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Case Report

Testicular Mass in Children: A Case Report

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ABSTRACT

Testicular sarcomas constitute only 1–2% of all testicular tumors and are mostly associated with germ cell tumor. Primary intratesticular localization is very rare and must be differentiated from paratesticular locations. Accurate diagnosis and early treatment is essential as it is an aggressive tumor with high metastatic potential and poor prognosis.The optimal treatment is based on radical inguinal orchidectomy and chemotherapy.

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INTRODUCTION

Rhabdomyosarcoma is the most common soft-tissue tumor in children.[1] It occurs predominantly in three regions: the head and neck, genitourinary tract and extremities. The majority of cases of rhabdomyosarcoma in the genitourinary tract occurs in the bladder and paratesticular organs.[1] Primary intratesticular rhabdomyosarcoma is very rare.[2,3] Here, we report a case of a ten-year-old child with primary pure intratesticular rhabdomyosarcoma.

CASE REPORT

A 10 year-male presented in our Out Patient Department with left testicular mass since 6 months, insidious in onset, gradually progressive, non painful for initial 3 then became mildly painful at touch. The patient did not have any medico-surgical or trauma history. He had also no personal or family history of cancer. Physical examination revealed there was a mass in the left scrotal region measuring 12×12 cm in size, firm to hard in consistency, irregular in shape, mildly tender on palpation, mobile and not fixed to the scrotal skin (fig. 1). Left testis was not palpable separately. Other systemic examination was normal. Baseline blood reports, serum alpha-fetoprotein (1.85 ng/ml), beta-human chorionic gonadotropin (0.300mIU/L) and lactate dehydrogenase (297 U/L) were within normal limits. Scrotal and abdominal ultrasound examination showed a large hypoechoic intratesticular mass measuring 12*12cm in the left scrotum. Right testis appeared normal. The liver, biliary system, pancreas, kidney, and spleen were normal. Retroperitoneal lymph nodes were not enlarged. CECT neck, chest and abdomen showed 12*12cm heterogeneously enhancing mass in left testis, few left para-aortic lymphnodes approx 11*7mm, few mesenteric nodes measuring 10*7 mm (fig. 2).

Figure 1: Gross picture of the left scrotal mass.

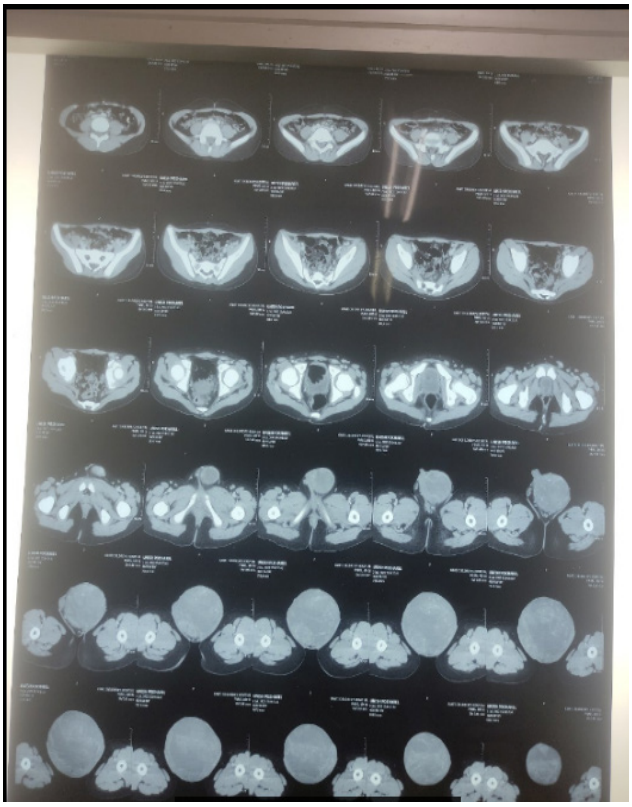

Figure2: CECT chest and pelvis showing at the pelvic level left scrotal mass.

The patient underwent 3 cycles of chemotherapy (Etoposide, Carboplatin and bleomycin) within 3 months. Post chemo serum alpha-fetoprotein (0.5 1IU/ml), beta-human chorionic gonadotropin (0.300 mIU/l) were within normal limits. Patient underwent left radical orchidectomy (fig. 3). Retroperitoneal lymph node dissection (RPLND) was not performed in our case as radiologically there was no lymphadenopathy. Grossly, the testis was enlarged and measured 12*12cm. The tumor was completely contained within the tunica albuginea. The cut surface was whitish and firm in consistency. The testis was almost completely replaced by tumor (fig. 4).

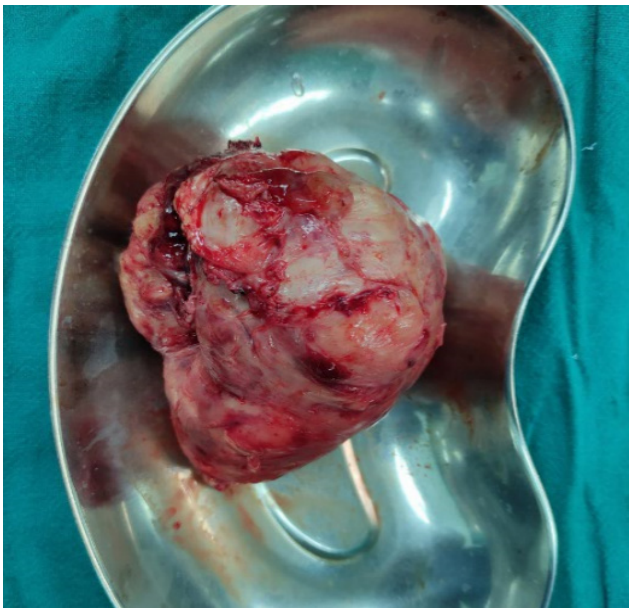

Figure 3: Intra operative and gross specimen picture showing huge left scrotal mass.

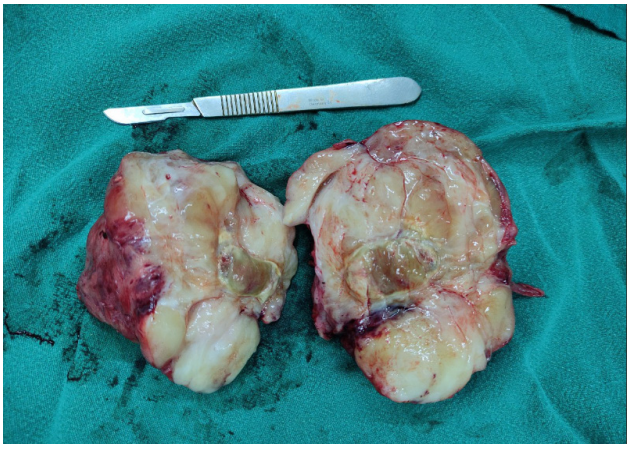

Figure4: Cut section of the resected specimen.

His post operative days were uneventful. Histopathological examination revealed hypocellular and hypercellular areas with proliferation of moderately pleomorphic atypical spindle shaped cells having scant cytoplasm, oval to elongated nuclei with coarse chromatin and inconspicuous nucleoli. Occasional bizarre multinucleated tumor cells are also identified. Mitotic figure constitutes 3-4/HPF in mitotically active cellular areas, adjacent areas show necrosis. Tumor is limited to testis with unremarkable cord resection margin. Immunohistochemistry showed positive results for Desmin, Myogenin, SMA, CD117, negative for CK and CD30. Diagnosis of primary intratesticular rhabdomyosarcoma was made. Patient is planned for post operative chemotherapy.

DISCUSSION

Rhabdomyosarcoma is the most common soft tissue sarcoma in children and adolescents, with 4.5 cases per one million people aged 0–20 years.[4] It can occur in all organs, but preferentially affects the head and neck (44 %), genitourinary tract (34 %) and extremities (14 %). Unlike paratesticular rhabdomyosarcoma,

primary intra-testicular localization is very rare and only 22 cases have been reported in the literature till today.[5-8] The origin of intratesticular rhabdomyosarcoma is uncertain. However, two theories link their development to undifferentiated mesenchyme having the capacity for rhabdomyoblastic differentiation, or to embryonal muscle tissue that has been displaced during the early stages of development. Trauma, teratoma of the testes, exogenous maternal estrogen and cryptorchidism have been reported as predisposing factors.[9]

Clinically primary intratesticular rhabdomyosarcoma presents usually as a short history of a progressively increasing painless testicular mass, generally in children and young adults, with no preference for any particular race. The painful mass has been reported in a few cases. Tumor is usually unilateral with no predilection aside.[8] Laboratory studies including a-fetoprotein (AFP), b-human chorionic gonadotropin (b-HCG) and lactate dehydrogenase (LDH) revealed nonspecific results, they are generally negative, unlike germ cell tumors. Scrotal ultrasound is useful for the diagnosis of the testicular mass, and for examining adjacent organs. Thoracicabdominopelvic computed tomography (CT) scan is usually used for detecting lymph nodes and distant metastases.[6] Anatomopathological examination is the gold standard for RMS diagnosis and for confirming the intratesticular origin of the tumor.

The main differential diagnoses of intratesticular rhabdomyosarcoma are paratesticular rhabdomyosarcoma which can be eliminated by radiological investigations and gross appearance. Other intratesticular spindle cell sarcomas also represent a differential diagnosis. It is also important to differentiate RMS from germ cell tumors with sarcomatous components by a meticulous examination of specimen, and immunohistochemical study.[10]

It was reported that the optimal treatment for intra-testicular RMS is radical inguinal orchiectomy followed by chemotherapy. The recommended chemotherapy agents are vincristine, actinomycin-D and cyclophosphamide (VAC).[5] Retroperitoneal lymphnode dissection for accurate staging or initial treatment is a source of controversy, especially in the absence of positive lymph nodes in radiological investigation. However, it has been reported that retroperitoneal lymphnode dissection has a role in debulking disease if nodes persist after Chemotherapy.[6]

CONCLUSIONS

Primary intratesticular rhabdomyosarcoma is a rare malignant tumor occurring in children and young adults. Radical inguinal orchiectomy followed by chemotherapy is the treatment of choice. Early diagnosis and treatment improve the prognosis.

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