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PERIOSTEAL OSTEOSARCOMA – DOES INTRAMEDULLARY EXTENSION PORTEND MORE AGGRESSIVE BIOLOGY?

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INTRODUCTION: Osteosarcoma is the most common primary malignant tumor of the bone accounting for approximately 35% of all primary malignant bone tumors. Periosteal osteosarcoma an uncommon variant of osteosarcoma is an intermediate grade sarcoma usually arising from the surface in the diaphysio-metaphyseal region of a long bone and accounts for less than 2% of all osteosarcomas. The term "periosteal osteogenic sarcoma" and its distinct clinico-pathological description was given by Unni and colleagues in 1976 who initially believed that intramedullary extension excluded the diagnosis of periosteal osteosarcoma. Medullary involvement in periosteal osteosarcoma was later recognized by Spjut in 1977 and confirmed by Hall in 1985. Though some authors mention that extension into the underlying endosteum is extremely rare, intramedullary extension has been reported to be as high as 69 % in some series. While wide surgical excision is the treatment of choice in periosteal osteosarcoma, the role of chemotherapy in the management of these lesions is unclear.

QUESTIONS: We asked if intramedullary extension in periosteal osteosarcoma was indicative of more aggressive biological behaviour leading to poorer overall survival in these patients

PATIENTS AND METHODS: 1226 cases of osteosarcoma were retrospectively identified from our prospectively maintained data base of more than 7000 cases operated between January 2001 and December 2012. These included 20 cases (1.6%) of periosteal osteosarcoma. There were 14 males and 6 female patients. The mean age at presentation was 16 years (range 5-26 years). Tibia was affected in 10 cases, femur in 8 cases followed by humerus and radius in one case each.

One patient had a prior curettage. In the absence of original imaging it was not possible to determine whether there was initial intramedullary involvement. Hence 19 cases were evaluated for intramedullary involvement. All the patients were non metastatic at presentation as determined by staging studies which included a CT scan of the chest and total body scintigraphy.

As per institutional protocol, treatment with 3 to 4 cycles of neoadjuvant multiagent chemotherapy followed by surgery and subsequent adjuvant chemotherapy had been recommended for all patients. Eighteen cases received chemotherapy as per the existing hospital protocol. In one patient chemotherapy was deferred due to associated co-morbidities and another patient was unwilling for chemotherapy.

Eighteen patients had a limb sparing resection, 1 had an amputation and 1 patient had a rotationplasty. The resected specimens were assessed for surgical margins and marrow involvement. Surgical margins were reported free in all patients.

RESULTS: On histopathology evaluation, intramedullary involvement was found in 8 of 19 patients (42%). All patients were available for follow-up. Subsequent pulmonary metastasis occurred in 4 cases (20%) at 57 months, 12 months, 11 months and 9 months follow-up. Two of these patients also had local recurrence. Intramedullary involvement was seen in 3 of these 4 cases with pulmonary metastasis. All four patients had unresectable pulmonary metastasis and eventually succumbed to the disease.

Fifteen patients are currently alive and continuously disease free. The median follow up of survivors was 82 months (24-140 months). Overall survival at 5 years was 78 %. Patients without marrow involvement had a better overall survival at 5 years as compared to patients with marrow involvement (90% vs. 63%), however the log-rank test showed no significant difference (p=0.203) .

DISCUSSION: Our study is a retrospective study with small numbers. The paucity of numbers is unavoidable in a single institutional study reporting on an uncommon lesion. The median follow up of survivors at 82 months (though midterm) does not appear to be a drawback as there does not seem to be a substantial risk of late recurrence in periosteal sarcomas, as all instances of local recurrence and progression to metastatic disease generally occur within 36 months of presentation. We believe that this study can add to the increasing understanding of the outcome of this uncommon variant of osteosarcoma. Such single institutional observational studies help in generating and supplementing outcome data which eventually may help establish guidelines for the management of these lesions.

CONCLUSIONS: While surgical excision remains the cornerstone of treatment of periosteal osteosarcoma, our data suggests that intramedullary involvement may be indicative of more aggressive biology of the disease. As the role of chemotherapy is still unproven in this lesion, whether this is a subgroup of patients that may benefit with more aggressive treatment remains an avenue to be explored. This is best done in the setting of a prospective multicentre trial in view of the uncommon nature of this variant of osteosarcoma.