

Osteosarcoma – A Clandestine Enigma

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Bone tumors form a small part of all human cancers. As per the SEER data about 2570 new cases of bone sarcomas were diagnosed in the United States in the year 2005.

Osteosarcoma, earlier called as “Osteogenic Sarcoma” is the most common primary bone tumor in humans, which has a predilection for metaphysis of long bones in children, adolescents, and young adults and most commonly involves the bones around the knee joint in about 65% of cases. The earliest published literature takes us to 1879, where in his publication, Gross advocated early amputation as the only treatment modality for extremity osteosarcoma with dismal survival outcomes. Since then the overall outcome of osteosarcoma has seen a sea change. This journey of evolution of treatment of osteosarcoma has been a roller coaster ride with its ups and downs. The world has seen remarkable survival improvements with some rapid strides in 1970s and 1980s. Introduction of multi-agent chemotherapy improved the 5 year survival from a dismal 20% to almost 70%. This fast paced growth reached a stagnant phase with no further improvements in the survival in almost last three decades. Though multiple agents have been tested in both phase II and phase III randomized controlled trials, none has been significant enough to be incorporated in clinical practice. Similar to the oncological outcomes, functional outcomes have also seen a dramatic improvement over last half a century. Limb salvage has become a norm in today's orthopedic oncology practice, which wasn't so in 1970s. Advent of neo-adjuvant chemotherapy, refinements in surgical skills, availability of durable metallic endo-

prosthesis have led to a “limb salvage revolution” where about 85% to 90% of extremity osteosarcoma patients use their own extremity at the end of the treatment. The exponential growth in function preservation still continues with more technology driven innovations and solutions making the commonly encountered implant related complications like aseptic loosening and frequent breakage, a thing of the past. The current standards of care warrant a multi-disciplinary approach in the management of osteosarcoma. The approach is not only required in treatment phase involving a multi-agent neo adjuvant chemotherapy followed by optimal surgical resection & reconstruction and adjuvant chemotherapy, but also in evaluation, diagnosis and staging process. A seamless integration between musculoskeletal surgeon, musculoskeletal radiologist and sarcoma pathologist can achieve higher levels of accuracy in diagnosis in order to initiate the optimum line of treatment within an ideal time frame.

The presence of metastatic disease at presentation is one of the most significant negative prognostic factors. Western data have shown that about 15% to 20% of patients will have clinically detectable metastases at presentation with lung being the most common site of metastasis in about 85% of cases followed by bone as the second most common site. These figures may be higher in developing countries as patients typically present with large volume disease. Lack of awareness, belief in alternative medicine and poor socio economic status are some of the factors contributing to higher percentages in these countries. Delay in diagnosis due to lack

of suspicion and inappropriate initial evaluation as well as management has also led to dismal outcomes. The present symposium on Osteosarcoma tries to address the above issues and provide evidence based robust data, which will help the clinicians to understand the principles for evaluation and management of extremity osteosarcoma. The importance of understanding the presenting symptomatology and clinical evaluation is well scripted in the first article [1]. This article also stresses the importance of multi disciplinary strategy to diagnose a suspected bone lesion correctly. It discusses in depth the role of sequential radiological and histopathological evaluation of a suspected case of osteosarcoma. Staging of osteosarcoma is also discussed, which eventually helps clinicians to plan the treatment and estimate the prognosis. Radiological evaluation, whether it is with radiographs or with high end cross sectional imaging, has been the cornerstone for diagnosis and the local staging of the disease. Osteosarcoma exhibits various radiological and histological forms, which have deep implications on their treatment. These varied radiological presentations are discussed in the second article, which gives a tabulated comparison of various characteristics and their differentials [2].

The modern era is dominated by technology driven tools and this surge is quite evident in evaluation of bone tumors too. Emergence of PET scan as a “one stop shop” for the evaluation of bone tumors has created some whirlpools, leading to unending debates in recent era. Though more data is being collected to prove its worth in osteosarcoma, it is now being used to replace invasive investigations in other tumors like Ewing sarcoma and chondrosarcoma. The third article discusses the use of PET scan as a single modality to stage as well as to assess the chemo response evaluation in osteosarcoma [3]. The recent advances of this bio-imaging tools and the probable futuristic avenues are addressed in the third article.

Complete surgical resection has been the single most important criteria to achieve

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adequate local control. Local relapses are associated with very poor overall survivals. The surgeons need to work to achieve a fine balance between complete tumor resection and retaining function. Over the years this has been addressed by “concept of margins” which was first popularized by the godfather of musculoskeletal oncology, Dr. W.F. Enneking. The concept was further revisited and modified by Kawaguchi, who gave the concept of “barrier effects” and challenged the traditionally propagated concept of “quantitative margins” and replaced it with a new concept of “qualitative margins”. The fourth article in the symposium address the similar issues regarding the adequacy of margins in the resection of osteosarcoma [4]. The article also explains the relationship of local failures with respect to resection margins

and tumor necrosis.

As discussed earlier multi-agent chemotherapy forms an integral part of management of osteosarcoma. The next article details the evolution of various chemotherapy protocols and current standards of chemotherapy for osteosarcoma [5]. Osteosarcoma is considered as a radio-resistant tumor, thus radiotherapy had a limited role in the management of osteosarcoma. Similar to the other specialties of medicine, radiation oncology has seen major changes in understanding the mechanism of radio tumor kill and also in the development of the delivery system. The advent of high-end technique like carbon ion and proton beam radiotherapy with their high accuracy, ability to give very high focused dosage and reduced side effects have made

radiotherapy a new tool in the armamentarium for local control in osteosarcoma. Though these techniques are more useful in non-resectable lesions of axial skeleton, these are becoming increasingly popular in margin positive cases to avoid amputations. The latest updates regarding the use of this modality are explained in the last article of the symposium [6]. This symposium on osteosarcoma has been divided in to two sets, this first set encompasses articles related to evaluation and overall management of extremity osteosarcoma. Next issue will contain the second half of the symposium which will have articles on surgical management and advances in the management of osteosarcoma.

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