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Review

Chondrosarcoma Pelvis; A Review Article

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Abstract

The pelvis is three dimensional structures and is the most common site of Chondrosarcoma. Chondrosarcoma is the 2nd most common malignant bone tumor. Peak age of presentation is 50 to 60 year of age but it can appear in childhood as well. Chondrosarcoma can be primary and secondary and graded as I, II, III depending on the cellular atypia and aggressiveness. Pelvic Chondrosarcoma presents with large bumps because of no clear compartmental boundary to restrict its growth. Chondrosarcoma can present with pain, urinary obstruction, constipation and sciatic nerve palsy. Radiograph shows the punctuate calcified lesion with cauliflower like lesion, computerized tomography delineate the extent of pelvic bone and magnetic resonance imaging shows the thickness of cartilage cap more than 2.5 cm likely to be indicative of the Chondrosarcoma . Treating the pelvic Chondrosarcoma is the real challenge for the surgeon. Wide margin resection is the best treatment option followed by reconstruction with Allograft, Frozen bone, prosthesis, Arthodesis, internal hemipelvectomy and external hemipelvectomy. All methods of reconstruction have complications. Prognosis of the Chondrosarcoma depends on the grade and staging of the tumor.

Conclusion: Chondrosarcoma pelvis is the challenging problem for surgeon. Resection followed by reconstruction must be selective. Patient must be counseled about the complications.

Introduction

The pelvis is three dimensional structures and is the most common site of Chondrosarcoma. Chondrosarcoma are malignant bone tumor with pure type hyaline cartilage differentiation; myxoid changes, calcification, or ossification is present [1].Chondrosarcoma is the 2nd most common malignant bone tumor and if we add myeloma in list it becomes the third most [2].The pelvis is the most common site of Chondrosarcoma [3]. Peak age of presentation is 50 to 60 year of age but it can appear in childhood as well with slight male predominance [4].Chondrosarcoma accounts for approximately 20% to 25 % of bone sarcoma [5].

Classification

Chondrosarcoma are classified as central and peripheral according to site of involvement. Chondrosarcoma can be primary and secondary [6]. Primary Chondrosarcoma occurs in normal bone and secondary appears in enchondroams, osteochondroma, hereditary multiple exostosis (olliers disease) [7, 8]. Maffucci syndrome [9, 10] and fibrous dysplasia [11]. Chondrosarcoma can be well differentiated and undifferentiated [12]. Pathologically types of Chondrosarcoma are clear cell, mesenchymal, myxoid and dedifferentiated [13].

Pathologically Lichtenstein and Jaffe describe the pathological criteria for Chondrosarcoma that includes cells with pulp nuclei, more tha occasional cell with two such nuclei, giant cartilage cells with large single or multiple nuclei or with clumped chromatin [14].

Grades

Grading of CS depends upon the number of cell alterations plus the presence and absence of calcification and enchondral ossification as described by O'Neil& Ackerman. Chondrosarcoma is graded as Grade I that includes pauciarticular hyaline cartilage, small densely staining nuclei, rare or no binucleate cells with absent mitotic figures. Grade2 shows the increase peripheral cellularity, moderate size nuclei with nucleate atypia, bizarre nuclei and low mitotic figures. Grade3 shows more mitosis with dense peripheral cellularity. De

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differentiated CS are considered as high grade non cartilaginous tumors with bad prognosis and more likely to develop metastasis [15, 21]. Mesenchymal CS is rare high grade tumor with typical biphasic pattern consisting of both small cells and islands of atypical cartilage [16]. Molecular Genetic play role in differentiating peripheral and central CS .Mutation in EXT1 and EXT2 in multiple osteochondroma turn into malignant peripheral CS variant. While in central CS Ext gene is not involved. [17, 18]

Clinical Presentation

Clinically patients with Chondrosarcoma pelvis presents with Huge Lumps as there is no limited boundary or compartment to limit the spread of the tumor [19]. CS also can present with pain with mild to severe depends upon size and involvement of surrounding structures, urine retention, constipation, foot drop secondary to sciatic nerve palsy and L5 nerve root involvement, fixed flexion deformity hip (secondary contracture) [20].

Radiology

Radiologically, Chondrosarcoma present with typical "ring and arc"chondroid matrix mineralization or scattered calcifications in the cartilaginous part of the tumor [22, 23] Radiologically. Chondrosarcoma can present with the aggressive Osteolytic lesion with areas of mineralization. MRI may show the fluid levels on T2 weighted images [24]

Treatment

CS is the surgical disease as these tumor are chemotherapy and radiotherapy resistant because of their extracellular matrix, low percentage of dividing cells and poor vascularity [25]. Radiotherapy is only considered in patient with residual disease and inaccessible sites [26]. Radiotherapy may have some role in Mesenchymal Chondrosarcoma. CS is chemo resistant may be because of expression of multidrug resistance P-glycoprotein expression [27].

Surgical treatment

Pelvic CS treatment is challenging problem for the surgeon. Surgery is the only method of curing the CS [28].Wide margin resection is the best option of treatment [29, 30] Resection is all depend upon the involvement of Pelvis. Pelvis resection is divided into four levels. Levels of resection depend upon the involvement of level [31]. Level I can be resected with or without any reconstruction and level II can be resected without reconstruction. But the key is the CS In level II where acetabulum get involved and resection is challenging in functional point of view. Level IV is the sacral resection. Two surgical options are available for the treatment of CS pelvis on is Limb sparing and other is the amputation. Limb sparing can be internal hemipelvectomy, iliac resection, acetabular resection and pubis and ischium resection. Round the way External hemipelvectomy is the amputation. Alternate method is the hind quarter amputation [38]. Following the wide margin resection of the CS pelvis reconstruction around the pelvis is another challenge because of complications associated with the methods of reconstruction. Options available for the pelvic reconstruction are hemi pelvic allograft, autoclaved graft [32, 33] autograft for sacroiliac Arthodesis [34] iliosacral, iliofemoral, ischiofemoral Arthodesis [35]. Charnley Mueller method of reconstruction after resection of tumor around the acetabulum by implanting the total hip replacement and pelvic ring reconstruction with cement reinforced by Kuntscher rods and heavy K wires [36,41] Intraregional resection of pelvic Chondrosarcoma grade 1 can be done but with the risk of local recurrence of 100% that's why it should be avoided [37, 39].Pelvic reconstruction after periacetabular resection also can be done with saddle prosthesis with advantage of shorter operative time, rapid recovery with moderate postoperative function but associated with high risk of complications including dislocation, infection, extensive upward migration of prosthesis and prosthesis dissociation [42].Pelvic allograft are associated with high complication rates mostly infections [43]. Recently Computer assisted navigation aid in accurate resection of pelvic tumor and with addition of new technique using fluro CT matching may allow more accurate resection of the margins [40]. Prognosis of low grade conventional Chondrosarcoma is up to 87% and for the pelvic and sacral ranging from 25 to 54% [44, 45].

Conclusion

In conclusion pelvic Chondrosarcoma is surgical problem and real challenge for the surgeon because of large lumps and important structures lying nearby. Another challenge is method of reconstruction of pelvis those have high complication rate so reconstruction should be done in selected patient.

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