Case report

Primary intra-testicular rhabdomyosarcoma: Case report

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ABSTRACT

Introduction: RMS is a highly aggressive neoplasm that represents the most common soft tissue sarcoma in children and adolescents. It occurs most frequently in the head, neck, and genitourinary tract. Intra-testicular localization remains exceptional and very few cases have been reported in the literature.

Case presentation: We represent a case of a 16-year-old patient, with no medical and surgical history, who presented with a painless right scrotal mass. Imaging revealed a suspicious testicular mass with no other secondary location. The patient underwent a right inguinal orchidectomy and the anatomopathological study confirmed the diagnosis of intratesticular rhabdomyosarcoma. The patient subsequently was referred to the oncology department for chemotherapy (Agha et al., 2020).

Discussion: Intra-testicular rhabdomyosarcoma is a very rare malignant mesenchymal tumor showing skeletal muscle differentiation. They are subclassified into four major histological subtypes: Embryonal, alveolar, pleomorphic and Spindle cell/sclerosing rhabdomyosarcoma. ERMS is the most common subtype. Diagnosis of ITRMS is based on anatomopathological examination. Surgery and chemotherapy remain the mainstay of treatment. Radiotherapy is useful for local recurrence and metastasis.

Conclusion: In this report we aim to describe the clinical, pathological and immunohistochemical features of this rare entity, and to highlight the importance of an early diagnosis followed by treatment combining Radical inguinal orchietomy and chemotherapy and its role in improving prognosis.

1. Introduction

Rhabdomyosarcoma is a malignant mesenchymal tumor showing skeletal muscle differentiation. Primary intratesticular localization is very rare and must be differentiated from paratesticular locations. The Immunohistochemical study is essential. The optimal treatment is based on radical inguinal orchietomy and adjuvant chemotherapy. This case has been reported in line with SCARE criteria [11].

2. Case report

We report the case of a 16 years old patient who presented to our structure for painless scrotal mass that has been gradually increasing in size over 4 months. The patient did not have any medico-surgical or trauma history. He had also no personal or family history of cancer. Physical examination revealed a renitent and suspicious mass. The left testis was normal. Scrotal ultrasound revealed a heterogeneous, intra-testicular mass measuring 93 × 81 mm. Blood investigations including α-fetoprotein (AFP), β-human chorionic gonadotropin (β-HCG), and lactate dehydrogenase (LDH) were normal.

A thoracic-abdominopelvic CT scan showed at the pelvic level a 94 × 81 × 70 mm mass within the right testis. No lesions were identified in the left testicle. There was also no evidence of pulmonary, abdominal, or lymph-node lesions (Fig. 1).

In front of these radiological aspects that suggested testicular malignancy, the patient underwent a right inguinal orchidectomy. He had no postoperative complications and was discharged one week later.

The macroscopic examination of the specimen showed tumor mass arising from the body of the testis primarily without infiltrating the tunica albuginea. The cut surface shows a pale-tan and firm mass with areas of necrosis (Fig. 2).

Histopathologic examination revealed a mesenchymal profilation
(proliferation) with alternating areas of loose and dense cellularity. It is composed of sheets of spindled, and round cells with scant eosinophilic cytoplasm and small oval nuclei with inconspicuous nucleoli. We note focally scattered differentiated rhabdomyoblasts.

Immunohistochemically, tumor cells showed positive staining for vimentin, desmin and myogenin, and negative staining for CD34, OCT ¾, SALL 4 and MDM2 (Fig. 3).

These pathology results were consistent with the diagnosis of intratesticular embryonal rhabdomyosarcoma grade III of FNCLCC. The tumor was staged as pT1 N0 M0 (AJCC 7th edition staging).

3 weeks later, the patient was referred to the oncology department where he received chemotherapy consisting of vincristine, actinomycin D and cyclophosphamide. One year later, the patient is still stable with no recurrence or metastasis.

3. Discussion

RMS is the most common soft tissue sarcoma in children and adolescents, with 4.5 cases per one million people aged 0–20 years [1]. It can occur in all organs, but preferentially affects the head and neck (44 %), genitourinary tract (34 %) and extremities (14 %). Unlike para-testicular rhabdomyosarcoma, primary intra-testicular localization is very rare and only 22 cases have been reported in the literature till today [2–5].

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 [6].

 Clinically PI RMS 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 [5]. 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 tumours. Scrotal ultrasound is useful for the diagnosis of the testicular mass, and for examining adjacent organs. Thoracic-abdominopelvic computed tomography (CT) scan is usually used for detecting lymph nodes and distant metastases [3]. Anatomopathological examination is the gold standard for RMS diagnosis and for confirming the intratesticular origin of the tumor while macroscopic examination. Histologically, there are four histological types [9]:

Fig. 1. A thoracic-abdominopelvic CT scan showing at the pelvic level a right scrotal mass.

Fig. 2. Cut section of the resected specimen.
Embryonal RMS is the most common subtype, it is composed of round and spindle cells with scattered differentiated rhabdomyoblasts proliferating in patternless sheets with altering areas of loose and dense cellularity. Immunohistochemically, cells are positive for desmin, myogenin and MY0D1. These stains are less diffuse than in alveolar rhabdomyosarcoma.

Alveolar RMS: this is a neoplasm with a poor prognosis. It presents histologically as a dense and monomorphic proliferation composed of uniform round cells with features of arrested myogenesis. Neoplastic cells are generally in nests separated by fibrovascular septa. Immunohistochemically, tumor cells show a strong homogeneous nuclear stain for myogenin and heterogeneous staining for desmin and MY0D1.

Spindle Cell/Sclerosing Rhabdomyosarcoma is a rare subtype. It is characterized by fascicles of spindle cells with an intersecting growth pattern.

Pleomorphic RMS is composed of sheets of large and pleomorphic cells with eosinophilic cytoplasm.

The main differential diagnoses of ITRMS 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 tumours with sarcomatous components by a meticulous examination of specimen, and immunohistochemical study [8].

It was reported that the optimal treatment for ITRMS is radical inguinal orchietomy followed by chemotherapy. The recommended chemotherapy agents are vincristine, actinomycin-D and cyclophosphamide (VAC) [2].

RPLND 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 RPLND has a role in debulking disease if nodes persist after Chemotherapy [3]. Radiotherapy can be used in case of metastasis, local recurrence, macroscopic or microscopic postoperative tumor residue, unresectable tumor, and unfavorable histological types including alveolar RMS [9]. ITRMS is a highly malignant tumor with a poor diagnosis with the potential for distant metastases [3]. Recurrences can occur 2 years after initial diagnosis hence the interest in patient follow-up [8]. Survival after recurrence is poor [10]. Tumor size, age, histological type, and lymph node involvement are important risk factors [2]. Children have the best outcome than adolescents and young adults [1]. ERMS is a favorable prognostic factor [7].

4. Conclusion

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

Abbreviations

| Abbreviation | Description |
|--------------|-------------|
| ITRMS        | intratesticular rhabdomyosarcoma |
| ERMS         | embryonal rhabdomyosarcoma |
| CD           | cluster designation or cluster of differentiation; OCT: octamer-binding transcription factor |
| SALL4        | Spalt-like transcription factor 4 |
| MDM2         | murine double minute 2 |
| RPLND        | Performing Post-orchidectomy Retroperitoneal Lymph Node Dissection |

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.
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Ethical approval

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Consent

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Availability of data and materials

Not applicable.

Author contributions

IE and SE analyzed and interpreted the patient data and wrote the manuscript. AM made the figures. KZ and ZB proposed the study, supervised IE and revised the manuscript. All authors read and approved the final manuscript.

Registration of research studies

Not applicable.

Guarantor

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