CASE STUDY

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Huge Chondrosarcoma of Shoulder

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Background: Chondrosarcoma is the second most frequent primary malignant tumor of bone. It is considered relatively an uncommon tumor. Generally it is not common for chondrosarcoma to attend huge size; however, one type was described as mesenchymal chondrosarcoma of low grade cartilaginous cells, with female predominance which can grow to exceptionally large size.
Case Presentation: We report a case of Chondrosarcoma, in 30 years old woman of 4 years duration, with a huge mass (weighted 16 Kg) and roughly measured 40x30 x20cm, around the right shoulder
Investigations: Repeated MRI of the shoulder and trunk revealed a clear plane of cleavage between the tumor mass and the chest and abdominal walls and absence of invasion of the abdomen. Ultra sonography revealed clear abdominal cavity and CT of the chest revealed clear lungs of any metastasis.
Treatment: A right forequarter amputation accomplished successfully and the whole tumor with the right upper extremity in one mass was removed.
Outcome and Follow-up: The post operative period was uneventful. The pathological examination is consistent with well differentiated chondrosarcoma.
Conclusions: It was difficult to decide whether the tumor was a primary or secondary; because of poor clinical history and her reluctance to seek medical advice, she is living in a small town 50 km south of Baghdad and there is good medical services in her area; this could reflects the social and psychological attitude towards amputation of a limb as a treatment.

Introduction:

Chondrosarcoma is the second most frequent primary malignant tumor of bone [1-4], with an incidence about half of osteosarcoma [5]. It constituted 7.6% of the primary malignant bone tumors in the Registry of Bone Sarcoma of the American College of Surgeons [6]. It is considered relatively an uncommon tumor [7].

The peak age incidence is in adulthood and old age between 20 and 70 years. More than half of tumors localized anatomically in pelvic girdle and ribs; 84% found in upper Femur and upper humerus with low incidence in the peripheral portions of extremities [7].

There is clear male predilection than female; in a study of 212 cases 138 were males and 74 were females [7]. Because of subtle histological changes necessary for predicting malignant clinical course, it requires a great correlation between clinical picture and histological picture to decide about labeling malignant chondroid lesion [6]; keeping the preserved specimens for many years for review new microscopic section to correlate and establish a pattern of natural history of the disease, clinical pictures of recurrence or local extension is sufficient evidence of malignancy [7].

Secondary chondrosarcoma arise at the site of preexisting benign lesion, in up to 25%, most frequently in cases of multiple enchondroma and multiple hereditary exostoses [5].

In secondary chondrosarcoma the presentation as a hard swelling or lytic lesion on plane radiological examination, pain rarely is a presenting symptom, usually with pathological fracture, and rapid increase in size [5].

In 11 out of 212 cases metastasis occurred within 2 years of 1st surgical procedure, however, metastasis can still occur up to 10 years after definitive surgery. Radiation did not seem to alter the final course of the disease, it may relief pain or regress the size [7].

Generally it is not common for chondrosarcoma to attend huge size; however, one type was described as mesenchymal chondrosarcoma of low grade cartilaginous cells, with female predominance and can grow to exceptionally large size [8].

Case Presentation:

M. A. Z., a 30 years old mother of 3 children presented to Al Wasity hospital in July 2009 with a huge mass around the right shoulder of 4 years duration, the swelling was generally irregularly oval in shape, roughly measures

40x30 x20cm involving the scapular region, chest wall and abdominal wall, swelling was painful with lobulated surface, hard in consistency with central deep ulcer with moderate amount of bleeding and infection.

Function of the right shoulder was completely lost, while the functions of the elbow and of the hand were preserved.

Investigations:

General serological and biochemical investigation were within normal limits apart from moderate chronic hypochromic anaemia with Hb of 9.0 g/dl, ESR was 60 ml /hr.

Previous attempt of doing a biopsy was not conclusive and not documented because it was done more than 3 years ago, and the family had lost the report.

The clinical picture, with progressive growth of the tumor to a huge size and the destruction of bone on plane radiological examination (Figure1) pointed to the malignant behavior of the tumor.

Figure 1: a plain radiograph of right shoulder showing the nearly complete destruction of the joint and scaoula with surrounding clouds of calcifications.

Repeated MRI of the shoulder and trunk revealed a clear plane of cleavage between the tumor mass and the chest and abdominal walls and absence of invasion of the abdomen. Ultra sonography revealed clear abdominal cavity and CT of the chest (Figure2) revealed clear lungs of any metastasis.

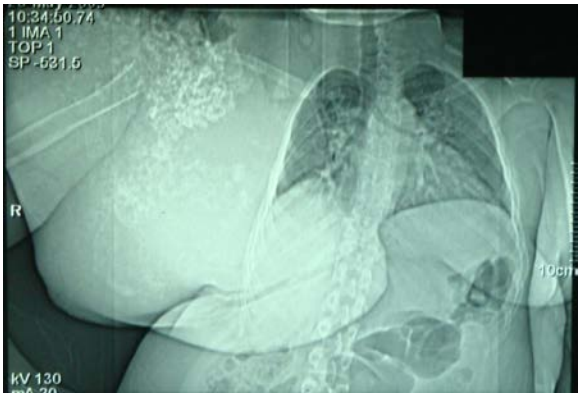
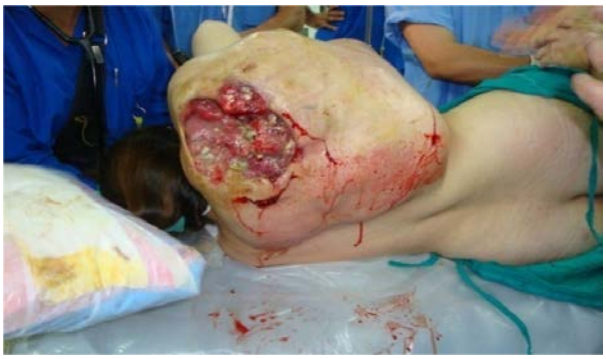


Figure 2: computerized radiography showing the extent of tumor pushing the chest and abdomen.

Treatment:

Decision was made to remove the tumor with a good safe margin (Figure 3) to improve the quality of life and give the patient a great hope of cure.



A

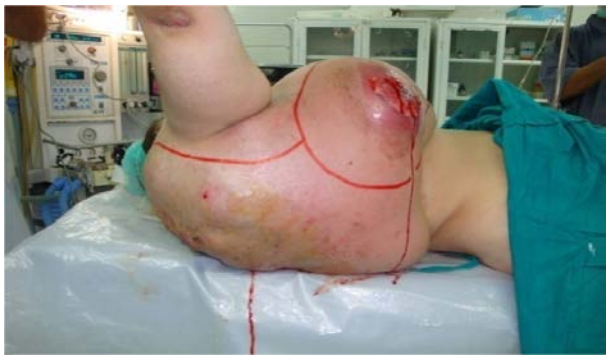


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Figure 3: A and B; posterior and lateral views with drawing the lines of incision

Supportive measures to build the energy and defense of the body to withstand the trauma of the coming operation were undertaken by giving tonics and correction of anemia. Chest surgeon and a plastic surgeon were consulted to participate with the orthopedic surgeons to accomplish a safe procedure .The anaesthesiologist prepared an intensive care bed.

A right forequarter amputation accomplished successfully and the whole tumor with the right upper extremity in one mass was removed, the weight of the whole tumor was 16 kg (Figure 4 and 5).

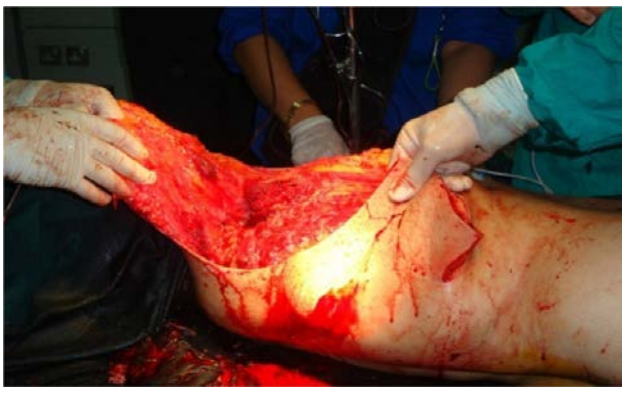


Figure 4: after the forequarter amputation and removal of the tumor.

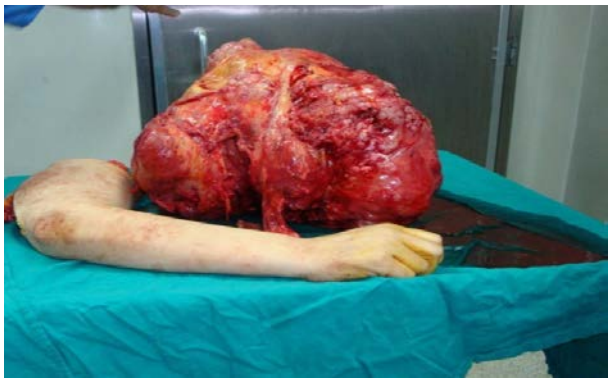
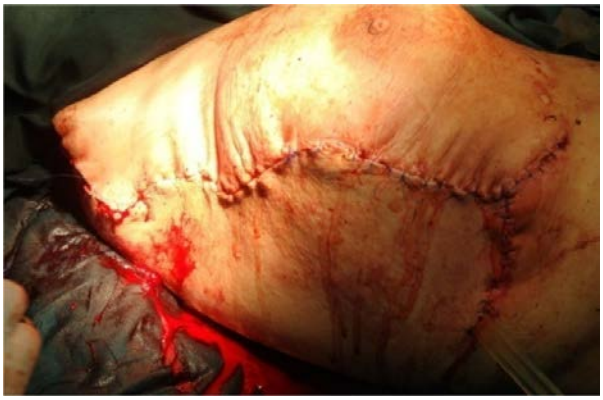


Figure 5: the excised tumor and upper limb.

Fashioning of the skin were enough to close the wound primarily over a non vacuum drain (Figure 6). An enlarged lymph node removed from the supraclavicular region as well.



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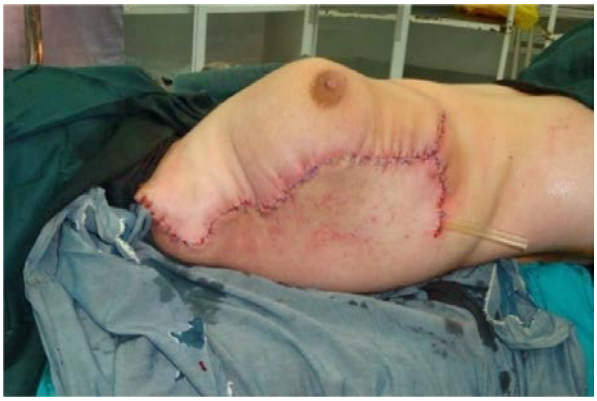


B

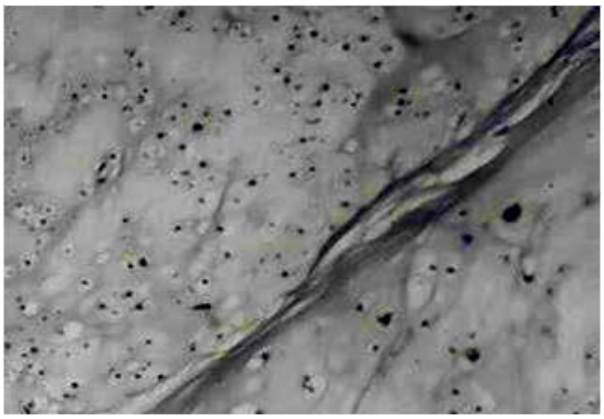
Figure 6: A and B; the wound is closed over drains.

Outcome and Follow-up:

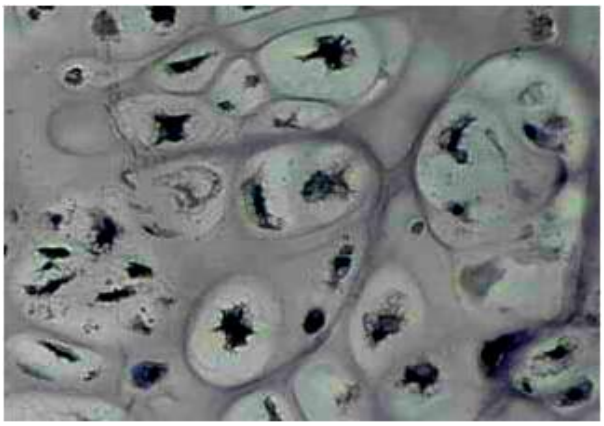
The post operative period was uneventful, drain removed on the third post operative day and the stitches of wound removed on the 15th day, no infection occurred but seroma continued to discharge for further three weeks.

The pathologist examined the whole specimen and chose many pieces from appropriate sites for histological examinations, the sections (Figure 7) showed lobules of cartilage separated by fibrovascular septae, chondrocytes show signs of atypea as hyperchromasia, more than one cell in each lacunae, more than one nucleus in some of the cells and abnormal mitosis, the picture is consistent with well differentiated chondrosarcoma. Sections of the lymph node show features of sinus hyperplasia, no secondary metastasis seen.

Tumor staging= $T_3N_0M_x$ (stage III)
Tumor grade = grade one
Tumor markers: (S 100 Protein) strongly positive.



A



B

Figure 7: A and B; light microscopic sections showing the cartilaginous nature and cellular atypea.

Discussion:

Chondrosarcoma is an uncommon tumor, yet it is considered the second most frequent primary malignant tumor of bone [1-4,7], it occurs mostly near the center of the body in pelvic girdle and ribs, it has male predilection [7].

In our case the exciting feature is the huge size of the tumor which weighted 16 KG, searching the literature, we couldn't find such big size, however, Giudici mentioned mesenchymal type of chondrosarcoma that can grow exceptionally large [8], Giudici found this type in low grade tumors and in females, consistent with our case, a 30 years old female and had a grade I tumor.

A second point in our case is the long history of four years duration with massive destruction of the Scapula and extension to chest and abdominal walls, there is no metastasis to nearby lymph nodes neither to the lungs by blood stream, it is mentioned in the literature that spread can be delayed up to ten years [7], low grade tumors are known to metastasize slowly, our case had grad I tumor.

It was difficult to decide whether the tumor was a primary or secondary; because of poor clinical history and her reluctance to seek medical advice, she is living in a small town 50 km south of Baghdad and there is good medical services in her area; this reflects the social and psychological attitude towards amputation of a limb as a treatment.

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