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Case Report

Two Cases of Extraskeletal Osteosarcoma with Divergent Pathogenesis

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ABSTRACT

Background- Extraskeletal osteosarcoma (ESOS) is a rare, aggressive, malignant high grade mesenchymal neoplasm, predominantly occurs in lower extremities and rarely involves the visceral organs. Till now only 6 cases of mesenteric ESOs have been reported in the literature. ESOS affects older age group than osteosarcoma of bone. ESOS has poor outcome with high rate of mortality around 70-80%. Our both the patients are now dead.

Case Presentation- we hereby present two cases of ESOS. Case 1 is 75 year old male presented with acute abdomen diagnosed with mesenteric ESOS. Case 2 is 65 year old female presented with globoid mass over gluteal region diagnosed as ESOS with history of therapeutic radiation 5 years back for carcinoma of cervix. Both the cases were diagnosed on histopathology and one case was supported with IHC findings.

Conclusion- ESOS should be considered as a differential diagnosis for any malignant mesenchymal tumor of abdominal cavity and tumor developing in post radiation cases specially the area which is irradiated. Radical surgical excision is mainstay of treatment and more aggressive treatment using neoadjuvant chemotherapy and radiotherapy as a multimodality treatment for management is necessary

Keywords- Osteosarcoma, Extraskeletal osteosarcoma, Histopathology, Malignant mesenchymal neoplasm, Outcome.

INTRODUCTION

Extraskeletal Osteosarcoma (ESOS) is a malignant mesenchymal neoplasm capable of osteoid, bone or chondroid matrix production, located in the soft tissues and without connection to the skeleton as determined by radio-imaging and intra-operative findings. [1-5] ESOS are rare malignancies account for 1% of all soft tissue sarcomas. [3,6,7]The term was first used by Wilson in1941. [6]

In contrast to osteosarcoma of bone which primarily affects adolescents and young adults, ESOS commonly affects older age group. [6,8] Etiology of ESOS is unclear. [5] Evolution of ESOS from myositis ossificans, therapeutic radiation, chronic irritation, trauma, at site of intramuscular injection and malignant transformation in previously benign heterotopic bone has also been reported. [4,6] ESOS generally involves extremities and rarely visceral organs. [3]

ESOS have poor prognosis with 70-80% mortality. [2]

We hereby present two cases of extraskeletal osteogenic sarcoma with divergent pathogenesis reported within two years.

Case I – Mesenteric ESOS

A 75 year old male presented with acute abdomen. Patient was known case of diabetes mellitus and hypertension and was on the treatment for the same. Clinical diagnosis of acute intestinal perforation was made and was subjected to laparotomy. No radio-imaging was done as patient was undertaken for emergency operation. Patient had no history of radiation or trauma.

Intraoperative findings were multiple, small, firm to hard nodules over jejunal mesentery and two jejunal perforations were noted. Jejunal resection was performed. Grossly approximately 60 cm long jejunal loop, serosal aspect of which showed multiple grey white, firm to hard globular nodules largest measuring 2.5cm in diameter. On cut section nodules showed solid grey white appearance. At two places jejunal segment showed perforation.(fig 1)

Microscopically sections from the nodules reveal a tumor composed of abundant lacy osteoid lined by neoplastic cells(fig 2,3,and 4). Tumor cells are large, pleomorphic, spindle or polygonal having large hyperchromatic nuclei and scanty eosinophilic cytoplasm. Many bizzare cells, tumor giant cells and mitotic figures are seen. Areas of necrosis and foci of calcification are also seen. On IHC tumor gave immunopositivity for vimentin (fig 5) and negativity for cytokeratin, desmin, CD34, muscle specific antigen

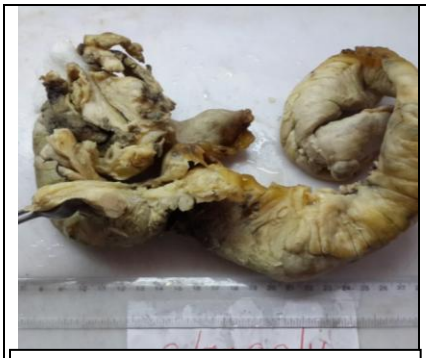

Fig 1- Gross- jejunum showing multiple small firm to hard nodules on mesentery

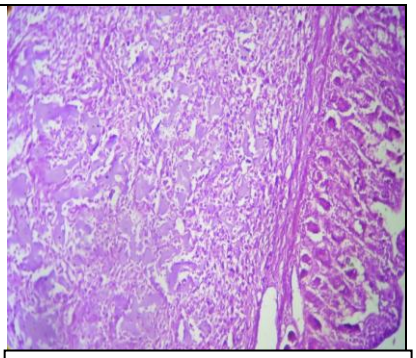

Fig 2- Section from jejunaum with mesentery showing tumor in mesentery. (H&E 100x)

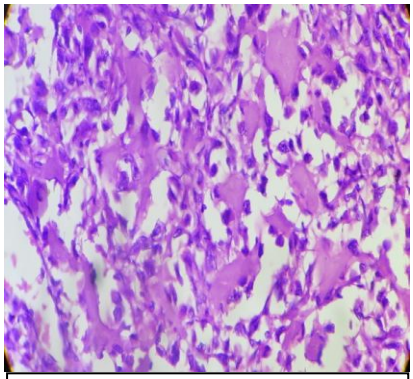

Fig 3- Neoplastic cells are lining the lacy osteoid. (H&E 100x)

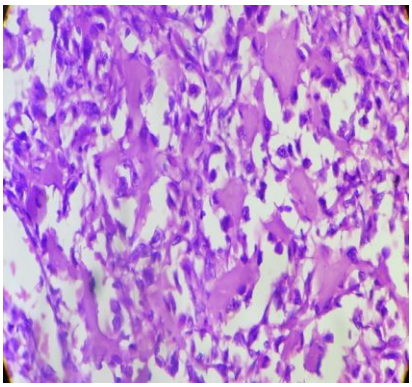

Fig 4- Neoplastic cells are lining the lacy osteoid. (H&E 400x)

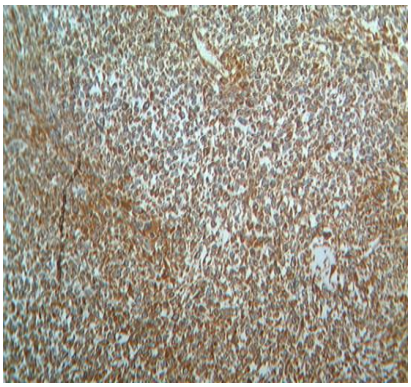

Fig 5-Tumor is immunopositive for Vimentin.

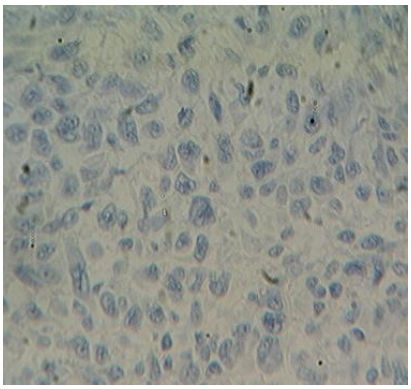

Fig 6- Immunonegative for Desmin

Case II

A 55 year female presented with globoid mass over gluteal region of 6 months duration.

Clinically mass was 5x4 cm in size over gluteal region. Patient was a diagnosed case of carcinoma of cervix 5 years back and completed radiation for the same.

CT scan revealed an ossified lesion with no connection with near bone i.e. pelvic bone.(fig E)

Grossly it was a single well circumscribed gray white nodular mass measuring 5.5x4.5x3cm. Cut section was solid gray white in colour with areas of calcification. (fig A)

Microscopically the tumor revealed plenty of lacy osteoid lined by neoplastic cells. Tumor cells are large, pleomorphic, spindle or polygonal having large hyperchromatic nuclei and scanty eosinophilic cytoplasm. Many bizzare cells are seen. Areas of calcification were noted.(fig B, C and D)

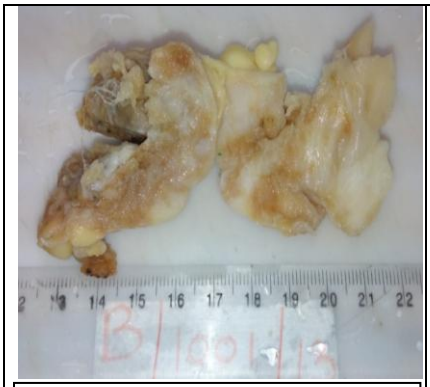

Fig A- Gross- globoid tumor cut section of which showing hard gray white

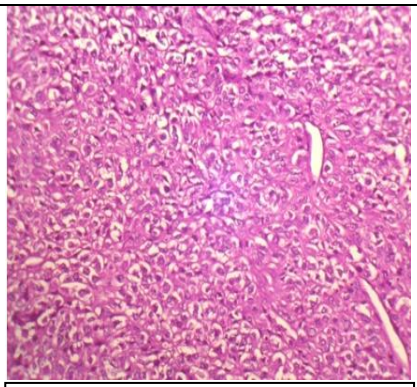

Fig B- Section showing neoplastic cells with osteoid. (H&E 40x)

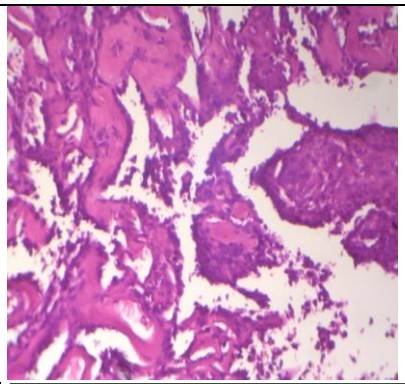

Fig C- Neoplastic cells are lining the lacy osteoid. (H&E 100x)

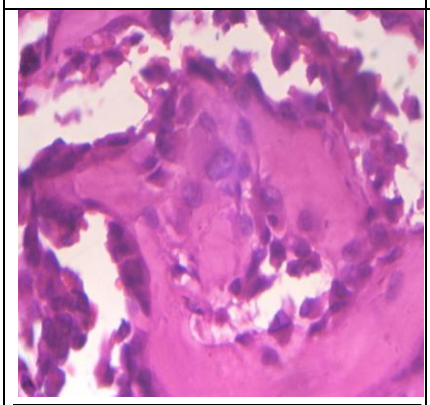

Fig D- Neoplastic cells are lining the lacy osteoid. (H&E 400x)

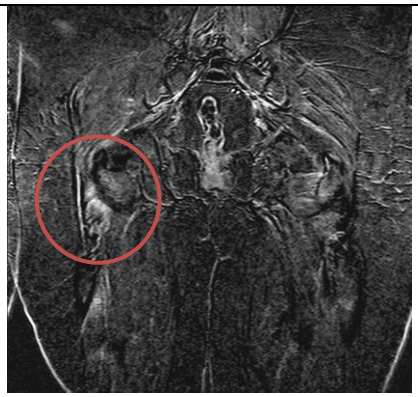

Fig E- CT scan revealed ossified mass with no connection with pelvic bone.

DISCUSSION

Extraskeletal osteosarcoma is a rare soft tissue tumor which has been reported to constitute approximately 4% of osteosarcomas. [8]

ESOS affects an older age group than osteosarcoma of bone with male preponderance. [2,6,8,9] Male to female ratio is 1.9:1. [1] Median age of presentation is 61

years. We present 1 male and 1 female case in old age greater than 60 year.

Tumor affects principally extremities: thigh and buttocks being the most common sites followed by retroperitoneum. [4] One of our cases is seen arising as a common site i.e. buttock, however site of the other case i.e. mesentery is extremely rare; only 6 cases have been reported in the literature till now. ESOS arising from unusual sites like kidney, esophagus, small intestine, liver, gall bladder, mesentery, parotid, breast, uterus, larynx, hand and cerebellum have been reported. [5]

Common presenting symptoms of ESOS are swelling and pain. [4] One of the two cases presented with similar complaints and other case had very rare presentation as acute abdomen. X ray, CT scan and MRI these radio-imagings show soft tissue masses with focal to massive areas of mineralization and lack of osseous involvement. Many other soft tissue sarcoma and carcinoma can also present with significant areas of calcification therefore, Histopathological examination is must for definite diagnosis. [3,5] Microscopy reveals tumor/neoplastic spindle cell proliferation with presence of lace like pattern of osteoid matrix/immature bone formed by neoplastic cells. Histologically six types are known – [3] osteoblastic, fibroblast, chondroblastic, telangiectatic, small cell and well differentiated type. IHC supports the diagnosis. Neoplastic cells give immunopositivity for vimentin and other markers like desmine, epithelial membrane antigen, cytokeratin give immunonegativity. [3] IHC was possible only in one of two cases and findings were similar as reported in other studies i.e. vimentin positive and cytokeratin, CD34, desmin, epithelial membrane antigen are negative.

Differential diagnosis: Various benign and malignant soft tissue osteogenic lesions can mimic osteogenic sarcoma. [10] ESOS can be mistaken for MO which is a benign ossifying process usually occurs in young adults as a single, well circumscribed mass with skeletal muscles and microscopically shows zoning effect with peripheral differentiation into well formed bone with no cytological atypia. But malignant osteoid tend to show reverse zonation by localizing towards the center, in contrast to MO. [6,10] ESOS may resemble MFH in cases where osteoid is absent. So careful sampling is necessary to avoid misdiagnosis of ESOS as MFH. Both with respect to clinicopathological features and therapeutic response ESOS behaves more like MFH than osteosarcoma of bone. [4,8]

Management

As ESOS are very aggressive tumors, recurrence and metastasis of tumor are well known even after adequate therapy. ESOS carries poor prognosis. Mortality is very high 70-80% cases. [4] Thus, more radical treatment is advised for ESOS. Our both the patients reported with ESOS are dead. Adjuvant chemotherapy, radiotherapy are part of multimodality approach in initial management of patient. [4] Tumor size and mitotic figures were found to be prognostic factors. Tumor size greater than 5 cm is predictor of worst outcome. [6,8 ,10] Etiology of ESOS is unclear. [5] Radiation is well documented as a predisposing factor for both bone and soft tissue sarcoma. [6,8,9] development of ESOS as a late effect of therapeutic irradiation is also documented. [7,9,11] One of our cases is of radiation induced sarcoma. Patient received cycles of radiation. Radiation induced ESOS develops at least 4 years following high dose radiation. [3,5] One of our two cases fits into this.

CONCLUSION

ESOS is a malignant, aggressive mesenchymal tumor capable of forming malignant osteoid with no connection to

bone, having poor prognosis with high rate of recurrence, metastasis and mortality. ESOS should be considered as a differential diagnosis for any malignant mesenchymal tumor of abdominal cavity and tumor developing in post radiation cases specially the area which is irradiated. Radical surgical excision is mainstay of treatment and more aggressive treatment using neoadjuvant chemotherapy and radiotherapy as a multimodality treatment for management is necessary.

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