A case of children with primary intratesticular rhabdomyosarcoma

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A B S T R A C T

In children, the most typical soft tissue tumor is a rhabdomyosarcoma (RMS). A primary intratesticular rhabdomyosarcoma is quite uncommon. An eleven-year-old had a painless left testicular tumor for two months. The patient’s left scrotum was found to have a firm, non-tender lump. A massive hypoechoic intratesticular mass measuring 8.0 × 7.5 × 6.0 cm was found in the left scrotum by scrotal and abdominal ultrasonography. The patient had an inguinal orchiectomy. Histopathological examination disclosed botryoid rhabdomyosarcoma. Primary intratesticular rhabdomyosarcomas are rare. Radical orchiectomy and chemotherapy are recommended. It may increase overall and disease-free survival.

1. Introduction

About 3% of children cancers and 1% of adult cancers are soft tissue sarcomas. The most frequent soft tissue tumor in children, a rhabdomyosarcoma (RMS), arises from mesenchymal cells. Rhabdomyosarcoma commonly occurs in the bladder and paratesticular organs. 1 The World Health Organization (WHO) study on categorization of RMS describes four histological subtypes of RMS: alveolar, embryonal, pleomorphic, and sclerosing or spindle cell. 2 Primary intratesticular rhabdomyosarcoma is very rare. 3 The etiology is unknown at this time, however it might be caused by an enlargement of the teratoma’s sarcomatous component. Here, we describe a case of primary intratesticular rhabdomyosarcoma in an eleven-year-old boy.

2. Case description

The eleven-year-old arrived with a two-month history of painless left testicular tumor. Left scrotum examination revealed a non-tender tumor. Complete blood counts were within the range of normal values. Due to our limitations serum AFP, β-HCG and DHL were not measured. A massive hypoechoic intratesticular mass measuring 8.0 × 7.5 × 6.0 cm was discovered during scrotal and abdominal ultrasonography evaluation of the left scrotum. There was no evidence of bleeding or necrosis inside the tumor. The right testis seemed healthy. Liver, biliary system, pancreas, kidney, and spleen were normal. The lymph nodes in the retroperitoneum were not swollen. The ultrasound results were indicative of testicular cancer.

After one week, the patient had a high inguinal orchiectomy. The testis was grossly swollen and measured 9 × 8 × 7 cm (Fig. 1A). Tunica albuginea totally covered the tumor. The cut edge was white and inconsistently hard (Fig. 1B). A minor fraction of normal testis remained after the tumor’s replacement of normal testis. A high grade cellular tumor constituted of pleomorphic cells comprising round to spindle, hyperplastic oval, and mostly compacted cells grouped in fasciculus was shown by histopathological investigation (Fig. 1C). Few big cells have a substantial amount of granular eosinophilic cytoplasm. Figures of mitosis were noticed (Fig. 1D).

Orchiectomy was followed by chemotherapy. Patient underwent chemotherapy with Bleomycin-Etoposide-Cisplatin after surgery. It was decided not to do a retroperitoneal lymph node dissection (RPLND) since there was no radiological evidence of lymphadenopathy. The patient was monitored following surgery and was still doing well as of the time this article was published.

3. Discussion

The percentage of testicular tumors that are testicular sarcomas is about 1%–2%. Paratesticular and intratesticular tumors are the two categories by which intrascrotal sarcomas are often classified. 3 Most pediatric cases of paratesticular sarcoma are rhabdomyosarcomas. Intratesticular sarcomas are uncommon and usually caused by germ cell tumors. 7 Intratesticular rhabdomyosarcoma is uncommon. 2

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Intratesticular rhabdomyosarcoma etiology is still indecisive. Possibly teratomatous with primitive germ cell rhabdomyoblastic growth. Other hypotheses include undifferentiated mesenchyme capable of differentiating into rhabdomyoblasts or embryonal muscle tissue displaced during the early stages of tissue development. Trauma, cryptorchidism, and exogenous maternal estrogen have all been linked to its development.

Intrascrotal rhabdomyosarcoma appears clinically as a painless lump that may grow for a few weeks before becoming uncomfortable, and a few instances have been observed to present with discomfort. Trauma is estimated to be a contributing factor in just 7% of cases and is sometimes related solely to draw attention to the tumor. Patients develop a slow-growing intrascrotal mass for the first six months before diagnosis with tumor size averaged 6.8cm (range 0.2–15cm) and all were completely contained within the tunica vaginalis. Sometimes, serum levels of AFP, β-HCG and DHL are normal. Serial serologic evaluation with β-HCG and AFP are needed to exclude serologic relapse from undetected germ cell tumor metastases.

Different methods are used to diagnose intrascrotal rhabdomyosarcoma. Ultrasound, Computed Tomography (CT), Magnetic Resonance Imaging (MRI), IHC staining, gross appearance, and histology are needed to determine whether the scrotal mass has an intratesticular or paratesticular origin, scrotal ultrasound and an abdominal CT scan may be used. Ultrasound is the most used imaging technique for evaluating testicular malignancy and adjacent organs. Germ cell tumors that have rhabdomyoblastic differentiation and other intratesticular spindle cell sarcomas may only be eliminated with the use of immunohistochemical markers.

Embryonal rhabdomyosarcoma, which typically develops in the vagina, urinary bladder, or extra hepatic bile ducts, was previously known as botryoid. Now, with the use of immunohistochemistry in diagnosis, the term has changed. Botryoid lesions are a better prognosis variation of embryonal rhabdomyosarcoma. Histologically, various kinds may be distinguished, including pleomorphic, botryoidal (an embryonal variation), alveolar (with its solid variety), and subtypes like fusocellular and sclerosing pseudovascular. The most aggressive variety, alveolar rhabdomyosarcoma, progresses quickly and commonly develops early metastatic lesions, increasing mortality as compared to embryonal rhabdomyosarcoma.

All patients must have a radical orchiectomy to ensure that the main tumor has been completely removed. Because there is little information on the management of RMS in adults, the IRS provides management advice (Intergroup Rhabdomyosarcoma Study). The IRS management

Table 1

| Stage | Site of primary tumor | Tumor size | Lymph nodes | Distant metastases |
|-------|-----------------------|------------|-------------|--------------------|
| 1     | Orbit, non-PM head/neck; GU nonbladder/prostate; biliary tract | Any size   | N0, N1      | M0                 |
| 2     | All other sites       | <5cm       | N0          | M0                 |
| 3     | All other sites       | <5cm <5cm | N1 or N1    | M0                 |
| 4     | Any site              | Any size   | N0 or N1    | M1                 |

PM: parameningeal; GU: genitourinary; N0: no nodal involvement; N1: regional lymph node involvement; M0: no metastasis; M1: distant metastasis. According to Raney et al.

Fig. 1. (A) Enlarged Testicle. (B) White firm-edged tumor. A little amount of normal testis remained. (C) Testicular malignancy with predominant spindle cell characteristics (100 × ). (D) Rhabdomyoblasts with big nuclei, prominent nucleoli, and glossy cytoplasm with crisscrossed grooves.
recommendations are designed with a pediatric population in mind (Table 1).

The prognosis for intratesticular rhabdomyosarcoma has significantly improved with the use of multidisciplinary treatment methods. RPLND was not advised since IRS group 1 tumors are not frequently used in Europe. The IRS grouping system is shown in Table 2, which divides patients into groups based on the extent of tumor still present following initial surgery but prior to starting chemotherapy and radiation therapy. Chemotherapy supplements surgery such as Bleomycin, etoposide, and cisplatin are utilized. Downgrading unresectable tumors with chemotherapy should lead to surgery. Metastasis sufferers need radiation. Patients with IRS group 1 have not demonstrated any benefit from radiotherapy.

4. Conclusion

Primary intratesticular rhabdomyosarcomas are uncommon, with just 23 recorded cases globally. Radical orchiectomy followed by chemotherapy is the recommended course of treatment. It may increase the rates of disease-free survival and overall survival. Prognosis improves with early diagnosis and treatment.

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Ethical approval and consent

The patient’s written informed consent was acquired before to publication and the use of any associated photos. It was sent to the journal’s chief editor. Due to the nature of case reports, our institution decided not to need ethical clearance.

Author contributions

Ghani Ikhsan Majid created the concept, planned the research, gathered patient data, and wrote the draft. Alyadi Rachmadiyan conceived the idea, collected the patient data. Irvan Octavian and Muhammad Ilhamul Karim validated the diagnosis, conducted the operation, reviewed and directed the case treatment. Friska Mardianty conducted the histological examination. Irvan Octavian, Muhammad Ilhamul Karim, and Friska Mardianty evaluated and corrected the final draft of the manuscript. Irvan Octavian was the project’s supervisor. All authors approved the final manuscript.

Registration of research studies

N/a.

Table 2

| Group | Definition |
|-------|------------|
| 1     | Localized tumor, completely removed with pathologically clear margins and no regional lymph node involvement. |
| 2     | Localized tumor, grossly removed with (a) microscopically involved margins Localized tumor, grossly removed with (b) involved, grossly resected regional lymph nodes Localized tumor, grossly removed with (c) both. |
| 3     | Localized tumor, with gross residual disease after grossly incomplete removal, or biopsy only. |
| 4     | Distant metastases present at diagnosis |

Based on Raney RB, Maurer HM, Anderson JR et al. The Intergroup Rhabdomyosarcoma Study Group (IRSG): Major Lessons From the IRS-I Through IRS-IV Studies as Background for the Current IRS-V Treatment Protocols. Sarcoma. 2001; 5(1):9–15. https://doi.org/10.1080/13577140120048890.

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