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Case Report

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Extra Skeletal Ewings Sarcoma Presenting as MUO Neck

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Introduction

Ewings sarcoma (ES) is a locally aggressive bone malignancy that is observed most frequently in long bones. The tumor is rarely noticed in the head and neck region and includes only 1% to 4% of all cases [1]. Extraskeletal Ewing's sarcoma (EES) is an uncommon, rapidly growing, round-cell malignancy of uncharacterized mesenchymal cell origin. Tefft et al first described EES in 1969 to be histologically similar to primary Ewing's sarcoma of bone [2]. It occurs predominantly in adolescents and young adults between the ages of 10 and 30 years and it follows an aggressive course with a high rate of recurrence. Distant metastases is also common in EES. We present a case of EES as a neck mass in a 32-year-old female.

Case Report

A 32-year-old woman presented to us with a left neck swelling. Physical examination showed a 3 × 3 cm firm, non-tender, mobile mass in the left posterior cervical region. There was no other abnormality in the head and neck. CT scan of the head and neck showed no evidence of primary elsewhere and an excision biopsy of the lesion was done.

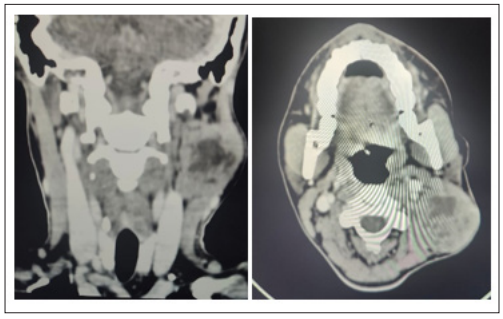

Figure 1: CT Images of nodal mass

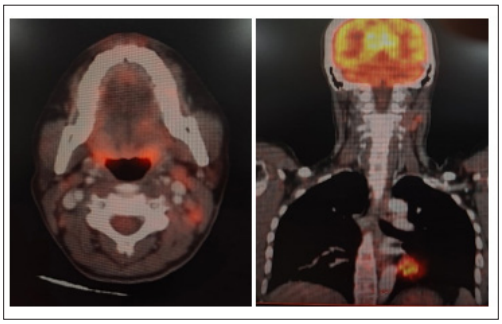

Figure 2: PET CT showing no other pathological uptake

Macroscopically, the tumor was mucoid cystic mass and histopathological examination showed features consistent with small round cell tumor. Immunohistochemistry showed immunoreactivity to MIC 2 protein, CD99 and vimentin and nonreactivity to cytokeratin, actin, neuron-specific enolase and leukocyte common antigen. Thus, histological and immunohistochemical findings were confirmatory for EES.

Whole body PET scan revealed no other metabolically active lesions apart from a few similar lymph nodes in level II and III in the left side of the neck. The patient was treated with multiagent chemotherapy (VACM protocol) followed by left Modified Neck dissection (MNDIII). After 12 months of treatment, the patient was tumor free with no clinical or radiological evidence of tumor recurrence.

Discussion

EES is a rare malignant neoplasm of uncharacterized mesenchymal cell origin. EES usually arises in the soft tissues of the trunk, paravertebral region, intercostal area, lower extremities and pelvis [3]. The head and neck region is an

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unusual primary site for this type of tumor. There is no sex predilection for this condition prior to adolescence, however, the number of males affected is slightly higher than the number of females. Allam et al has reported a male to female ratio of 2.4:1 [5]. The histological image of EES is a small, blue, round cell with a scanty cytoplasm and it is often confused with other small round cell tumors. Angerwall and Enzinger first reported the pathological features and behaviour of ESS, which are round or oval cell sarcoma of soft tissue and are histologically indistinguishable from ES of bone [3].

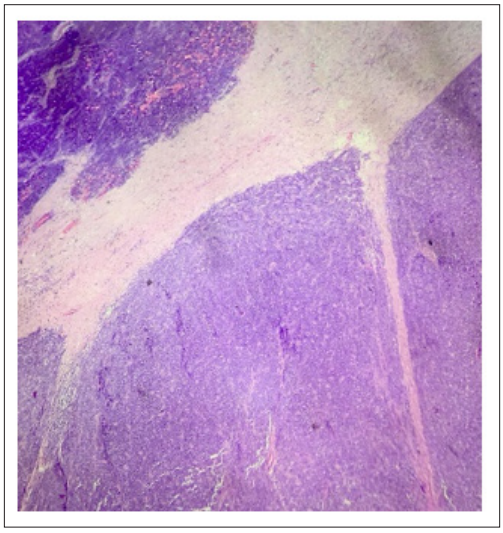

Figure 3: 4X: Tumour Cells arranged in islands

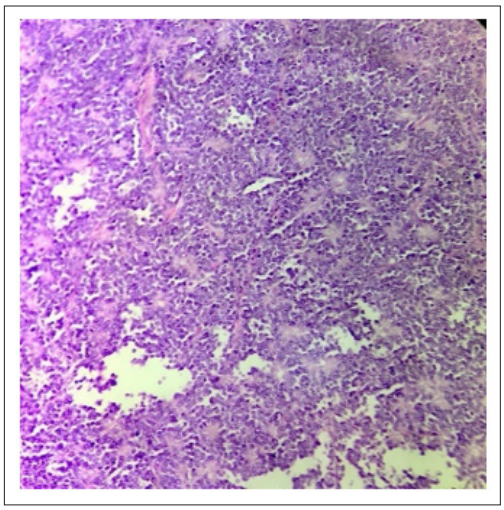

Figure 4: 20X: Showing small round blue cells suggestive of round cell tumour

Tumours share expression of glycoprotein surface antigen p30/32 (MIC2) CD99, a cell membrane protein of unknown function [6]. EES is sensitive to multimodality treatment. Early awareness and wide resection followed by chemotherapy and radiotherapy might improve the long-term survival of patients with extra skeletal Ewing's sarcoma. The rate of survival in EES has improved from 8% to nearly 65% in patients with extremity lesions and to 83% in patients with truncal lesions, and the prognosis is now approaching that of skeletal Ewing's tumors [4,7]. The aims of surgery are to improve or preserve the quality of life, achieve locoregional control and long-term survival.

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