Primary intratesticular rhabdomyosarcoma in pediatrics

Jitendra G. Nasit, Biren Parikh[1], Priti Trivedi[1], Manoj Shah[1]

Department of Pathology, P.D.U. Medical College, Rajkot,[1] Gujarat Cancer and Research Institute, Ahmadabad, Gujarat, India

ABSTRACT

Testicular sarcomas constitute only 1–2% of all testicular tumors and are mostly associated with germ cell tumor. Primary intratesticular rhabdomyosarcoma is rare and only 14 cases have been reported in the literature till date. It should be differentiated from germ cell tumor with sarcomatous component, other intratesticular spindle-cell sarcomas and paratesticular rhabdomyosarcoma. Accurate diagnosis and early treatment is essential as it is an aggressive tumor with high metastatic potential and poor prognosis. Orchidectomy is the treatment of choice. Chemo-radiotherapy is recommended in case of recurrence and metastasis.

Key words: Intratesticular rhabdomyosarcoma, orchidectomy, prognosis

INTRODUCTION

Rhabdomyosarcoma is the most common soft-tissue tumor in children.[1,2] 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.[3,4] The etiology is uncertain, however, the origin is presumed to be an overgrowth of the sarcomatous component of teratoma.[1,4] Here, we report a case of a two-year-old child with primary pure intratesticular rhabdomyosarcoma.

CASE REPORT

A two-year old child presented with a slow-growing painless right testicular mass since two months. Physical examination revealed a firm, non-tender mass in the right scrotum. Complete blood count, serum alpha-fetoprotein (2.0 ng/ml), beta-human chorionic gonadotropin (5.0 IU/L) and lactate dehydrogenase (120.0 IU/L) were within normal limits. Scrotal and abdominal ultrasound examination showed a large hypoechoic intratesticular mass measuring 6.8 × 4.5 × 3.5 cm in the right scrotum. Neither hemorrhage nor necrosis was identified within the mass. The left testis appeared normal. The liver, biliary system, pancreas, kidney, and spleen were normal. Retroperitoneal lymph nodes were not enlarged. Ultrasound findings were suggestive of testicular malignancy. Subsequently, after two days right testicular biopsy was performed. It was reported as high-grade spindle cell tumor; possibility of sarcomatous elements of malignant germ cell tumor should be ruled out.

The patient underwent high inguinal orchidectomy after one week. Grossly, the testis was enlarged and measured 6.4 × 4.0 × 3.0 cm. 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 with a small portion of residual normal testis [Figure 1]. Histopathological examination revealed a highly cellular tumor composed of pleomorphic cells including round to spindle, strap-like and racquet-shaped cells arranged in sheets. Few large cells having abundant granular eosinophilic cytoplasm with cross-striations, eccentrically situated large vesicular nucleus and prominent nuclei was evident. Mitotic figures were frequently observed [Figures 2a-c]. Numerous sections were studied in an attempt to find other elements of germ cell tumor, but none could be found. Immunohistochemistry

For correspondence: Dr. Jitendra G. Nasit, C/4, Suryadeep Society, Near Nutan School, B/H Chankyapuri Society, New Sama Road, Vadodara – 390 008, Gujarat, India.
E-mail: eagleeyenasit@gmail.com or jitupathologist@gmail.com

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showed positive results for vimentin, desmin and myogenin [Figure 2d, e] while negative for alpha-smooth muscle actin, placental alkaline phosphatase (PLAP), CD-117, S-100, cytokeratin, and epithelial membrane antigen. Diagnosis of primary intratesticular rhabdomyosarcoma was made. The postoperative course was uneventful.

Orchidectomy was followed by chemotherapy. The chemotherapy regimen included vincristine, actinomycin D and cyclophosphamide. Retroperitoneal lymph node dissection (RPLND) was not performed in our case as radiologically there was no lymphadenopathy. In spite of chemotherapy, patient developed multiple metastases to lung, pelvic and periaortic lymph nodes after 16 months and died after 20 months of diagnosis.

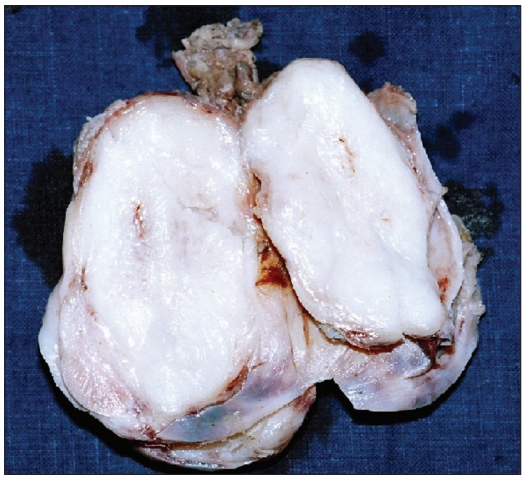

Figure 1: Testis was enlarged and almost completely replaced by tumor with whitish firm cut surface. Small portion of residual normal testis was also seen

DISCUSSION

Testicular sarcomas constitute only 1–2% of all testicular tumors.[5] Intrascrotal sarcomas are traditionally separated into paratesticular and intratesticular tumors.[6] Paratesticular sarcomas are mostly rhabdomyosarcoma, seen in children.[7] However, intratesticular sarcomas are very rare and mostly associated with a germ cell tumor.[6,8] Primary intratesticular rhabdomyosarcoma is very rare, with just 14 cases reported in the literature till date.[1,3,4]

The origin of intratesticular rhabdomyosarcoma is unclear. It is believed to be teratomatous with rhabdomyoblastic overgrowth of primitive germ cells.[4,9] Other hypotheses include undifferentiated mesenchyme having the capacity for rhabdomyoblastic differentiation, or perhaps from embryonal muscle tissue that has been displaced during the early stages of tissue development.[9] Trauma, cryptorchidism, and exogenous maternal estrogen (*in utero*) have all been associated with its development.[4]

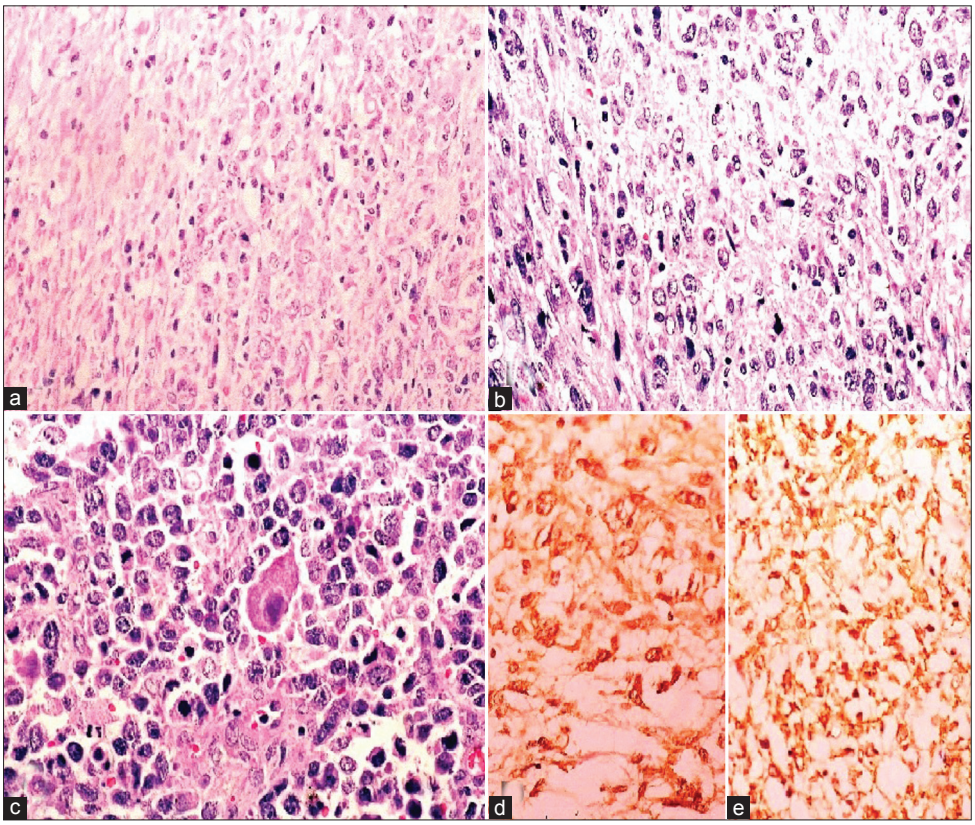

Figure 2: Primary intratesticular rhabdomyosarcoma: H and E stain: (a) and (b) Highly cellular tumor composed of pleomorphic cells including round to spindle, strap-like cells. Mitotic figures were seen (×100), (c) Racquet-shaped rhabdomyoblast showed abundant granular eosinophilic cytoplasm, eccentrically situated large vesicular nucleus with prominent nucleoli (×400). Immunohistochemistry showed desmin (d) and myogenin (e) positivity in tumor cells (×100)

Mean age of primary intratesticular sarcoma at presentation is 32 years (range 3–86 years).[9] In our case, the age of the patient was only two years. It presents as a slowly-growing, painless, firm to hard, testicular mass.[1] Average tumor size is 6.8 cm (range 0.2–15 cm).[9] Rhabdomyosarcoma is regarded as a highly malignant tumor with frequent recurrence and dissemination via the bloodstream and lymphatics.[1,4] Metastasis of rhabdomyosarcoma is common to the lung, bone marrow, brain, liver, omentum and lymph nodes.[7]

Primary intratesticular rhabdomyosarcoma should be differentiated from germ cell tumor with sarcomatous component, other intratesticular spindle cell sarcomas and paratesticular rhabdomyosarcoma. Diagnosis of primary intratesticular rhabdomyosarcoma should be made only after meticulous examination of specimen to exclude any germ cell component as striped muscle elements are frequent findings in testicular teratoma.[8] The accurate diagnosis of the sarcomatous component in germ cell tumor is very important because of the implications for therapy. Immunohistochemistry is helpful to exclude other intratesticular spindle cell sarcomas like fibrosarcoma and leiomyosarcoma.[10,11] Leiomyosarcoma shows strong immune reactivity for muscle-specific actin, smooth muscle actin, vimentin, calponin and alpha-l-chymotrypsin and negative reaction for desmin, cytokeratin, S-100 protein and Leu 7.[10,12,13] Fibrosarcoma shows positive result for vimentin, Type I collagen and negative result for smooth muscle marker and histiocytic markers.[11] A diagnosis of primary intratesticular sarcoma should only be made after exclusion of the more commonly seen paratesticular sarcomas.[10] Intratesticular rhabdomyosarcomas had better prognosis than paratesticular rhabdomyosarcomas.[9] Scrotal ultrasound and abdominal computed tomography imaging were used to exclude paratesticular and retroperitoneal pathology.[10] The tumor epicenter was found within the testis and no paratesticular pathology was seen in our case. Gross examination of the resected specimen showed tumor mass arising from the testis primarily, and tumor was not adherent to the scrotal wall or adjacent tissues.[13] However, to determine the specific site of origin of paratesticular and testicular sarcoma is not always possible because these structures are in continuity with each other.[8,13]

Multidisciplinary treatment approaches have greatly improved the prognosis of intratesticular rhabdomyosarcoma.[1,9] The Intergroup Rhabdomyosarcoma Study (IRS) Group management guidelines for the pediatric population have resulted in reducing morbidity and increasing survival from 25–70% over 20 years since 1970.[2,14] Orchidectomy followed by chemotherapy is the best treatment.[1,2] Post-orchidectomy RPLND used to be carried out as a routine staging procedure, more commonly for a therapeutic purpose.[1] RPLND combined with chemotherapy revealed excellent long-term results.[10] The chemotherapy agents used are vincristine, actinomycin D and cyclophosphamide. Patients with unresectable tumors who undergo treatment with chemotherapy should be considered for surgery after downgrading.[2] Radiotherapy is recommended more commonly to control local recurrence, metastasis or for unfavorable histology, such as alveolar rhabdomyosarcoma.[1,2] Recurrence may occur within two years so intensive surveillance and follow-up is necessary.[4] The prognosis is generally considered to be poor.[3,9]

CONCLUSION

Primary intratesticular sarcoma that is neither associated with germ cell elements nor paratesticular elements is a unique subset of intrascrotal sarcoma. Germ cell tumor component and paratesticular sarcoma must be excluded carefully before establishing the diagnosis. Primary intratesticular rhabdomyosarcoma is very rare, with an aggressive growth pattern. Radical orchidectomy with negative surgical margins followed by RPLND and chemotherapy is the recommended treatment. Radiotherapy is recommended for the control of local recurrence and metastasis.

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