are several factors that put people at a higher risk.

Certain hereditary conditions may make people more susceptible to chondrosarcomas. These include Ollier's Disease, Maffucci Syndrome, and Multiple Hereditary Exostoses (MHE, a.k.a., osteochondromatoses), People affected by these conditions are at a higher risk because they usually develop several benign bone tumors (sometimes called bone spurs in the case of MHE), which have a higher chance of becoming malignant. People with these hereditary conditions who experience sudden growth spurts or increases in hormone production, such as pregnancy, have a slight increased risk of a benign bone tumor changing into a chondrosarcoma. These patients should be followed by a bone tumor specialist for all of their lives. However, most patients with chondrosarcoma do not have any of these genetic conditions. Adults with Paget's disease, a non-cancerous condition characterized by abnormal development of new bone cells, may be at increased risk for chondrosarcoma. When chondrosarcoma occurs in children and young adults, it is often in patients who have had radiation or chemotherapy treatments for other conditions.

What are the different types of chondrosarcoma?

Chondrosarcomas are actually a heterogeneous group of tumors. They are classified as primary (or conventional chondrosarcoma) if they are unassociated with a pre-existing lesion and secondary if they develop from a pre-existing chondroid lesion, such as enchondroma or osteochondroma. More than 90% are designated conventional chondrosarcomas. They can be further sub classified as central when they arise from within the medullary cavity and peripheral...
when they arise from the surface of the bone. Conventional chondrosarcoma are nearly always central; secondary chondrosarcomas can be central but usually they are peripheral. They can also be classified by their location within the body (i.e., axial skeleton versus appendicular skeleton), whether it fits into a specific histological variant (i.e., clear cell, myxoid, mesenchymal, dedifferentiated).

The single most important factor to consider when evaluating the malignant potential of a chondrosarcoma is its cytologic and histologic grade, determined by the appearance of tumor material under the microscope combined with the clinical and radiologic presentation (Table 1) (5-10).

<table>
<thead>
<tr>
<th>Grade</th>
<th>Tumor</th>
<th>Symptoms</th>
<th>Prognosis</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign</td>
<td>Enchondroma</td>
<td>Usually no symptoms</td>
<td>Excellent</td>
<td>Surveillance, intralesional excision if symptomatic</td>
</tr>
<tr>
<td>Malignant (Low Grade)</td>
<td>Grade I Chondrosarcoma</td>
<td>60% are painful</td>
<td>Good</td>
<td>Controversial: Extended intralesional excision vs. rarely wide resection</td>
</tr>
<tr>
<td>Malignant (Intermediate Grade)</td>
<td>Grade II Chondrosarcoma</td>
<td>Up to 80% are painful</td>
<td>Fair</td>
<td>Wide resection</td>
</tr>
<tr>
<td>Malignant (High Grade)</td>
<td>Grade III Chondrosarcoma</td>
<td>Up to 80% are painful</td>
<td>Poor</td>
<td>Wide resection. Chemotherapy and radiation therapy in select cases</td>
</tr>
<tr>
<td>Malignant (High Grade)</td>
<td>Dedifferentiated Chondrosarcoma</td>
<td>Most are painful</td>
<td>Poor</td>
<td>Wide resection. Chemotherapy and radiation therapy in select cases</td>
</tr>
<tr>
<td>Malignant (High Grade)</td>
<td>Mesenchymal Chondrosarcoma</td>
<td>Pain and swelling</td>
<td>Poor</td>
<td>Wide resection. Chemotherapy and radiation therapy in select cases</td>
</tr>
</tbody>
</table>
While grading is very important in the management of chondrosarcoma, it can be contentious. Although several grading systems have been described, usually they are graded on a scale of 1-3. The grading is based primarily on nuclear size of tumor cells, nuclear staining (hyperchromasia, or darker staining of nuclear material) and cellularity (11). Approximately 90% are low grade to intermediate-grade tumors (grade 1 or 2) and only 5–10% of conventional chondrosarcomas are grade 3 lesions.

- Grade I (low grade) tumors are moderately cellular and contain hyperchromatic plump nuclei of uniform size and the cytology is very similar to enchondroma (a benign cartilage tumor). Occasionally binucleated cells may be seen.

- Grade II (intermediate grade) are more cellular with a greater degree of nuclear atypia, hyperchromasia and nuclear size.

- Grade III (high grade) tumors have significant areas of marked pleomorphism, large cells with more hyperchromatic nuclei than grade II, occasional giant cells and abundant necrosis. Mitoses are frequently detected.

Chondrosarcoma variants are rare and include the following:

- Mesenchymal chondrosarcoma is a highly malignant tumor characterized by the presence of solid, highly cellular areas composed of round or slightly spindled primitive mesenchymal cells and is histologically similar to Ewing's sarcoma with foci of cartilaginous differentiation. This is a very aggressive type of chondrosarcoma with a high risk of local recurrence and distant metastasis.

- Dedifferentiated chondrosarcoma contains two clearly defined components, a well differentiated cartilage tumor, either an enchondroma or a low grade chondrosarcoma, juxtaposed to a high grade noncartilaginous sarcoma. It is typically thought of as a tumor in which a low grade chondrosarcoma or benign cartilage tumor transforms into a high grade sarcoma with features of osteosarcoma, fibrosarcoma, or malignant fibrous histiocytoma. Radiographically the tumor produces an ill defined, lytic, interosseous lesion associated with cortical disruption and extension into the soft tissues. They are aggressive neoplasms and have a dismal prognosis.

- Clear cell chondrosarcomas are low-grade tumors with significant amounts of glycogen. They typically involve the epiphyseal end of a long bone. Histologically, cells have abundant clear cytoplasm embedded in a loose hyaline cartilaginous matrix and an infiltrative growth pattern. Radiographs show a lytic defect at epiphyseal end of long bones that is sharply demarcated with sclerotic margins. They carry a low recurrence rate and a good prognosis with wide resection.

- Myxoid chondrosarcoma is a slow-growing tumor characterized by prominent myxoid degeneration histologically and prolonged course, despite a high incidence of local recurrence and metastasis clinically.

**What is known about the genetics of chondrosarcoma?**
As evolving molecular techniques are available, several genotypic and phenotypic markers for chondrosarcoma have been tested to see if they assist in determining tumor grade prognosis. There is considerable complexity and heterogeneity in the pathologic and clinical behavior of chondrosarcomas. This is reflected in the diversity of cytogenetic and molecular genetic characteristics that have been described in these tumors. Please see Sandberg and Bridge (2003) (12), Sandberg (2004), and Bovee et al. (2005) (13) for a thorough review.

The genetic changes specific to chondrosarcoma continue to be investigated extensively. Although studies have not yet established a specific or recurrent karyotypic feature for any of these tumors, different chondrosarcomas have demonstrated anomalies in several tumor suppressor genes, oncogenes, and transcription factors, including TP53, RAS, EXT1, EXT2, and Sox9. Available cytogenetic and comparative genomic hybridization (CGH) studies reveal changes in some chondrosarcomas, but fail to do so in others. These studies are thus far difficult to interpret.

Based on the available studies, it is likely that chondrosarcomas are generated by a coordinated, multi-step process involving primarily tumor suppressor genes. In fact, the complexity and variety of genetic changes seen in chondrosarcomas may indicate several distinct genetic pathways. Some of the same genes may be involved in each, but the order and manner in which they are affected may differ among chondrosarcomas. Establishing the genes that initiate the neoplastic processes, and that are subsequently involved along the pathways leading to chondrosarcoma may lead to therapies addressing these molecular changes, as has been accomplished for several other sarcomas.

**How can one differentiate a low grade chondrosarcoma from a benign cartilage tumor?**

It is important to understand the difference between a benign and malignant cartilage tumor. Chondrosarcoma is a sarcoma, or malignant tumor of connective tissue. An enchondroma is a benign bone tumor. Benign bone tumors are not sarcomas. Benign bone tumors do not spread to other tissues and organs, and are not life threatening. They are generally left alone or cured by surgical removal if they cause symptoms such as tenderness via pressure on surrounding muscles, tendons or nerves. Low grade chondrosarcoma also has relatively little potential to spread but can recur locally if not treated appropriately. If these tumors due recur they can start to behave more aggressively.

Distinguishing chondrosarcoma from its benign counterpart, enchondroma, is crucial to the patient treatment and prognosis but can be difficult at times. The diagnosis of cartilage lesions requires expert evaluation. First, clinical and radiographic features such as the age of the patient, symptoms, localization in the skeleton, and the pattern of bone destruction or mineralization should be scrutinized. The role of biopsy in low grade lesions is quite contentious as sampling errors are a distinct possibility. A wait and see approach is generally recommended at first. If symptoms worsen or there is a progression on radiographic examination then consideration for intralesional surgical treatment may be indicated. Because of the possibility that the tumor is a low grade chondrosarcoma, it should be removed by an experienced musculoskeletal oncologist at a designated sarcoma center with expert pathology and radiology support. The removed specimen must be thoroughly evaluated for features concerning for malignancy (14, 15).
Distinguishing between enchondroma and chondrosarcoma is one of the most frequent diagnostic dilemmas facing orthopaedic oncologists and their colleagues in diagnostic radiology and pathology (16).

• **Clinical Presentation**

Patients with chondrosarcoma experience a wide range of clinical courses, from slow insidious tumor growth over years in low grade lesions to rapid neoplastic progression, metastasis, and death in higher grade lesions. The vast majority of chondrosarcomas is low grade and accordingly is very slow to progress. Clinical symptoms may be helpful in the initial evaluation of a cartilaginous tumor. Pain is more common with chondrosarcoma than with the benign enchondroma. In a review from Murphey et al 95% of patients with chondrosarcoma experienced pain (17). Eefting et al reported that 35% of patients with enchondromas and 62% of the patients with central grade I chondrosarcoma presented with spontaneous pain. It has been reported that in patients with grade I chondrosarcoma, 60% have night pain or rest pain, 21% have vague regional pain, and only 19% have painless tumors (18). This difference is significant, although presentation without pain does not exclude malignancy and should not delay additional analysis (19). Pathologic fractures occur in 3% to 8% of patients with chondrosarcoma (18).

• **Imaging**

**Radiographs** - Enchondromas and low grade intramedullary chondrosarcomas of long bones can have similar radiologic appearances. Both types of tumors demonstrate stippled calcifications, and both may display endosteal scalloping on plain radiographs (20). Calcification is manifested by punctuate mineralization or popcorn like calcification (Figure 1).

![Figure 1](http://sarcomahelp.org/low_grade_chondrosarcoma.html)

The margins of the tumor should be examined for osteolysis and endosteal scalloping.
Chondrosarcoma can demonstrate adaptive and aggressive radiologic signs. Cortical expansion and thickening are adaptive changes, and cortical disruption and soft-tissue masses are aggressive changes associated with chondrosarcoma. The extent and degree of endosteal scalloping correlate with the likelihood of the lesion being a chondrosarcoma. The imaging characteristics that should suggest chondrosarcoma are endosteal scalloping depth and extent (greater than two-thirds of cortical thickness and along more than two thirds of the lesion), extent of matrix mineralization (within less than two-thirds of the lesion as seen on radiographs), presence of cortical remodeling or destruction and thickening, periosteal reaction, pathologic fracture, and associated soft-tissue mass (17). There is an overlap in the radiographic size of both, however a lesion larger than 5-6 cm in diameter are much more likely to represent a chondrosarcoma.

Anatomic tumor localization is different but should not be considered of diagnostic value. Enchondromas are most common in the hands or feet while chondrosarcomas are common in the axial skeleton (spine and pelvis), typically with large associated soft-tissue masses. The ilium is the most frequently involved bone followed by the proximal femur, proximal humerus, distal femur and ribs.

**Advanced imaging**

**Computed Tomography (CT)** – CT is helpful in detecting more details of the lesion. Enchondromas as a rule should not progress during adulthood and any change in size or radiographic appearance of an enchondroma should be considered as a red flag for the presence of a malignant tumor. CT is superior to radiography for detecting focal areas of scalloping and is considered the best modality to detect mineralization characteristic of a chondroid neoplasm. Murphey et al reported that all enchondromas showed evidence of calcification on CT scans but only 95% revealed these areas on radiographs (17).

**Magnetic Resonance Imaging (MRI)** – MRI is particularly useful in determining the nonmineralized intramedullary extent of the tumor and soft-tissue extension (Figure 2).

![Figure 2](http://sarcomahelp.org/low_grade_chondrosarcoma.html)
performed. Greater than 90% medullary involvement can be suggestive of chondrosarcoma, while the absence of 90% medullary involvement of non-contiguous areas of cartilage within the bone can suggest the presence of an enchondroma. The relationship of a soft-tissue mass to important paraosseous structures, such as the joint capsule and the neurovascular bundle, is accurately demonstrated on MR images. Fast contrast-enhanced MR imaging has the potential to help differentiation between enchondroma and chondrosarcoma. MR imaging results should be seen as an additive tool and may not be considered alone. The use of gadolinium-enhanced MR imaging adds substantially to the characterization of cartilaginous tumors. The timing and progression of gadolinium contrast enhancement patterns may help direct a clinician toward or away from a diagnosis of malignancy. Early enhancement (within 10 seconds of arterial enhancement) may be seen in chondrosarcoma but not in enchondroma. Differentiation of malignancy on the basis of early and exponential enhancement has been demonstrated to have a sensitivity of 61%, specificity of 95%, positive predictive value of 92% and negative predictive value of 72% (21). Gadolinium-enhanced MR imaging shows septal enhancement in low-grade chondrosarcomas, corresponding to fibrovascular septation between lobules of hyaline cartilage. This feature is usually absent in benign cartilaginous lesions (22, 23).

**Bone Scan** - This test works by injecting a small amount of radioactive material into the bloodstream and taking images using a gamma camera to detect uptake of radioactive material. Bone scintigraphy has limited application in the differential diagnosis of tumors with low biologic activity such as Grade-I chondrosarcoma. A whole-body bone scan with a high degree of radionuclide uptake within the lesion compared with an internal standard, such as the anterior superior iliac spine or acromioclavicular joint, has been considered as more consistent with chondrosarcoma than enchondroma (17) (Figure 3). Great caution should therefore be used in drawing conclusions from bone scan results, but these results can add to the overall picture, and better inform the decision making process.
**PET Scan** - Recently, there has been some research into the use of a specialized radiographic test called fluorine-18 fluorodeoxyglucose positron emission tomography (FDG PET) for grading of tumors in patients with chondrosarcoma. Preliminary studies showed that (FDG PET) could be an objective and quantitative adjunct in differentiating chondrosarcomas from enchondromas and osteochondromas and in assessing the grade of chondrosarcomas (24). In a more recent study, FDG PET imaging was found a useful parameter for tumor grading and prediction of outcome in chondrosarcoma patients (25). However, the measurement of glucose metabolism by positron emission tomography alone cannot distinguish between benign and grade-I malignant cartilaginous tumors. It is important to understand the advantages and disadvantages of imaging modalities for accurate interpretation of results. Although positron emission tomography has limitations, it may be useful for predicting high-grade chondrosarcomas from benign chondroid lesions and grade-I chondrosarcoma (26).

**What if a low grade chondrosarcoma is suspected?**

After all of the imaging modalities are obtained, an experienced orthopaedic oncologist must decide whether to observe the patient or intervene. If a low grade chondrosarcoma is suspected, then an aggressive curettage is indicated. When a more aggressive lesion is suspected, a sampling biopsy is indicated as surgical excision is more extensive and so the diagnosis of a high grade cancer should be established. When fresh tissue from a chondrosarcoma is viewed under a microscope after a biopsy, lower grade chondrosarcomas will exhibit increasing amounts of relatively acellular cartilage stroma as well as regions of modestly increased cellularity. By contrast, higher-grade lesions tend to harbor regions of densely packed hyperchromatic malignant looking cells.

Histologic features to distinguish benign from malignant cartilaginous tumors were advocated in the past although the diagnostic value of individual morphologic criteria or how they are handled in clinical practice is controversial (27). Recent studies have tried to establish a more precise standard method for distinction of cartilage neoplasms in long bones. The American Skeletal Lesions Interobserver Correlation among Expert Diagnosticians (SLICED) Study Group showed that radiologic and pathologic interpretations of cartilaginous lesions in long bones are not optimally reliable, even among specialized experts (28). Another recent study reported that distinction between a high grade chondrosarcoma seems to be less problematic, however differentiation of grade I chondrosarcoma from enchondroma remains a diagnostic challenge. In that study there was considerable interobserver variability, even among expert bone tumor pathologists, in the histologic diagnosis of enchondroma and low-grade chondrosarcoma. In an attempt to distinguish between low-grade chondrosarcoma and enchondroma, the authors identified four most important histologic parameters, as following: host bone entrapment, high cellularity, marked nuclear pleomorphism, and irregular distribution of cells. The differential diagnosis could be assessed with a higher great degree of accuracy if the biopsy specimen contained a combination of: presence of host bone entrapment, open chromatin, mucoid matrix egeneration, and the patient’s age was above 45 years (19).

**What are the current options of treatment for low grade chondrosarcoma?**
The management of low grade chondrosarcoma can be a challenge. Grade I chondrosarcomas may not all behave in the same way clinically or radiographically. In some cases a painful cartilaginous lesion in a long bone has the radiologic appearance of a low-grade chondrosarcoma (e.g., lytic areas or high-grade endosteal scalloping without adaptive or aggressive radiographic changes) and the histologic appearance of an enchondroma. This borderline type of lesion has been referred as grade 0.5 chondrosarcoma, atypical enchondroma, grade 0 chondrosarcoma and chondrosarcoma in situ by different authors (9, 13, 18). The decision making process about the management of these lesions should involve the following considerations:

- **Intralesional excision** grossly removes the tumor but conceivably leaves microscopic and macroscopic tumor in the tumor bed. Intralesional excision with some sort of adjuvant therapy (e.g. liquid nitrogen, argon beam, hydrogen peroxide, phenol) has the potential benefits of adjacent bone and joint preservation and better functional outcome.

- **Wide resection** includes a cuff of normal tissue surrounding the tumor completely. Depending on the location, wide resection may cause increased morbidity and require complex reconstruction. Generally, this is reserved for higher grade lesions.

The key for long term successful treatment of these lesions is to adequately identify those patients who would benefit from less aggressive surgical resection without the risks of local recurrence and distant metastasis.

The local recurrence rate and potential for metastasis in low-grade chondrosarcoma are low, so limited surgery (intralesional resection) with adjuvant therapy has been advocated for less aggressive-appearing lesions (5, 7, 8, 14, 18). Leerapun et al (14) recently report that intralesional excision with adjuvant therapy only works well in confined bony defects. In their series, tumors that had no associated cortical perforation or soft tissue mass were successfully treated with intralesional curettage. Patients with more aggressive lesions (eg, cortical disruption, Stage IB) were selected for en bloc resection and had excellent local control. They excluded lesions of the central skeleton because these historically behave differently from long bone lesions. Marco et al described a subset of patients with low-grade chondrosarcoma that could be treated with intralesional excision with adjuvant therapy without compromise of the oncologic outcome: painful, intramedullary low-grade chondrosarcoma (stage IA) of the appendicular skeleton, which can demonstrate a high degree of endosteal scalloping without adaptive or aggressive radiologic signs. The only local recurrence in their series was in a patient with cortical disruption and expansion, as well as a soft tissue mass. Bauer (5) reported on patients with low-grade intramedullary chondrosarcoma of a long bone treated by intralesional excision and followed for 2-25 years. The authors concluded that a central grade I chondrosarcoma of long bones can be treated with curettage and filling with either bone graft or bone cement (methylmethacrylate) but distal more radiographic aggressive lesions requires en bloc resection. Lesions of the pelvis and shoulder girdle should also be treated aggressively since they have a high risk of recurrence. In another recent study Van Der Geest et al (29) reported excellent oncological and functional results the use of cryosurgery as an adjuvant in the surgical treatment of active or aggressive enchondromas and chondrosarcomas grade I. Post-operative fracture was seen as the most common complication in their series.
Most authors agree that adequate surgical margins lower the risk of local recurrence in patients with chondrosarcoma. (7, 8, 14, 18, 30). Wide resection should be implemented for the treatment of low grade chondrosarcoma in a long bone if it shows radiographic signs of adaptive changes (cortical expansion or thickening) or aggressive changes (cortical disruption, stage IB) and if it's associated with a soft tissue mass. Wide resection is recommended for virtually all low grade chondrosarcomas of the pelvis and sacrum because in these locations there is a higher chance of local recurrence and metastasis (14, 31, 32).

Is there a need to further treatment of low grade chondrosarcoma by radiation or chemotherapy?

For higher-grade tumors, with a worse prognosis for recurrence and metastasis, adjuvant therapies may be considered. Adjuvant radiation and chemotherapy have been reserved for patients who have a mesenchymal chondrosarcoma or a dedifferentiated chondrosarcoma or for those who have had inadequate operative treatment. Unfortunately, to date, studies have not shown adjuvant treatments such as chemotherapy or radiation to have any significant impact on patient morbidity or mortality in the majority of isolated primary lesions. Proton beam radiation is generally reserved for refractory tumors in high risk anatomic areas such as the skull base and axial skeleton (33).

There is no indication for the use of chemotherapy or radiation in the management of low grade chondrosarcoma as adequate initial surgical resection can virtually be successful in all patients.

Clinical Trials

Earlier this decade, a national clinical trial was opened in the United States. However, due to the National Cancer Institute’s budgetary constraints and the relative rarity of chondrosarcoma compared to other cancers, the trial had to be closed prematurely. Investigators hope that the trial can be reinitiated soon.

Are there any emerging therapies in the treatment of chondrosarcoma?

During the past several years, substantial new insights have been gained about molecular cell biology, molecular cytogenetics, and immunopathology (34). These have led to a better understanding of chondrosarcoma development at the molecular level and will ultimately lead to the development of targeted treatments. Though they are at present highly experimental, researchers are investigating several new treatments for chondrosarcoma. Examples include agents targeting estrogen receptors (35), new chemotherapeutic agents, such as ET-743 (36), and agents effecting cytogenetic pathways (13).

Where is the best place to go to receive appropriate treatment?

Patients with chondrosarcoma are best treated at major Sarcoma centers with specialized diagnostic and treatment facilities and the availability of Musculoskeletal Tumor Specialists or Orthopedic Oncologists. Because this, like many other bone cancers, are not common, it is often a
What are the chances for cure and survival from low grade chondrosarcoma?

In general, the prognosis for chondrosarcoma depends on the grade of the tumor and the attainment of complete excision of the tumor and other conditions the patient has such as diabetes, lupus, and clotting and coagulation problems. Table 2 below shows a comparison between the prognosis based on the tumor grade. Pathological fracture, metastasis, local recurrence, and death are usually more common in patients with a high grade chondrosarcoma.

For low grade chondrosarcomas, prognosis is excellent after adequate excision, with very low rates of recurrence or spread when treated at an established sarcoma center. In a review of 70 patients with low grade chondrosarcoma of the appendicular skeleton only three presented with metastasis (14).

### Table 2: Prognosis by Tumor Grade

<table>
<thead>
<tr>
<th>Grade</th>
<th>Five-Year Survival</th>
<th>Metastatic Potential</th>
<th>Recurrence Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grade I</td>
<td>90%</td>
<td>0%</td>
<td>Low</td>
</tr>
<tr>
<td>Grade II</td>
<td>81%</td>
<td>10-15%</td>
<td>Fair</td>
</tr>
<tr>
<td>Grade III</td>
<td>29%</td>
<td>&gt;50%</td>
<td>High</td>
</tr>
<tr>
<td>Dedifferentiated</td>
<td>&lt;10% (1 year)</td>
<td>Most</td>
<td>High</td>
</tr>
</tbody>
</table>

Summary

Cartilaginous lesions of the human skeleton exist on a continuum, spanning from the completely benign embryonic inclusion, to the far less common but dangerously aggressive neoplastic process. Differentiating a benign enchondroma lesion from a low grade (grade I) chondrosarcoma is a challenging task even for the more experienced team of Sarcoma specialists. Based upon imaging characteristics, for less aggressive lesions, a wait and watch approach is the best initial management. At follow-up, if there appears to be progression, intervention should be considered.

The majority of patients with less radiographically aggressive low grade chondrosarcoma may be safely treated with a limited surgical interventional (intralesional resection). Patients with any signs of a radiographically aggressive lesion should be treated with a staged biopsy and a more extensive wide resection surgery to keep the local recurrence rate and potential for metastases low. In order to determine the appropriate treatment for each individual lesion, musculoskeletal...
oncologists must take into account the clinical, radiographic, histologic and soon the molecular biologic characteristics of the tumor.

It is important for patients to seek treatment for these tumors at a recognized sarcoma center with availability of specialists possessing a sound understanding of these lesions and a firm grasp of the evolving treatment options. The health care team at these centers will keep patients informed about the details of the treatment course in both the short and long term. Understanding and recognizing the spectrum of appearances of the various types of chondrosarcoma allow improved patient assessment and are vital for optimal clinical management including diagnosis, biopsy, staging, treatment and prognosis.

References


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