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An adult case of paratesticular spindle cell rhabdomyosarcoma: rare phenomena

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Abstract: Paratesticular rhabdomyosarcoma (RMS) is an extremely rare malignant neoplasm with aggressive behaviour. It accounts for only 7% of all RMS and usually presents as painless scrotal swelling. Multimodality treatment includes surgery, chemotherapy and radiotherapy have improved the disease outcome .we herein report a case of spindle cell variant of paratesticular RMS in a 20 year old gentleman being treated in our department.
Keywords: Spindle cell, rhabdomyosarcoma, paratesticular, chemotherapy, radiotherapy.

INTRODUCTION

Paratesticular rhabdomyosarcoma (RMS) is the most common non germinal malignant tumor. RMS arises from the mesenchymal tissues of the spermatic cord, epididymis and testicular envelopes. It occurs frequently in children but extremely rare in adults. It is a highly malignant tumor originating from immature striated muscle. It is characterized by the presence of cells having an identifiable striated muscular differentiation with rhabdomyoblasts. Embryonal RMS is the most common subtype of paratesticular RMS. Spindle cell is an uncommon variant of embryonic RMS and is associated with a favourable prognosis in children. We present a case of spindle cell variant of paratesticular RMS in a 25-year-old gentleman.

CASE PRESENTATION

A 25 year old healthy boy presented with prior history of high inguinal orchidectomy for a painless left scrotal mass of 3 week duration diagnosed as paratesticular rhabdomyosarcoma and having received nine cycles of adjuvant Chemotherapy [Ifosfamide, Etoposide/ Vincristine, Adriamycin, Cyclophosphamide (IE/VAC) regimen, 3weekly in view retroperitoneal adenopathy] with good response. Patient was disease free for four months and then presented to us with pain in left flank region since 2 months.

Previous records were reviewed and the radiological and pathological reports were reassessed. High resonance ultrasonography (HRUSG) of scrotum revealed a left intrascrotal heterogeneous lesion with eccentrically displaced left testis, measuring 6.5 x 6.2 cm with epididymis not visualised separately. Epididymis was the probable site of origin. Contrast enhanced computed tomography of thorax abdomen & pelvis (CECT-TAP) showed a large heterogeneously enhancing mixed attenuation lesion measuring 9.5x 7.5 cm in left hemiscrotum with contiguous extension along ipsilateral spermatic cord. Ipsilateral testis was compressed eccentrically but normal. Another heterogeneous lesion measuring 3.4x2.8cm was seen in paraaortic region on left side just proximal to aortic bifurcation. Tumour markers were within normal limits (beta-human chorionic gonadotropin [β HCG], alpha-fetoprotein [AFP], lactate dehydrogenase [LDH]. Metastatic workup was negative. A high inguinal orchidectomy was performed. Cross section revealed structure of testis, epididymis and a highly cellular area in the paratesticular soft tissue. Microscopy demonstrated tumor with variable degree of cellularity with alternating densely packed hyper cellular areas and loosely textured myxoid areas. Tumor consisted of poorly oriented ovoid or elongated or spindle shaped cells with ovoid nuclei and eosinophilic stained cytoplasm. Tumor also had areas of haemorrhage and necrosis and was surrounded by tubules of epididymis extensively. IHC was strongly positive for vimentin & whole muscle actin, focally positive for desmin and myogenin and negative for CK, CD 99, and LCA. Final diagnosis of spindle cell variant of rhabdomyosarcoma was made. Patient was given 9 cycles of adjuvant Chemotherapy [Ifosfamide, Etoposide / Vincristine, Adriamycin, Cyclophosphamide (IE/VAC) regimen, 3weekly] with very good response leading to disappearance of disease in follow up PET-CT scan only reports [no images of the radiological investigations were available].

Physical examination revealed no obvious abnormality. CECT scan (abdomen and pelvis) revealed moderate sized hypodense enhancing lesion measuring 7.2 x 5 cm is seen in pre and left paravertebral region and infrarenal location with mild dilatation of left side pelvicalyceal system (fig 1a & 1b).

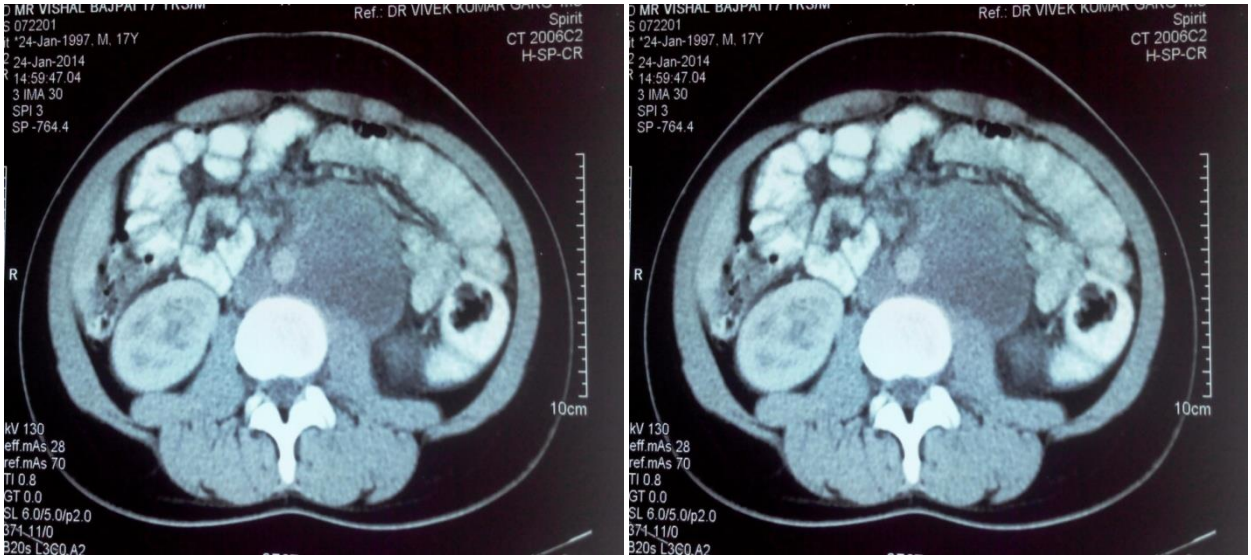

Fig- 1a & 1b: CECT scan (abdomen and pelvis) revealed moderate sized hypodense enhancing lesion measuring in pre and left paravertebral region and infrarenal location with mild dilatation of left side pelvicalyceal system.

Tumor markers were within normal limit. Metastatic workup was negative. Patient was restarted chemotherapy, received 12 cycles of VAC (Vincristine, Adriamycin, and Cyclophosphamide) regimen. Post chemotherapy CECT scan showed isodense, mildly enhancing lesion is seen in left paraaortic region below left renal hilum measuring 4 x 2.2 cm with adjacent fascial planes with aorta and left psoas well maintained (partial response) (fig 2a & 2b).

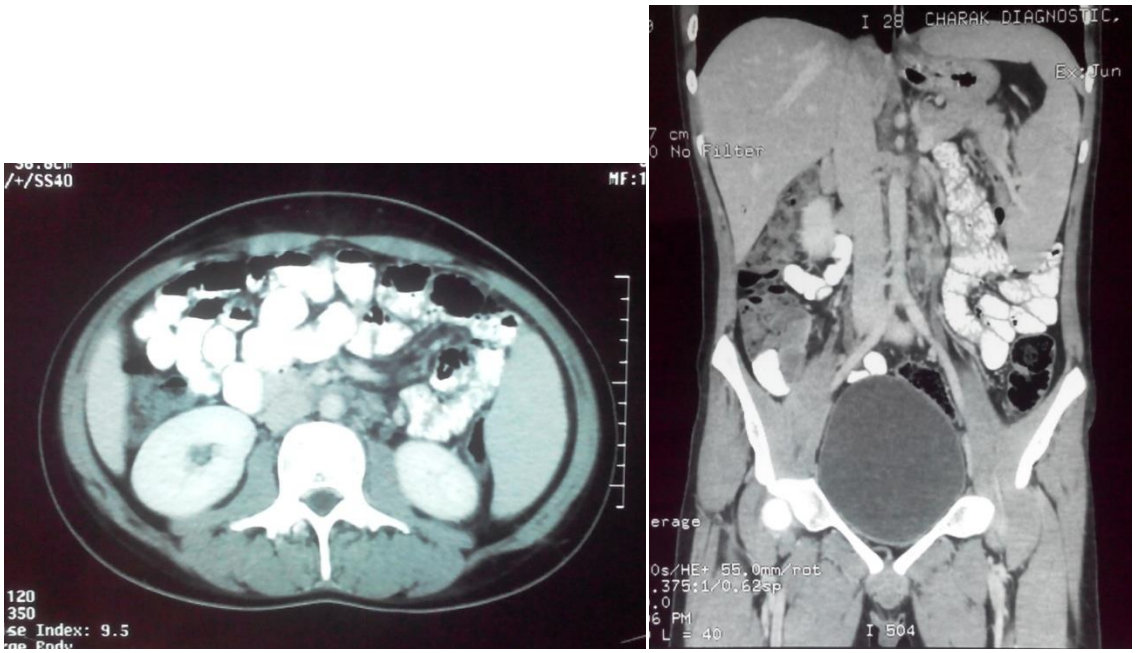

Fig- 2a & 2b: Post chemotherapy CECT scan showed isodense enhancing lesion is seen in left paraaortic region below left renal hilum (decrease in tumour size- partial response).

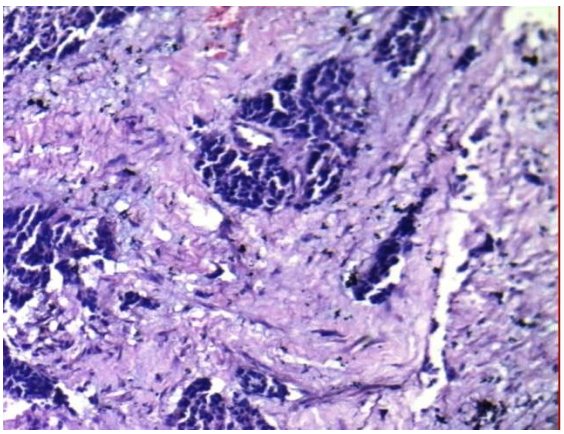

Fig- 3a: Photomicrograph revealed sheets of spindle shaped cells with elongated nucleoli suggestive sarcoma.

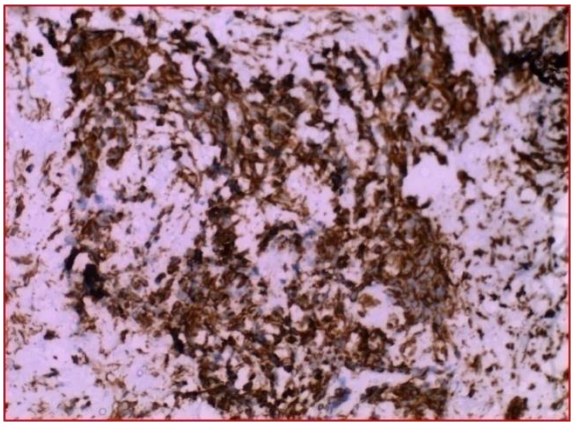

Fig- 3b: IHC examination showed tumour cells positive for Vimentin

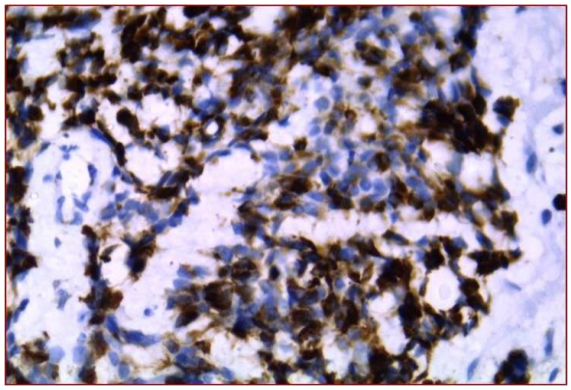

Fig- 3c: Tumor cells showed positivity for Desmin.

In view of good response and no major vessel involvement, patient underwent retroperitoneal lymph node dissection. Histopathology examination revealed well encapsulated highly cellular neoplasm composed of sheets of spindle shaped cells with elongated nucleoli (fig 3a), brisk mitotic activity (>50/50 HPF), large areas of necrosis and fibrosis. IHC was strongly positive for vimentin (fig 3b) & whole muscle actin and focally positive for desmin (fig 3c) and myogenin.

Final diagnosis of spindle cell variant of rhabdomyosarcoma was made. Postoperative course was uneventful and patient remained disease free for 3 months on follow up. After 3 months patient presented with abdomen distension and features suggestive of sub acute intestinal obstruction. On examination, patient was severely cachexic with abdominal distension. CECT scan of abdomen and pelvis showed multiple bilobar hepatic metastases and conglomerated bulky metastatic adenopathy in the retroperitoneum, encasing major vascular structures, omental and peritoneal metastasis with moderate to gross ascitis (fig 4a). There was also moderate sized rounded hypodense lesion in anterior cortex of left kidney (fig 4b), retroperitoneal nodal mass and ascitis and patient later on succumbed to it.

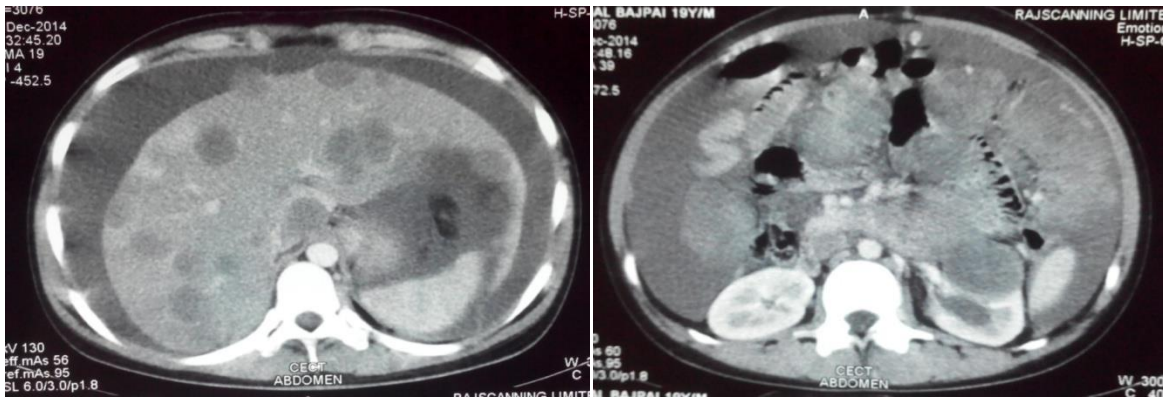

Fig- 4a & 4b: CECT scan (abdomen and pelvis) showed multiple bilobar hepatic metastases and conglomerated bulky metastatic adenopathy in the retroperitoneum with moderate to gross ascitis (fig 4a). There was also moderate sized rounded hypodense lesion in anterior cortex of left kidney (fig 4b)

DISCUSSION

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children, accounting for 3% to 4% of all cases of childhood cancers [1]. Rhabdomyosarcoma arises from a primitive mesenchymal cell and can be found in multiple areas of the body. Most common affected anatomic regions include the head and neck (including the orbit and parameningeal areas, (35%), genitourinary tract (including the bladder, prostate, vagina, vulva, uterus, and paratesticular area; 22%), and extremities (18%) [1] Malignant tumours of the paratesticular region are uncommon. Approximately 30% tumours of paratesticular region are malignant and are sarcomas mainly. Most common histological subtypes of sarcomas are leiomyosarcoma (32%), rhabdomyosarcoma (RMS, 24%), liposarcoma (20%), and malignant fibrous histiocytoma (MFH, 13%) [2]. about 7%-10% of RBS involve the paratesticular region [3]. Paratesticular RMS includes several subtypes, namely embryonal [most common variant], alveolar, and pleomorphic. [4] Spindle cell type is a rare variant of embryonal RMS, and was first reported in 1992 by Cavazzana. Adult spindle cell cases account for only 3% of all RMSs. To the best of our knowledge, only a few case reports exist in the literatures which describe a paratesticular lesion in an adult.

Clinically paratesticular tumour presents as an intrascrotal mass, usually large, sometimes reaching up to the external inguinal ring and compressing the testis and epididymis. In large tumours it may be clinically indistinguishable from testicular tumours. Majority of the patients present with painless testicular swelling [5]. Pain has been reported only in 7% of cases and a hydrocele can be occasionally present [2]. Involvement of retroperitoneal lymph nodes in paratesticular tumours is common and occurs in up to 30% of cases and lung is the common site of distant metastases [seen in 14% of cases] [6]. Differential diagnoses include testicular torsion, epididymorchitis, scrotal abscess and, rarely, testicular tuberculosis.

A testicular ultrasound is routinely performed for a scrotal swelling. This imaging modality shows a mass with heterogeneous echogenicity and inguinoscrotal extension in 80% of cases. [7] This allows the nature of the intrascrotal tissue mass to be determined and specifies the exact topography. CECT scan of thoraco-abdomino-pelvic region allows assessing loco-regional spread especially paraaortic and pelvic metastases as well as possible metastases to the liver or lung. Magnetic resonance imaging is an efficient imaging modality when using surface coils. Tumor appears homogeneous in T1-weighted images and heterogeneous in T2-weighted images with signal intensity similar to that in a normal testis. The low signal intensity of the tunica albuginea in T2-weighted images allows the visualization of a clear separation of the mass from the testis [1, 8, 9]. Tumor markers including alpha-fetoprotein, beta-human chorionic gonadotropin and lactate dehydrogenase levels are usually normal in rhabdomyosarcoma,

A malignant tumour might be suspected in masses present in the distal cord with a hard and irregular form adhering to surrounding structures; a rapid increase in tumor volume may be noted. Final diagnosis is based on histolopathological and IHC examination. Microscopically, spindle cell RMS often displays fascicular elongated cells with central nuclei and eosinophilic fibrillary cytoplasm with a small proportion of rhabdomyoblasts, featuring more eccentric nuclei, striations, and sharper eosinophilia. Immunohistochemical analysis are conducted using a panel of antibodies including myosin and desmin for diagnosis spindle cell RBS. On IHC examination, spindle cell rhabdomyosarcoma stains positive for desmin & myogenin and is virtually confirmed (specificity close to 100% and sensitivity of 95%). [9, 10]

The optimal management of paratesticular RMS remains unclear because of the rarity of the disease in adults. Treatment strategies reviewed in the

literature include radical high inguinal orchidectomy, chemotherapy (CT), radiotherapy (RT) and retroperitoneal lymph node dissection (RPLND). Hermans and colleagues described 19 paratesticular RMS patients treated with RPLND, and claimed that a combination of RPLND and systemic CT afforded a high cure rate [11]. Ferrari and colleagues reported on 44 patients with paratesticular RMS who did not underwent RPLND [4]. Majority , however, consider RPLND as unnecessary for localized disease because of the sensitivity afforded by computed tomography for early detection, the potential RPLND-associated morbidity, the low rate of retroperitoneal recurrence and the presumed efficacy of chemotherapy in controlling of microscopic disease. An alternative approach toward the treatment of clinically enlarged retroperitoneal lymph nodes involves the use of a more intensive adjuvant chemotherapy regimen. Such an approach is based on results obtained in the IRS-III trial, which showed that patients experienced poor outcomes if treated with RPLND followed by Chemotherapy [12]. Chemotherapy can control micrometastases into retroperitoneal nodes when a primary tumor has been completely resected. Ferrari and colleagues reported that CT was effective to treat childhood RMS, in adjuvant setting [4]. The role for adjuvant CT in adults, however remains poorly understood [2, 4]. Vincristine, dactinomycin, cyclophosphamide, adriamycin, epirubicin, ifosfamide, carboplatin and etoposide have been used in different combinations, and with varying dose schedules. CT should be offered as a component of multimodal therapy in patients with paratesticular RMS to control retroperitoneal dissemination and to minimize such dissemination. Generally, spindle cell type paratesticular RMS is associated with a favourable prognosis, with 5-year survival rate of 95% [4]. Additionally, lymph node metastasis is less common in the spindle cell variant compared with non spindle cell variants (16% vs. 36%).

CONCLUSION

Paratesticular RMS is a very rare tumor with suspicious findings on ultra-sonographic examination including solid masses with increasing size and vascularity. Oncologists should have a high level of suspicion for these lesions and need to consider it in the differential diagnosis. Optimal treatment may be radical orchidectomy followed by adjuvant chemo radiation with ill defined role of RPLND.

Abbreviations

RMS- Rhabdomyosarcoma; HRUSG- High resonance Ultrasonography, CECT- contrast enhanced computed tomography; FNAC- Fine needle aspiration cytology; HCG-human chorionic gonadotropin; AFP- alpha-fetoprotein; LDH- lactate dehydrogenase IHC-Immunohistochemistry; CK- Cytokeratin; LCA-leukocyte common antigen CD- Cluster of differentiation; CT- Chemotherapy; RT- radiotherapy; RPLND- Retroperitoneal lymph node dissection; IE/VAC- Ifosfamide, Etoposide / Vincristine, Adriamycin, Cyclophosphamide

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