International Surgery Journal
Khan MM et al. Int Surg J. 2017 Oct;4(10):3567-3569
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: http://dx.doi.org/10.18203/2349-2902.isj20174542

Case Report

Huge low-grade chondrosarcoma arising from rib cage: a case report

Manal M. Khan[1], Yasmee Khan[2], Rohit Kumar Namdev[1]*

[1]Department of Burns and Plastic Surgery, AIIMS, Bhopal, Madhya Pradesh, India
[2]Department of Medicine, AIIMS, Bhopal, Madhya Pradesh, India

Received: 14 August 2017
Accepted: 07 September 2017

***Correspondence:**
Dr. Rohit Kumar Namdev,
E-mail: dr.rknamdev@gmail.com

ABSTRACT

We report a case of 35 years old male who presented with a huge, gradually progressing swelling of right side of chest and abdomen for 5 years. Dimensions of swelling were 40cmx30cm. Radiological findings suggested characteristic appearance of a chondrosarcoma which was arising from right lower ribs. Lesion has involved whole liver, gall bladder, right half of colon and right kidney. On histopathology, we found low grade chondrosarcoma. Though low-grade chondrosarcoma is a potentially curable neoplasm in its early stages, the present patient reported very late. As famous dictum said "the ignorant are ignorant of their ignorance ", this is a classic example of patient's ignorance which converted a curable disease into incurable.

Keywords: Giant chondrosarcoma, Rib cage tumour

INTRODUCTION

Chondrosarcoma is a type of sarcoma, which is composed of transformed cartilage producing cells. It is the most common malignant tumor of cartilage and second most common primary spindle cell tumor of bone. Classically there is no evidence of bone formation in chondrosarcoma and if found they are classified as osteosarcoma. Osteosarcoma can be intramedullary, peripheral, mesenchymal, differentiated and clear cell. Clinically they can vary for low grade well differentiated locally invasive tumor to high grade metastatic tumors. Chondrosarcoma usually occurs in pt more than 40 years of age. The most common site is long bones and pelvis. They are most common primary malignant tumor of sternum and scapula. They are usually painless and pain arises when they involve nerves or compresses surrounding structures. Pain is considered an ominous sign in case of benign cartilaginous tumor. Histologically they are classified into grade I, II, III. Metastatic potential tends to correlate with histological grades. Survival rates also decrease as grade of tumor increases. Classically chondrosarcoma is considered as radio and chemo resistant tumor. However, some studies show that some of these lesions are radio curable specially when that involve axial Skelton or when the patient is medically unfit for surgery.[1,2]

CASE REPORT

A 35 years old man came to us with a huge swelling over his right side of chest and abdomen. Swelling was associated with recent onset mild pain. He has a history of small swelling which first appeared over his right lower rib cage. Because this swelling was initially painless he ignored it and didn't take any medical advice. Dimension of swelling is 40X30 cm. It was lobulated, hard in consistency and fixed to underlying rib cage (Figure 1, 2).

Radiology- a contrast enhanced spiral CT of chest abdomen and pelvis was performed. STIR coronal sequence of the MRI is also done to evaluate the matrix of the lesion. We found a huge neoplasm of size 36X31x30 cm arising from lateral and anterior ends of the right lower ribs. Neoplasm was involving chest and

abdominal wall with intra-abdominal extension. Intra abdominally lesion was encasing entire liver right half of colon and right kidney. Neoplasm has a characteristic ring and arch appearance and show typical chondroid matrix. Chondroid was hyper intense on the STIR weighted image of the MRI. No parenchyma or mediastinal abnormality found.

Figure 1: Clinical images.

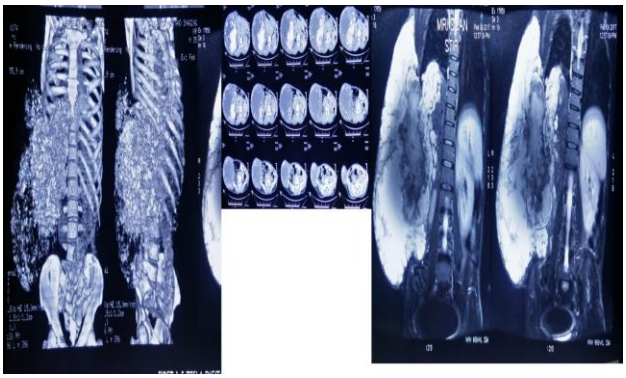

Figure 2: CECT abdomen and STIR MRI.

DISCUSSION

Chondrosarcoma (CHS) is a rare malignancy. The estimated overall incidence of CHS is 1 in 200,000 per year.[3] majority of CHS occurs in older individuals(>50years) with a slight male predilection (M:F=1.5:1). Chondrosarcoma may arise de novo or may arise from preexisting benign tumor of cartilage. Swelling is the most common presenting feature followed by local pain. Patient may present with pathological fractures. Histologically CHS ranges from grade I to grade III. Grading is done on the basis of nuclear size, cellularity, hyperchromasia and mitoses. As the grade of CHS increases cellularity increases.[4] Immunohistochemically, the cartilaginous area is strongly positive for S-100. CHS is consistently positive for CD99 and may stain for vimentin and Leu. CHS is negative for osteocalcin, actin, cytokeratin, and epithelial membrane antigen (EMA).[5,6] Chondrosarcoma is common in patients with Maffucci syndrome and Ollier's disease. Both syndromes are characterized by multiple enchondromas with frequent malignant transformations. Chondrosarcoma of the chest wall is the most frequent primary malignant chest wall tumor. Surgery remains the only effective treatment. Widhe B et all studied 106 patients of chest wall CHS over a period of 22 years. Ninety-seven patients were treated with a curative intent. Patients operated with wide surgical margins had a 10-years survival of 92% compared with 47% for those with intralesional resections. The 10-years survival was 75% for patients treated at sarcoma centers and 59% for those treated by thoracic or general surgeons. Local recurrence rate was highly dependent of the surgical margins-4% after wide resections and 73% after intralesional resections. The proportion of intralesional resections was higher outside sarcoma centers. Prognostic factors (multivariate analysis) for local recurrence included surgical margin and histological grade; for metastases, prognostic factors included histological grade, tumor size, and local recurrence. Metastases occurred in 21 of the patients and only 2 were cured.[7] Radiotherapy has a limited role in CHS. Proton radiotherapy has some proven role in the case of partially resected CHS of the skull base and axial skeleton. Hug EB et all reported local control rates of 85% to 100% in cases of CHS of skull base. There are several structural barriers of chemotherapy like poor vascularity, low perfusion index and abundant extracellular matrix. Further it is also hypothesized that CHS cell may harbor multidrug-resistance 1 gene (P-glycoprotein), which confers resistance to doxorubicin.[9] However some chemo response has been noted in dedifferentiated and mesenchymal subtypes of chondrosarcoma.[10] Lastly patient awareness is utmost important. Present patient came to us when the tumor invaded liver and other intra-abdominal structure. This is a classic example of ignorance which converted a potentially respectable and curable tumor to nonresectable and lethal. At this stage only palliative measures can be taken.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Harwood AR, Krajbich JI, Fornasier VL. Radiotherapy of chondrosarcoma of bone. Cancer. 1980;45(11):2769-77.
2. Treating Specific Bone Cancers. Available at https://www.cancer.org/cancer/bone-cancer/treating/treating-specific-bone-cancers.html
3. Fletcher CD. Pathology and genetics of tumors of soft tissue and bone. World Health Organization Classification Tumors. 2002;4:35-46.
4. Evans HL, Ayala AG, Romsdahl MM. Prognostic factors in chondrosarcoma of bone. A clinicopathologic analysis with emphasis on histologic grading. Cancer. 1977;40(2):818-31.
5. Devoe K, Weidner N. Immunohistochemistry of small round-cell tumors. In Seminars in diagnostic Pathology. 2000;17(3):216-224.

6. Fanburg-Smith JC, Auerbach A, Marwaha JS, Wang Z, Rushing EJ. Reappraisal of mesenchymal chondrosarcoma: novel morphologic observations of the hyaline cartilage and endochondral ossification and β-catenin, Sox9, and osteocalcin immunostaining of 22 cases. Human Pathol. 2010;41(5):653-62.
7. Widhe B, Bauer HC, Scandinavian Sarcoma Group Surgical treatment is decisive for outcome in chondrosarcoma of the chest wall: a population-based Scandinavian Sarcoma Group study of 106 patients. J Thorac Cardiovasc Surg. 2009;137(3):610-4.
8. Hug EB, Loredo LN, Slater JD, DeVries A, Grove RI, Schaefer RA, et al. Proton radiation therapy for chordomas and chondrosarcomas of the skull base. J Neurosurg. 1999;91(3):4329.
9. Cakir O, Topal U, Bayram AS, Tolunay S. Sarcomas: rare primary malignant tumors of the thorax. Diagn Interv Radiol. 2005;11(1):23-7.
10. Huvos AG, Rosen G, Dabska M, Marcove RC. Mesenchymal chondrosarcoma. A clinicopathologic analysis of 35 patients with emphasis on treatment. Cancer. 1983;51(7):1230-7.

Cite this article as: Khan MM, Khan Y, Namdev RK. Huge low-grade chondrosarcoma arising from rib cage: a case report. Int Surg J 2017;4:3567-9.