INTRODUCTION

Extraskeletal Ewing’s Sarcoma (EES) is an uncommon, rapidly growing, round cell malignancy of uncharacterized mesenchymal origin. The most frequent sites of occurrence are the chest wall, lower extremities and paravertebral region. Less frequently, the tumor occur in the pelvis and hip region, the retroperitoneum and the upper extremities. It occurs predominantly in young adults and adolescents between the ages of 10 and 30 years and it follows an aggressive course with a high rate of recurrence. Distant metastases are also common in EES. The age at the time of diagnosis, unlike its osseous counterpart, has a wide range, from infancy to the elderly, and has a slight predominance in male patient. We present a case of rare tumor - Extraskeletal Ewing’s Sarcoma at a rare site.

CASE REPORT

A 15 year old boy presented with a non healing ulcer over right foot associated with pricking type of pain. History of itching and profuse bleeding was also present. On examination there was a swelling of 12x6 cm size with ulceration over lateral side of right foot. CECT showed a 12x6.8cm sized lobulated heterogeneously enhancing soft tissue mass over lateral side of right foot. FNAC was suggestive of a round cell malignant tumor. Excision biopsy revealed the diagnosis of Extraskeletal Ewing’s tumor, which was confirmed by special stain with PAS and immunohistochemistry for CD99 and Synaptophysin. Patient responded well with chemotherapy.

REFERENCES:


ABSTRACT

Introduction: Extraskeletal Ewing’s Sarcoma (EES) is an uncommon round cell malignant tumor of uncharacterized mesenchymal origin.

Case Report: We present a rare case of extraskeletal ewing’s sarcoma of leg in a 15 year old boy. FNAC was suggestive of a round cell malignant tumor. Excision biopsy revealed the diagnosis of Extraskeletal Ewing’s tumor, which was confirmed by special stain with PAS and immunohistochemistry for CD99 and Synaptophysin. Patient responded well with chemotherapy.

Conclusion: Extraskeletal Ewing’s sarcoma although rare in soft tissues, it should be considered in the differential diagnosis of round cell malignancies.

Key words: Extraskeletal, ewing’s sarcoma, immunohistochemistry.
tissue density mass lesion noted in soft tissues of lateral aspect of plantar surface right foot with ulceration suggestive of malignant lesion. Bone scintigraphy revealed primary lesion in the right foot.

FNAC was done which revealed predominantly singly scattered and few loosely cohesive clusters of monotonous small round ovoid cells with scant cytoplasm, round to ovoid dark nucleus, scanty fragile cytoplasm and occasional prominent nucleoli. A few rosettes noted. Occasional mitotic figures noted.

Edge biopsy was done and sent for histopathological examination which showed features consistent with malignant small round cell tumor. The cells show round to oval nucleus with fine powdery chromatin, 1-2 minute nucleoli and scant to moderate amount of pale eosinophilic to vacuolated cytoplasm.

Mitotic activity is 10-12 MF/ 10 HPF. There is admixture of small pale round cells with darker staining cells. Many vascular channels are noted and they are surrounded by blue round cells. Areas of hemorrhage and necrosis are noted.

Special stain with PAS showed positivity in the cytoplasmic vacuoles.

IHC showed immunoreactivity for CD-99 and Synaptophysin.
DISCUSSION

The most frequent sites of occurrence of Extraskeletal Ewing’s Sarcoma (EES) are the chest wall, lower extremities and paravertebral region.\[^{1}\] It occurs predominantly in young adults and adolescents and it follows an aggressive course with a high rate of recurrence.

EES is similar to osseous ES and is often confused with other round cell tumors but recent advances in IHC have helped in diagnosis of EES.\[^{2}\]

This family of tumors shares common cytogenetic and molecular changes which involve of Ewing’s sarcoma gene on chromosome 22(22q12) on to a number of other genes like FLI-1 on chromosome 11(11q24) in 90% of cases and ERG on chromosome 21( 21q22). The tumors also share expression of glycoprotein surface antigen p30/32 (MIC2) CD99, a cell membrane protein of unknown function.\[^{3}\]

Five-year survival rates of EES between 38 and 67%. Although the prognosis for this tumor is poor, an early and adequate surgical resection followed by adjunctive chemotherapy and radiotherapy for microscopically positive surgical margins improve the survival rate.\[^{1,2}\]

CONCLUSION

Extraskeletal Ewing’s sarcoma although rare in soft tissues, it should be considered in the differential diagnosis of round cell malignancies. Immuno histochemistry helps in the confirmation of histopathologically diagnosed cases.
CONFLICT OF INTEREST:
The authors declared no conflict of interest.

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