Case Report

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Primary Renal Ewing's Sarcoma with Orbital Metastasis: A Rare Case Report

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Abstract

Peripheral primitive neuroectodermal tumour/Ewing's sarcoma (PNET/EWS) is an aggressive type of sarcoma. It is second most common sarcoma in the paediatric–young adult age range. However it is extra ordinarily rare primary tumour in the kidney. The clinical presentation of this tumor is not specific, and other renal tumors may present with a similar histologic appearance. Histopathology and immunohistochemistry are useful in confirmation of the condition. Ewing sarcoma/primitive neuroectodermal tumor carries a strikingly dismal prognosis and early metastasis. Lung, liver, bone being common metastatic sites. However we present case of primary Ewing's Sarcoma with Orbital metastasis which has not been much reported.

Keywords: Extra Osseous Ewing's Sarcoma; Orbital metastasis

Introduction

Ewing's sarcoma and primitive peripheral neuroectodermal tumor (PNET) are high grade malignant tumours found in children and adolescents. PNET is a member of the Ewing's sarcoma/primitive neuroectodermal tumour family of tumours.[1] and is the second most common tumour of bone among children. However its occurrence in soft tissues is very rare. Primary Renal Ewing's sarcoma is a very rare tumour. It was first described in 1975 by Seemayer and colleagues.[2] It is a highly malignant neoplastic condition with tendency towards early metastasis. Most common sites of metastasis are lung and liver, also to bone and lymph nodes.[3] Metastasis to orbit is a very rare occurrence.

Case Report

A 13 year old girl presented with complaints of fever on and off, right sided abdominal pain and abdominal swelling for 2 months. MDCT Abdomen revealed 10.9X13.8X10cm partially necrotic mass lesion arising from Right Upper kidney with lower pole cortex infiltration. Extensive peri-tumoral fat stranding seen. Main Renal artery and vein markedly compressed and displaced by lesion. Multiple discrete lymph nodes seen in medial perinephric region, para-aortic region and mesentry initially presumed to be renal cell carcinoma. (Fig. 1) CT thorax was done which was unremarkable. No abnormality was present in renal function tests, liver profile, Serum electrolytes and complete blood count except normocytic hypochromic anaemia.

Patient then underwent Right radical nephrectomy. No complication was present during operation. Post op period was uneventful.

Histological sections from renal mass showed tumour composed of small round cells with scanty cytoplasm and rosette like bodies, focal areas of necrosis, hyalinization and calcification. (Fig. 2)

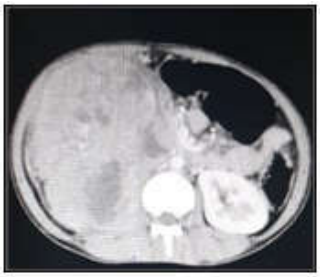

Fig. 1: MDCT Abdomen showing large mass lesion arising from Right upper kidney with lower pole cortex infiltration

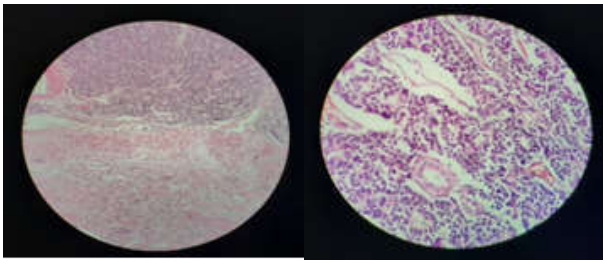

Fig. 2: Histopathological section showing small round cells, with scanty cytoplasm and rosette like bodies

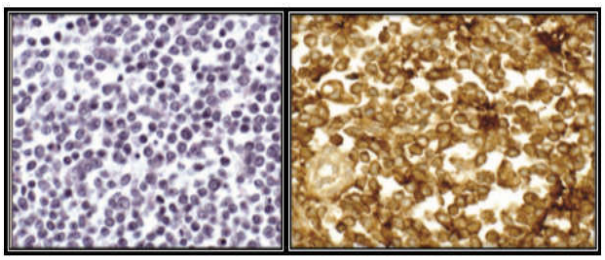

Fig. 3: Strong membranous staining with CD99

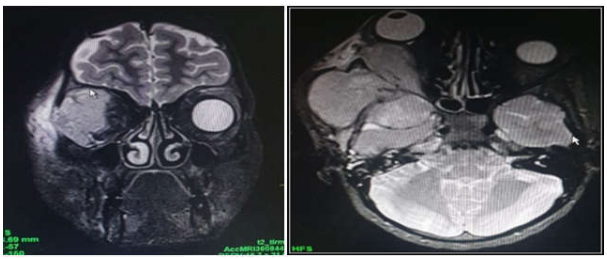

Fig. 4: MRI Orbit reveals soft tissue lesion with intra-orbital extension with invasion of intracanalicular segment of optic nerve

Immunohistochemical studies showed CD99, FL-1, Synaptophysin positive, WT-1 negative Suggestive of- Ewing's sarcoma/PNET. (Fig. 3)

Patient took no adjuvant treatment and lost to follow up. She presented one month later with complaint of pain in right eye and swelling along with diminished vision. On examination there was presence of Right lateral temporal swelling along with Right eye proptosis and conjunctival chemosis. No neck nodes were palpable and rest of clinical findings were normal.

Bone Marrow Smear examination was normocellular. MRI Orbit was suggestive of 6X6X5.7cm soft tissue mass lesion involving lesser and greater Right sphenoid wings with intracranial, extracranial and intra-orbital extension. Intra- orbital soft tissue extension seen along supero-lateral walls of Right orbit with mass effect with invasion of intracanalicular segment of optic nerve. (Fig. 4)

She was then started on chemotherapy receiving 4 cycles of vincristine/Adriamycin/ cyclophosphamide followed by 6 cycles of vincristine/doxorubicin/cyclophosphamide. MRI Orbit revealed partial response for orbital metastasis.

She was then started on palliative radiotherapy for orbital metastasis. Patient was relieved symptomatically of eye pain and swelling reduced. However she took 4# RT and then defaulted to treatment.

Discussion

Ewing's Sarcoma was first described by *Stout* in 1918 in ulnar nerve. It contained small round cells arranged into rosette. Then it was further characterized by John Ewing in Diaphysis of Long Bones. They are neural crest cells in origin belonging to family of primitive neuroectoderm tumours (PNETs). Ewing's sarcoma/primitive neuroectodermal tumour (ES/PNET) is an extra ordinarily rare primary tumour in the kidney.[4] It comprises of <1% solid renal tumours. Sources of primary renal EWS include neural cells that invaginate into the kidney during development.[1,5] There are also other theories according to which embryonic neural cells migrate into the kidney and undergo tumorigenesis.[6] It predominantly affects young adults (mean age 28–34 years) with slight male predominance.[3,6-8] In a case report and meta-analysis by *Hakky* et al[9] flank pain mimicking renal colic was one of the most common presenting symptoms. Our patient also presented with complaints of flank pain supporting their finding. The presence of systemic symptoms such as fever and weight loss are indicative of underlying malignant disease.

EWS/PNET usually histologically is composed of small round blue cells with hyperchromatic nuclei, scanty cytoplasm arranged in rosette.[10] Immunohistochemically it is strongly positive for CD99 and leukemia virus integration (FLI)-1. A large panel of immunochemistry markers is essential for excluding other round cell tumours.

It includes markers like cytokeratin, epithelial, membrane antigen, a WT1, synaptophysin, muscle specific actin, desmine, myogenin, LCA and CD45.

The differential diagnoses from other round cell tumours include rhabdomyosarcoma,Wilms tumour, neuroblastoma, clear cell sarcoma of the kidney, lymphoma, small cell variant of osteosarcoma, desmoplastic small round cell tumor, small cell anaplastic neuroendocrine carcinoma and nephroblastoma.[11]

The imaging characteristics of these tumours mostly comprises of ill-defined, large heterogeneous masses with necrotic and hemorrhagic areas.

Due to the rarity of this tumour, there is no standardized treatment strategy.Complete nephrectomy complemented with resection of othera-ectedorgansorvas culature is the standard surgical therapy[12] with adjuvant chemotherapy and radiotherapy[13,14] is the treatment protocol being followed. The effective chemotherapy agents are vincristine, doxorubicin, ifosfamide, etoposide, actinomycin D and cyclophosphamide. Addition of ifosfamide and etoposide to doxorubicin containing regimens confers a survival advantage with non-metastatic disease.[15] Radiation has shown success as salvage treatment therapy especially in cases of positive lymph nodes following surgery, extracapsular spread of tumours and tumour residues.[3,16]

The condition being highly aggressive, exhibits mostly as early metastatic disease and considered as systemic condition. It most commonly metastasizes to lungs followed by liver, bones and lymph nodes. However in this case we noticed presence of orbital metastasis. There have not been reported cases for Primary Renal Ewing's Sarcoma with Orbital Metastasis. The disease prognosis is very poor and hence low cure rates. One of the main challenges is proper diagnosis and adequate treatment in expeditedtime.[17] In our case the patient was treated with adjuvant chemotherapy followed by palliative radiotherapy for orbital metastasis. Symptomatic relief and partial response was achieved before patient defaulted to treatment.

Conclusions

Primary Ewing's sarcoma is a rare entity in kidney presenting only in <1% of solid tumours. It should be considered as a differential diagnosis in patients presenting with renal mass. The symptoms being non specific. CT and MRI findings are non specific but helpful in local assessment of resectability and metastasis detection. The diagnosis is based on Immunohistochemical and cytogenetic studies. It is an aggressive disease with propensity for early metastasis. Surgical excision with adjuvant chemotherapy and radiation is the treatment modality. The most common sites of metastasis being lungs, liver, bone and lymph nodes. However in our case there was presentation of orbital metastasis which has not been reported much till now. Despite all treatment the prognosis is poor.

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