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Well- Differentiated Osteosarcoma Of The Rib

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Abstract:

Primary osteosarcoma are one of the most common malignant bone tumors principally affecting the long bones in children and adolescent. Well differentiated osteosarcoma of the rib is extremely rare, there is no sex.

Predominance and the mean age is in the third decade. The tumor has strong predilection for the long bones of the extremities, especially the metaphysis but may occur in the diaphysis. Radiological, the lesion shows no distinctive features, often simulating fibrous dysplasia or desmoplastic fibroma.

I report a case of well differentiated osteosarcoma involving the (7^{th},8^{th},9^{th},) left ribs of 25-year-old male. This is a peculiar case of well differentiated osteosarcoma involving an unusual site.

This patient underwent a wide excision of the tumor of the ribs and the pleura and chest wall reconstruction. The patient followed by courses of chemotherapy, and follow up for 22-monthes with no sign of recurrence. Classic microscopic feature of a ramifying osteoid matrix amidst the tumor cells was diagnostic of an osteosarcoma.

Key words: male, chest wall tumors, osteosarcoma, ribs.

الخلاصة:

ورم العظم الخبيث (السرقوم) ورم خبيث ينشا في النسيج الضام .هو واحد من أورام العظام الخبيثة الأكثر شيوعا التي تؤثر أساسا في العظام الطويلة عند الأطفال والمراهقين. وورم سرطان الضلع يعتبر من الحالات النادرة الحدوث. ولا توجد دراسة حول نسبة الاصابة بين الذكور والاناث.

غالبية الاصابة هم من متوسط العمر هو في العقد الثالث. الورم لديه ميل قوي للعظام الطويلة من الأطراف وخاصة الكردوس ولكن قد تحدث في مناطق نمو العظام، الرقوق الشعاعية لم تظهر أي ملامح مميزة، تشبه في كثير من الأحيان خلل النسج الليفي أو ورام ليفية صلدة.

هنا حالة من سرطان العظم للأضلاع (**7، 8، 9**) من الصدر الايسر لذكر 25 عاما، هذه حالة غريبة من عظمية مختلفة تماما تنطوي على موقع غير عادي.

خضع هذا المريض لعملية استئصال الاضلاع مع غشاء الجنب واعادة تصليح جدار الصدر وتتبع بعدها بأخذ جرعات من المواد الكيمياوية المعالجة للسرطان. تمت متابعة حالته لمدة 22شهر وقد شفي تماماً، وكانت السمة الكلاسيكية مجهرية من مصفوفة عظماني متشعبة وسط الخلايا السرطانية التشخيص لعظمية .

Introduction:

The risk of being diagnosed with cancer increases as the individual ages, and 77% of all cancer are diagnosed in persons age 55-year and above. As a lifetime risk, the probability that an individual, over the course of a lifetime. Will develop a cancer is slightly less than one in two for men and a little more than one in three for women[1-3].

In the united states, for children aged 0-14 , and adolescents aged 15-19 years, the overall incidence rate for all cancers is (16.5)cases per 100.000 persons per year(1-4). The childhood and adolescent cancer incidence rate has increased from (11.5) per 100.000 person per year in 1975, to (14.8) per 100.000 in 2004.

Although this trend is recognized to be the result, in part, of improved diagnosis and reporting methods, it appears that there is a true increase in the occurrence of the some childhood cancer[2-5].

For any newborn, the risk of developing cancer by age 20-years is about one in 300 for males and one in 333 for females[1-6].

It has a peak of incidence at age (5-14).[7] . Childhood cancers account for no

more than 2% of all cancers[2]. Well differentiated osteosarcoma(OS) of the bone is rare , accounting for approximately 1.2% of all (OS)[8], and was first described by Unni et al. in 1977[9]. Presenting symptoms and signs are non-specific and typically consist of pain and swelling. Often symptoms have been present for more than one year, a rare occurrence for conventional, high grade (OS0)[10,11].

There is no obvious sex predilection, although one study suggested a female predominance[10]. Patients are distinctly older, on average, than those with conventional (OS). It often peaks out in the third decade of life, but affected individuals are often in their fourth, fifth, sixth, or even seventh decades[11].

They principally arise in the metaphysis of the long bones, particularly at the lower end of the femur and upper ends of the tibia and humorous[12]. Short bones, spine, and flat bones, such as, the ribs, pelvis, and craniofacial bones are less frequently involved. Secondary (OS) may develop in unusual bones, such as, vertebra and flat bones in patients treated with chemotherapy[12].

Primary (OS) of the rib, occurring in an adult, is extremely rare. I present one such case that posed a diagnostic difficulty, owing to its unusual location.

Materials and Methods:

Case History:

A 25-year-old man presented with a progressively increasing left -sided chest wall swelling for three weeks.

The swelling was accompanied by pain for the past three weeks. However, there was no fever, cough, breathlessness or loss of weight.

There was a history of emergency laparotomy operation due to perforated peptic ulcer.one month before the presentation of the chest wall pain. Physical examination revealed a bony hard mass in the left hemithorax measuring 8X6 cm extending from the anterior costal cartilage to the mid axillary line.

An X-ray examination revealed opacity in the left lateral chest wall involving the ribs and soft tissue. [Figure 1] A computed tomography (CT) scan showed a lobulated, extra pleural, soft tissue density, with calcification involving the seventh, eighth, and ninth ribs, which showed sclerosis and destruction [Figure 2]. The pleura were involved, but the lungs appeared normal.

A preoperative diagnosis of chondro- sarcoma was considered. A left anterolateral thoracotomy with tumor resection including the (7th, 8th, 9th.) ribs from the costal edge till the posterior costal parts with the affected pleura and chest wall reconstruction with prolene mesh was performed. The tumor was excised in Toto keeping the field clean from obvious massGross examination revealed a partly encapsulated bony hard mass measuring 10X8X6 cm, bearing the mass resected. The tumor was seen involving and encasing all the ribs [Figure 3].

The cut surface of the tumor was solid, gray-white, and fleshy. Microscopic examination showed a malignant neoplasm composed of pleomorphic, round-to-spindle shaped cells the spindle cells showed slightly hyerchromatic spindle shaped nuclei and indistinct cytoplasmic borders [Figure 4].Often the spindle cells revealed in an interlacing pattern and infiltration with permeation of the bone marrow. There was abundant cartilage formation.

The tumor was composed of newly formed irregular bony trabecular and hypocelluar spindle cells in fibrous stroma.

A diagnosis of conventional well differentiated osteoblastic osteosarcoma of the ribs and the pleurae. Although the soft tissue surgical margins were involved, the bone surgical margins were free. The patient was referred to a specialized cancer center for further adjuvant therapy.

Discussion:

Primary osteosarcomas usually originate in the metaphysis of the long bones. Approximately 10% of osteosarcomas are located in the flat bones, with the pelvis being the main site, and a mere 1 - 2% occur in the thoracic bones inclusive of the ribs, sternum, and clavicle[12].

OS are known to have a predilection to affect long bones, such as, the distal femur and proximal tibia, because these are the sites of greatest bone growth, where bone cell mitotic activity is at its peak. In fact there is a high incidence of these tumors in large dog breeds such as Great Danes and St. Bernard's, for the same reason [13].

Cases originating from the ribs are infrequent and have been reported mainly in the pediatric population [14,15,16] In a large study comprising of 49 cases of primary malignant chest wall tumors there were no osteosarcomas reported[17]. Involvement of flat bones may be seen as a metastatic process or secondary to chemotherapy, but a primary OS is rare[18].

OS originating from such a rare site poses a diagnostic challenge to the radiologist, pathologist, and the surgeons.

The typical 'sunburst' radiological pattern observed in the OS of the long bones may not be evident in the OS of the flat bones. [17] Owing to the exclusive site and varying radiological images, these tumors can be confused with other bone lesions and the differentials may include chondrosarcoma, fibrosarcoma or meta-static tumor.

Although a CT scan and magnetic resonance imaging (MRI) can evaluate the exact location and extent of involvement of the bone and adjacent structures, they may not be useful in defining the exact nature of the tumor.

Some authors opine that osteosarcoma should be suspected if the CT scan reveals a dense calcification within a mass that is centered in a rib[19]. Histopathological diagnosis is imperative in instituting a definite therapy.

The classic feature of a ramifying osteoid matrix laid down by the neoplastic cells clinches the diagnosis and enables one to exclude all the other possible differentials. Diagnostic difficulties may be encountered when the osteoid production is scant, when a diligent search for the same and extensive sampling of the tumor is warranted. A combined effort with radiological and clinical correlation is trustworthy and may serve to avoid pitfalls in the diagnosis.

Conventional osteosarcomas are the most aggressive osseous neoplasms. The overall prognosis of osteosarcoma in flat bones remains poor because of the difficulty of complete excision.

The guidelines for management and the prognosis and survival rates in rib primary OS is not clear due to the small number of cases studied. However, there is documentation of better survival in patients who have had a complete resection of their tumor at the time of surgery. [15] A local wide excision with removal of the involved ribs and subsequent reconstruction using a mesh followed by adjuvant chemotherapy and radiotherapy may improve survival in these patients. [15] Although OS of flat bones is rarely associated with metastasis, it is prudent to include a CT scan of the thorax, chest x-rays, and bone scans as part of the management protocol, in order to look for metastasis. [15].

Postoperative multi - agent chemotherapy has improved these patients' survival, reducing the risk of both local and distant relapse. However, there are also reviews which report no apparent difference between the patients treated with surgery alone and those treated with both surgery and chemotherapy. Overall survival despite modern adjuvant chemotherapy is noted to be 27% at 5 years [20].

Osteosarcoma of the flat bones is rarely associated with metastasis in contrast to osteosarcoma at other sites. In

this patient, despite the large tumour mass with aggressive features on imaging and good response to chemotherapy, there was no evidence of metastases after 22-months of the following this patient.

In summary, this paper presents a rare case of primary osteosarcoma of the rib and emphasizes that this condition should be considered in the differential diagnoses of children and adolescents presenting with a chest wall tumour. As metastases at presentation is uncommon, early diagnosis and aggressive surgical management will help to improve outcome. CT remains the essential imaging modality for accurate primary evaluation of the tumour mass, identification of metastatic disease, evaluation of tumour response to chemotherapy and postoperative evaluation of recurrent neoplasm, all of which have important prognostic implications

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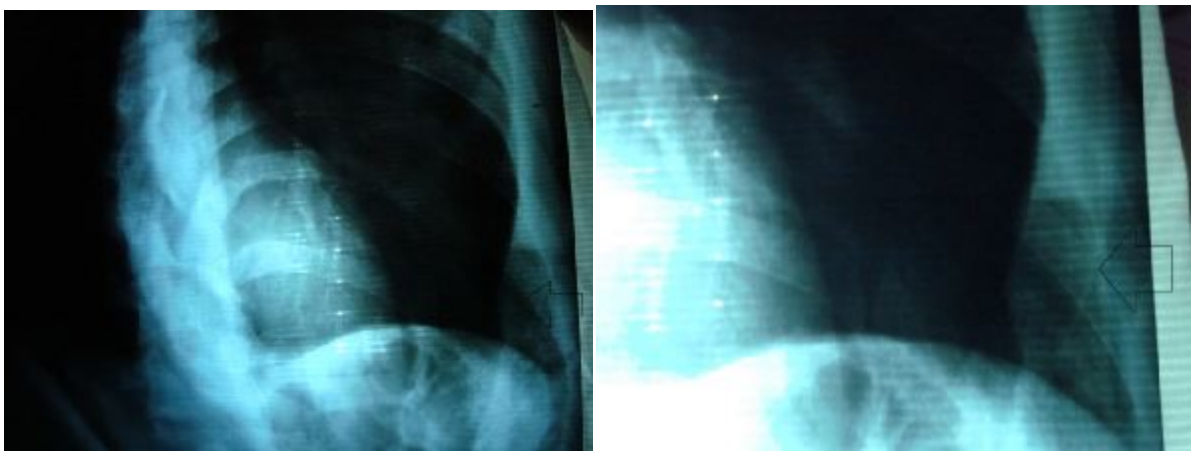

Figure-1: Plain chest X-Ray shows a mass in the left hemithorax.

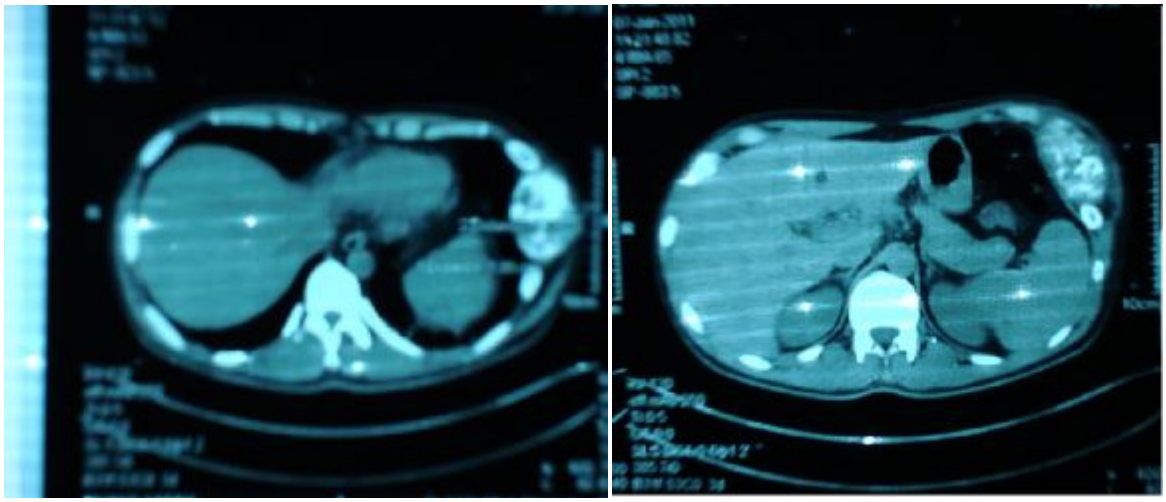

Figure-2: Axial CT image showing a lobulated, soft tissue density mass with calcificationand sclerosis, and is seen destroying the rib

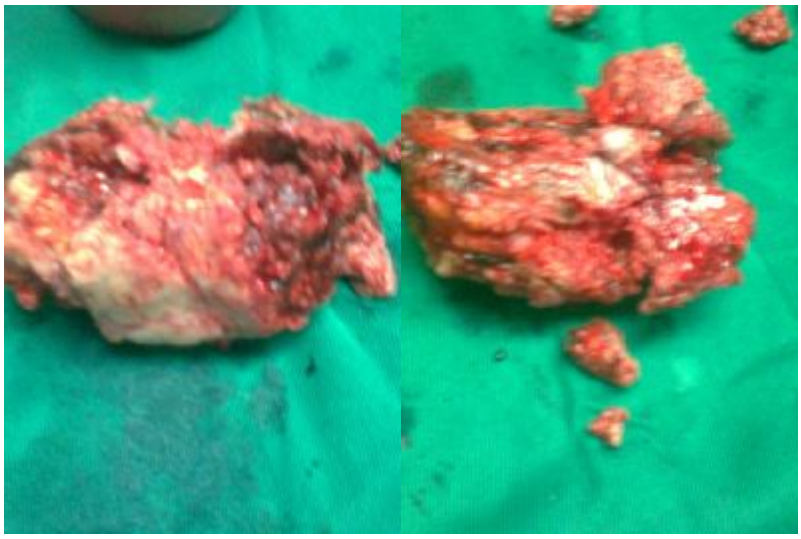

Figure-3: The tumour resected with attached ribs

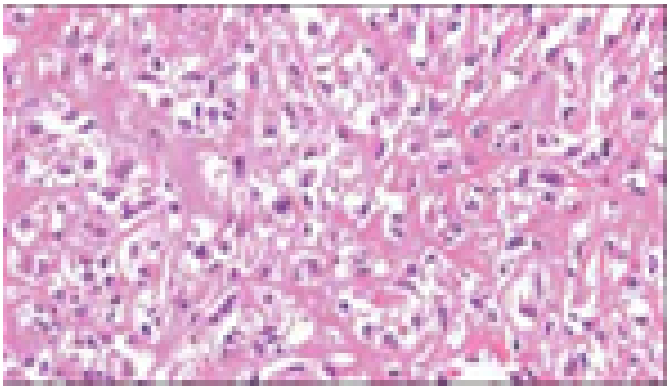

Figure-4 : Neoplastic cells rimming haphazardly laid osteoid (H and E, ×200]

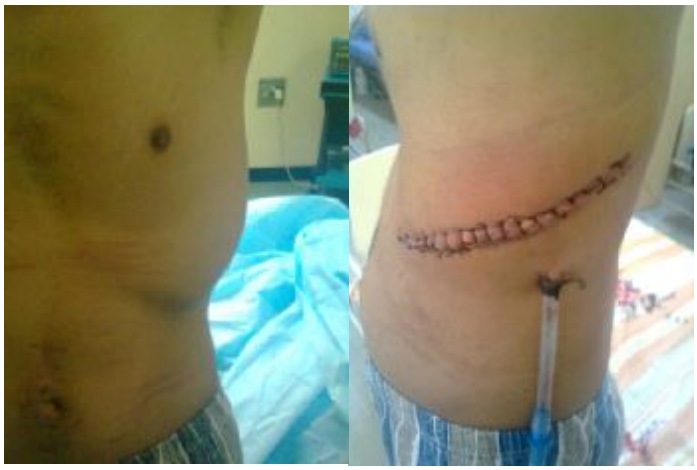

Figure-5: A, B: Showed the patient before the operation and after the operation

References:

1- Rics, L. A. G.; Mclbert, D. and Krapcho, M. et al. eds. SEER, R. cancert Statistics Review, 1975- 2004 National cancer Institute. January 2009.

2- American Cancer Society. Global cancer facts and figures. 2007 Atlanta GA; American Cancer Society January 2009.

3- S. Cancer Statistics Working Group. United States Cancer Statistics; Incidence and Mortality. Atlanta, GA; U.S. Department of Health and Human Services, Centers for Disease control and Prevention and National Cancer Institute: January 2009.

4- Lid, Thompson, T. D. and Miller, J. W. et al. Cancer incidence among children and adolescent in United States, 2001-2003. Pediatrics. 2008. Vol. 121. Pp: 1470-1477.

5- Steliarova Foucher, E.; Stiller, C. and Kaatsch, P. et al. Geographical patterns and time trends of cancer incidence and survival among children and adolescent in Europe since the 1970 (ACCIS Project):an epidemiological study. Lancet. 2004. Vol. 369. Pp: 2097-2105.

6- Minino, A. M. and Smith, B. L. Deaths: Preliminary data for 2000,Nal Vital Stat Rep. 2001. Vol.49. Pp:1-40.

7- Suravi, Mohanty, Y. K.; Inchara, Julian, A. Crasta, and Anuradha Ananthamurthy. An unusual case of primary osteosarcoma of the rib in an adult.Indian Journal of Medical and Paediatric Oncologg. 2010. Vol. 31 (97). Pp: 1-978.

8- Unni, K. K. Delinks bone tumors. General aspects and data on 11,087 cases. 5th ed. Philadelphia: Lippincott-Raven. 1996. Vol. 167.

9- Unni, K. K.; Delink, D. C.; McLeod, R. A. and Pritchard, D. J. Intra-osseous well differentiated osteo-sarcoma. Cancer 1977. Vol. 40. Pp: 1337-47.

10- Eilis, J. H.; Siegel, C. L.; Martel, W.; Weatherbee, L. and Dorfman, H. D. Radiologic features of well – differentiated osteosarcoma. Am J Roentgenol 1988. Vol.151. Pp:739-42

11- Kurt, A. M.; Unni, K. K.; McLeod, R. A. and Pritchard, D. J .Low-grade intraosseous osteosarcoma. Cancer 1990. Vol.65. Pp: 1418-28.

12- Mirra, J. M.; Gold, R. H.and Picci, P. Osseous tumors of intramedullary origin: In: Mirra JM, editor. Bone Tumors: Clinical, Radiological, and Pathological Correlations. Philadel-phia: Lea and Febiger; 1989. Pp:143-438.

13- Rosenberg, A. E. Bones, Joints, and Soft tissue Tumors. In: Robbins, Cotran, editors. Pathologic Basis of Disease. 7th ed. Philadelphia: Saunders; 2004. Pp: 1273-324 .

14- Yamaguchi, T.; Shimizu, K. Koguchi, Y.; Saotome, K. and Ueda, Y. Low-grade central osteosarcoma of the rib. Skeletal Radiol 2005.Vol. 34. Pp: 490-3.

15- Chattopadhyay, A.; Nagendhar, Y.and Kumar, V.Osteosarcoma of the rib. Indian J Pediatr 2004.Vol.71.Pp: 543-4.

16- Deitch, J.; Crawford, A. H. and Choudhury, S. Osteogenic sarcoma of the rib: A case presentation and literature review. Spine (Phila Pa 1976). 2003. Vol. 28. Pp: E74-7 .

17- Sabanathan, S.; Shah, R. and Mearns, A. J. Surgical treatment of primary malignant chest wall tumors. Eur J Cardiothorac Surg 1997.Vol. 11. Pp: 1011-6.

18- Kellie, S. J.; Pratt, C. B. ; Parham, D. M. ; Fleming, I.D.; Meyer, W. H. and Rao, B. N. Sarcomas (other than Ewing's) of flat bones in children and adolescents. Cancer 1990. Vol. 65. Pp: 1011-6.

19- Abdulrahman, R. E.; White, C. S.; Templeton, P. A.; Romney, B.; Moore, E. H. and Aisner, S. C. Primary osteosarcoma of the ribs: CT findings. Skeletal Radiol 1995. Vol. 24. Pp: 127-9.

20- Lee, H. C.; Hsieh, C. C.; Wu, Y. C.; Wang, L. S.; Huang, B. S.; Hsu, W. H. and Huang, M. Management of primary chest wall tumors: 14 years' clinical experience. J Chin Med Assoc 2006.