CASE REPORT

EXTRA OSSEOUS PRIMARY EWING'S SARCOMA

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The case of 20 years old boy with an extra osseous Ewing's sarcoma is described. He was initially diagnosed as a case of infiltrative malignant tumour of left suprarenal gland on the basis of preoperative workup but postoperative biopsy of surgically excised specimen confirmed Extra-osseous Ewing's Sarcoma (EES) suprarenal gland with no evidence of malignancy on skeletal scintiscan, bone marrow aspirate and histopathology. Suprarenal location of primary EES is unknown and probably has not been reported in literature. We report a unique case of EES.

Keywords: Ewing's sarcoma, extra osseous, suprarenal gland, management

INTRODUCTION

Ewing's sarcoma was first described by James Ewing in 1921.[1] It is a rare malignant tumour commonly arising in the nervous tissue of long bones or in soft tissues in children and young adults. Histologically it has both mesodermal and ectodermal elements hence its classification is difficult.[2] It is predominantly reported in males with male/female ratio of 1.6:1. A common genetic locus with chromosomal changes in a cell's DNA is responsible in large percentage of cases.[3] It can spread to adrenal gland and other soft tissues. Extra osseous Ewing's sarcoma is a rare soft tissue sarcoma and is composed of small undifferentiated round to oval cells histologically indistinguishable from osseous form. Its diagnosis is based on the basis of histological findings with no evidence of bony involvement at the time of presentation. There are only a few case reports of extra osseous Ewing's sarcoma which are diagnosed on the basis of MRI findings. Hence this is a first ever novel case reported locally.

CASE REPORT

A 20 year old healthy boy presented with a history of dull ache in left flank since last one month. There were no urinary or gastrointestinal symptoms. His vitals, general physical and systemic examinations including abdominal examination were unremarkable. There was no other associated co-morbid condition. Ultrasound revealed a mass at upper pole of left kidney suggestive of supra renal tumour. His serum cortisol, urinary VMA and urinary cortisol were within normal limits. CT scan chest was unremarkable however abdomen revealed a malignant mass in left adrenal gland with infiltration of upper pole of left kidney. MRI revealed a heterogeneous mass measuring 11×9 Cm at upper pole of left kidney. An ultrasound guided biopsy on histopathology revealed tiny foci of an infiltrating tumour composed of groups and sheets of atypical small round to oval cells with hyper chromatic nuclei and scanty cytoplasm. Tumour markers were not helpful. His baseline investigations were within normal limits. He underwent surgical excision of the Adrenal mass which was partially cystic and separated from upper pole of left kidney and entire mass was sent for histopathological evaluation (Figure-1). Gross specimen consists of a mass measuring 7.5×7×4.5 Cm in size. Cut section revealed a solid grey brown tumour with areas of haemorrhage and necrosis (Figure-2). Microscopic examination showed a neoplastic lesion composed of solid sheets of cells with small round hyperchromatic nuclei with clumped chromatin, small nucleoli and indistinct cell membrane. The solid sheets of neoplastic cells were separated by fibrous septae. Mitoses were frequent along with extensive necrosis (Figure-3). Further evaluation included immunostaining for CD99 and chromogranin. Morphological and immunohistochemical features favoured diagnosis of Ewing's sarcoma. Bone marrow biopsy and skeletal scintigraphy was also negative for metastatic involvement (Figure-4). He had an uneventful recovery and was discharged on 8th postoperative day. He was planned for chemotherapy but the patient was lost to follow-up.

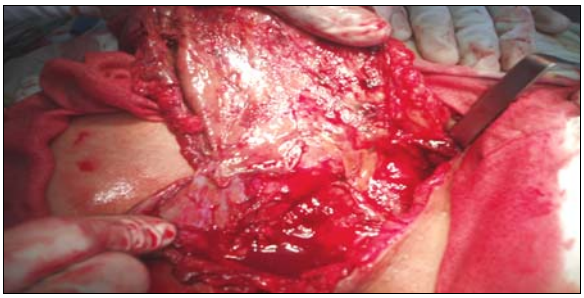

Figure-1: Adrenal mass as seen per-operatively

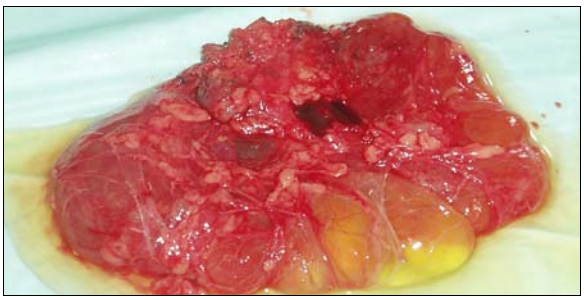

Figure-2: Pathological specimen of the excised adrenal mass

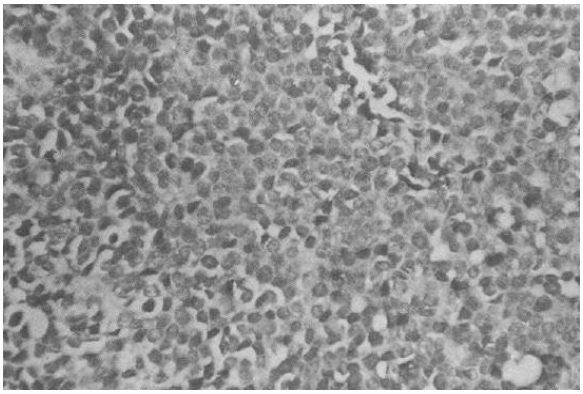

Figure-3: Microscopic examination showed a neoplastic lesion composed of solid sheets of cells with small round hyper chromatic nuclei with clumped chromatin, small nucleoli and indistinct cell membrane

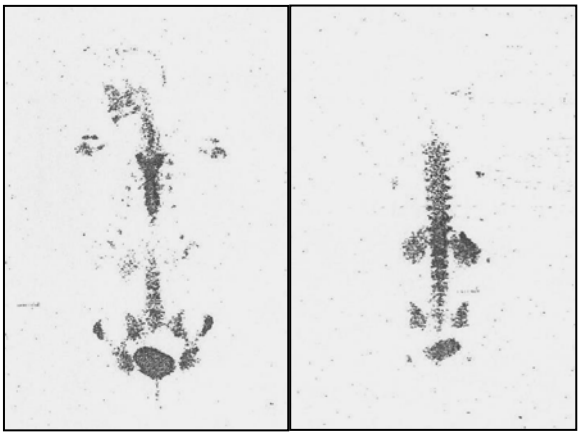

Figure-4: Skeletal scintigraphy (Anterior and Posterior view) findings were negative for metastatic involvement

DISCUSSION

EES has been considered a distinct clinic pathological entity despite its striking microscopic resemblance to ES of bone.[2] It has been described in different locations for example central nervous system, chest wall, retro peritoneum, skin, kidney, small intestine, pelvis, rectum, vagina, thumb, finger, arm, scalp, nasal fosse, lip, toe, Para vertebral and perineum.[4] EES is unknown in supra renal gland; however it can metastise to adrenal gland. The origin of EES has been a matter of research since the times of James Ewing (1866–1943). In recent reports presence of Homer-Wright like rosette, neurosecretory granules, microtubules, positive stain with neural markers, same chromosomal translocation suggest possibility of these belonging to the group of peripheral primitive Neuroectodermal tumours. They do not usually involve nodal sites.[5] The mainstay of treatment is complete excision of the tumour followed by adjuvant chemotherapy and often radiotherapy.[6–8] The systemic chemotherapy includes combination of cyclophosphamide, vincristine, doxorubicin, alternating with ifosfamide and etoposide.[8–9] Radiation is often used for local control postoperatively for positive margins or definitively if the primary cannot be completely excised.

Poor prognosis depends upon the age at the time of diagnosis, bulk of disease, site, and the presence of secondaries.[10] Unfortunately the patient is lost to follow-up despite of counselling for systemic chemotherapy to prevent recurrence. This case report is presented to emphasise the importance of extra osseous Ewing's sarcoma as a possible rare diagnosis and preoperative and postoperative workup to establish a correct diagnosis so that targeted management can be instituted thereby optimizing the potential for curative outcome.

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