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Para testicular spindle cell variant of embryonal rhabdomyosarcoma – A case report

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Abstract: Para testicular Embryonal Rhabdomyosarcoma is a rare soft tissue sarcoma, presenting most often in the first two decades of life. Most tumors present clinically as painless scrotal swellings which can be clinically indistinguishable from a testicular tumor. Spindle cell variant of embryonal rhabdomyosarcoma has an excellent prognosis. These tumors are managed with multispecialty approach. Radical orchidectomy with high ligation of spermatic cord is the standard surgical procedure followed by chemotherapeutic agents improving majority of the patient's survival outcome. We report the case of a spindle cell variant of Para testicular embryonal Rhabdomyosarcoma in a 19 year old adolescent presenting with progressive painless scrotal enlargement.
Keywords: Para testicular, Embryonal Rhabdomyosarcoma, Spindle cell, Orchidectomy.

INTRODUCTION:

Primary Para testicular tumors in children and adolescents are rare, accounting for 7 – 10% of primary genitourinary tumors [1]. Majority of the Para testicular tumors present clinically as painless, unilateral scrotal masses with or without associated hydrocele. Approximately 80% of the Para testicular rhabdomyosarcoma (RMS) occur before 21 years of age [2]. Embryonal RMS is the most frequent subtype of RMS in the Para testicular region [3].

CASE REPORT:

A 19 year old male patient presented with a progressively increasing painless scrotal swelling over 3 weeks duration. No history of fever and lymphadenopathy. Ultrasonographic studies of the abdomen showed no iliac, and para aortic lymphadenopathy or liver metastasis. Chest x- ray was normal. His serum tumor markers for β Human Chorionic Gonadotropin and alpha fetoproteins levels were within normal limits. Patient underwent high orchidectomy and specimen was sending for histopathological examination.

Gross examination revealed a markedly enlarged orchidectomy specimen measuring 14x9x8.5cms with attached spermatic cord measuring 6cms in length. Cut sections showed a small testis measuring 4x2x1.5cms pushed and compressed to one side by a large Para testicular growth measuring 13x8.5x 8cms.Cut section through the growth showed pale creamy yellow fleshy lobulated mass with large areas of necrosis and focal myxoid degeneration (Fig 1). Cut section through the testis appeared unremarkable (Fig 1 inset).

Microscopic examinations from the growth show a malignant spindle cell neoplasm having alternate cellular and myxoid areas composed of round to spindle shaped rhabdomyoblasts (Fig 2). Individual rhabdomyoblasts display mild to moderately pleomorphic hyperchromatic nuclei and scant to moderate eosinophilic cytoplasm (Fig 3a). Few of the rhabdomyoblasts have cytoplasmic tailing with occasional cross striations (Fig 3b). There are focal areas of necrosis and myxoid degeneration seen. There are increased mitotic figures (30 – 40/10 hpf) noted. Sections from the testicular parenchyma showed atrophic changes. Sections from the epididymis appeared histologically unremarkable. Section from the surgical end of spermatic cord was free of tumor deposits. Immunohistochemical examination showed strong positivity in tumor cells for Vimentin (Fig 3c), Desmin and Myogenin (Fig 3d). Tumor showed negative expression for Smooth muscle actin and S100. The proliferative index (Ki 67) was 40 - 50%.

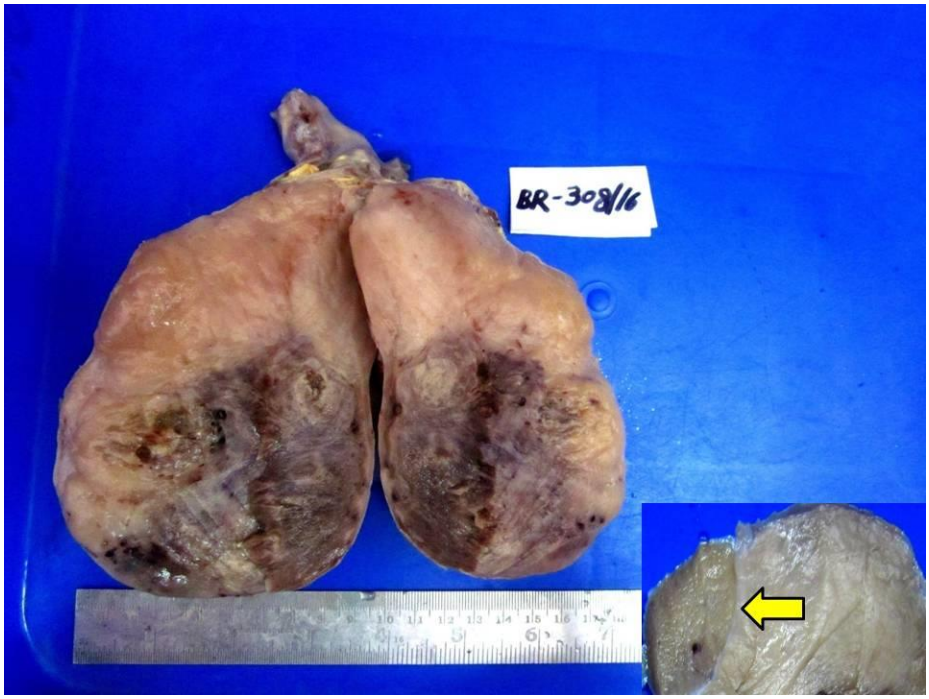

Fig 1: Gross picture of Para testicular tumour. Cut surface shows lobulated mass with focal myxoid degeneration and necrosis; Inset shows atrophic testis (yellow arrow).

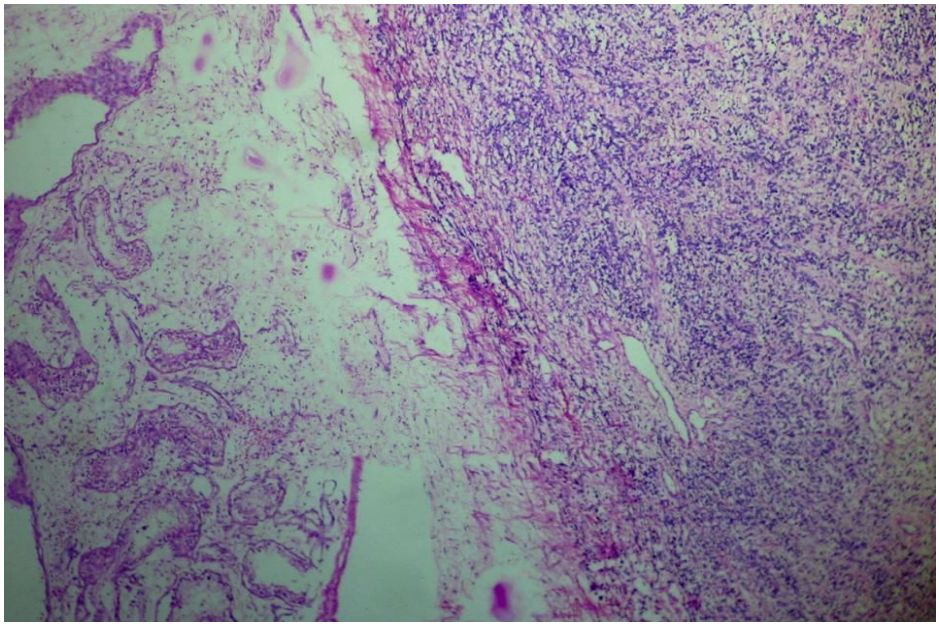

Fig 2: Spindled cell embryonal RMS and atrophic testicular parenchyma (H&E, x10).

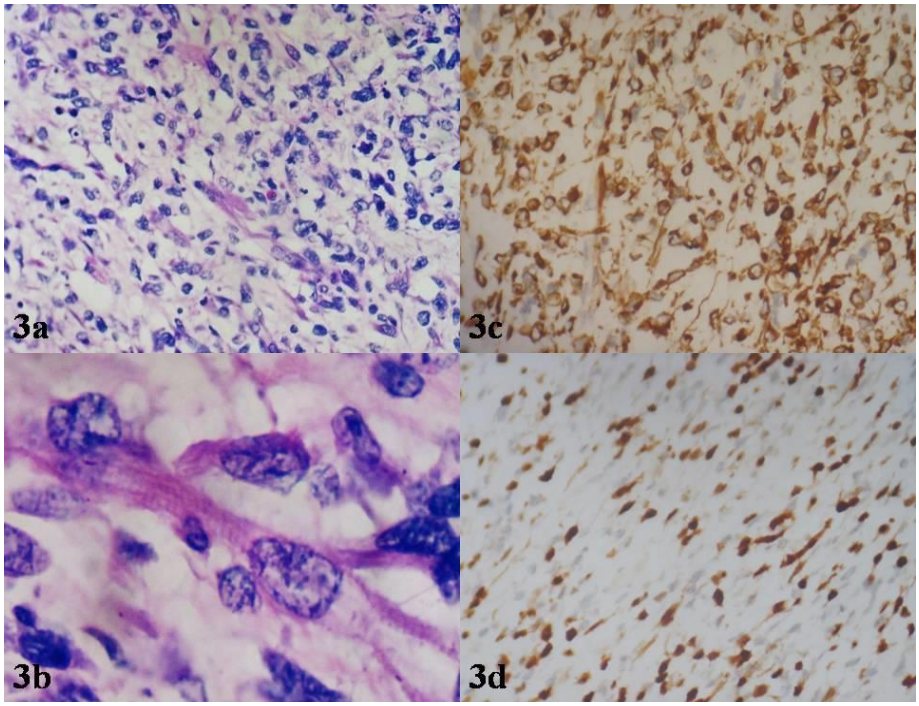

Fig 3: (a) Rhabdomyoblasts with focal cytoplasmic tailing (H&E, x10) (b) Rhabdomyoblast with cytoplasmic tailing and cross striations (H&E, x100), (c) Vimentin positivity in tumour cells (Vimentin, x10), (d) Myogenin positivity in tumour cells (Myogenin, x10).

Final histopathological diagnosis was embryonal RMS, spindle cell variant in Para testicular area. The TNM staging was T1N0M0. The patient was followed up post operatively with chemotherapy.

DISCUSSION:

The Para testicular region comprises of contents of spermatic cord, testicular tunics, epididymis and vestigial remnants. Histologically, tumors may be epithelial, mesenchymal or mesothelial origin. Rarely secondary tumor deposits can be seen in the Para testicular region [4]. Most tumors in the Para testicular region are of mesenchymal origin. Soft tissue sarcomas are the most common malignancies in this region. RMS is the commonest adult soft tissue sarcoma in the Para testicular region accounting for approximately 24%, of which approximately 80% occur before 21 years of age [2].

Para testicular RMS present clinically as a unilateral, painless scrotal mass with short clinical duration as seen in our patient. Most common mode of tumor spread is via lymphatics to iliac and para-aortic lymph nodes. Distant metastasis to liver, lungs and bone by hematogenous spread account for 20% of patients in initial presentation [3].

The Para testicular RMS can be further classified into embryonal, alveolar, pleomorphic and mixed subtypes. Embryonal RMS is the commonest subtype, accounting for 90% of all RMS in the Para testicular region with a peak incidence between 1 to 5 years of age [5]. Spindle cell variant of embryonal RMS (Spindle cell RMS) is a minor subset with excellent prognosis [6]. According to the Intergroup Rhabdomyosarcoma study (IRS), there was 30.6 % incidence of spindle cell variant of embryonal RMS in the Para testicular area out of 800 randomly selected patients in IRS group II [7].

Spindle cell variant of RMS can cause diagnostic challenge with leiomyosarcoma being a close differential diagnosis. Demonstration of cytoplasmic cross striations or rhabdomyoblasts on routine microscopy supported by positive immunohistochemical expression of Myogenin and Myo D1 (Myogenic nuclear transcription factors) confirms the skeletal muscle differentiation [8].

Management of embryonal RMS has shifted with multispecialty approach improving the survival chances upto 80% by incorporating chemotherapeutic drugs in addition to surgical intervention or radiation therapy. Radical orchidectomy with high ligation of spermatic cord is the standard surgical intervention. The site of tumor origin, lymphnode metastasis, distant metastasis, presence or absence of residual disease and surgical resection status will help in staging of RMS patients.

CONCLUSION:

Spindle cell variant of embryonal RMS is a rare tumor in the Para testicular region as was seen in our patient with excellent prognosis. Scrotal and abdominal ultrasound with serum tumor markers should be done as part of routine evaluation of rapidly

growing, painless scrotal mass to detect rare Para testicular neoplasms like embryonal RMS for timely multidisciplinary management resulting in better clinical outcome and prognosis.

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