International Journal of Surgery Case Reports 93 (2022) 106849

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

First reported case of adult paratesticular myxofibrosarcoma in Indonesia: Case report and literature review

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ARTICLE INFO

Keywords:
Myxofibrosarcoma
Paratesticular tumor
Radical orchidectomy
Case report

ABSTRACT

Introduction and importance: Myxofibrosarcoma is one of the rarest sarcoma types, found in para-testicular regions of the elderly. Although this tumor is detectable by MRI, there has been no specific guideline for managing its recurrence.

Case presentation: A 49-year-old male with a painless scrotal mass was studied. The patient had no other complaint, and the laboratory results showed unremarkable testicular tumor markers. Ultrasound examination of the right hemiscrotum shows a solid mass in the scrotum and right inguinal that compressed the right hemi-testicle. MRI examination of the scrotal region revealed a homogeneous solid mass, while at the lower abdomen, it showed a mass extending from the inguinal canal to the penis shaft and right testis. The patient had no signs of metastatic disease, but after high ligation orchidectomy, a rare paratesticular myxofibrosarcoma was revealed from histopathology examination.

Clinical discussion: Based on existing data and patient MRI imaging, total surgical excision with high ligation orchidectomy is the only curative therapeutic option for low-grade tumors. Furthermore, no recurrent mass was identified during follow-up, and adjuvant chemotherapy or radiotherapy was not administered. The patient was satisfied with the surgery and is on a 6-month routine follow-up to observe the physical symptoms.

Conclusion: This is the first adult paratesticular myxofibrosarcoma case in Indonesia, and it was performed with radical orchidectomy with high ligation due to its invasiveness. However, adjuvant chemotherapy was not provided because the benefit remained inconclusive. The result showed that this approach produces excellent outcome without any relapse.

1. Introduction

Most tumors found in the scrotal sac are malignant and originate in the gonads, while a smaller percentage are extra testicular and stem from paratesticular tissue [1–3]. Despite their rarity, paratesticular tumors have a significant benign rate of 70%, while the remaining 30% are malignant. The paratesticular area is most prevalent for rhabdomyosarcoma formation, contributing to 20% of all cases [3–6]. The ultrasound appearance on these tumors might be deceiving, due to the presence of homogenous, hyperechoic liposarcoma formations [7]. MRI helps to identify and triangulate the tumors better in conjunction with numerous paratesticular structures, which cannot always be conducted using ultrasonography [8]. The knowledge utilized to guide therapy is based on the experiences of a small number of individuals due to the unique nature of Adult paratesticular sarcoma [5]. The best local and systemic therapy for these tumors is still up for debate, although most experts agree that paratesticular sarcomas in adulthood should be treated with complete resection and high spermatic cord ligation [2]. The use of adjuvant chemotherapy for adults with high-grade sarcoma is still controversial. Overall, 5-year disease-specific survival rates for paratesticular tumors have improved from 58 to 80% [2]. High-grade tumors tend to have the worse prognosis, with 62% failing

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https://doi.org/10.1016/j.ijscr.2022.106849
Received 9 December 2021; Received in revised form 18 February 2022; Accepted 22 February 2022
Available online 25 February 2022

systemically or found the retroperitoneal nodes [8]. Therefore, in line with SCARE Guidelines, this study analyzed an adult paratesticular myxofibrosarcoma managed in a tertiaryr care teaching hospital [9].

2. Case presentation

A 49-year-old male arrived in the clinic at the tertiary care teaching hospital complaining of a right scrotal mass five-month-old. Although the mass painlessly grew, the patient was bothered and wanted it removed immediately. Other urinary tract symptoms were denied, and there was no history of fever or weight loss. The patients only had a prior history of hepatitis B, while other chronic diseases such as diabetes mellitus or hypertension were not present with an unremarkable family. Upon examination, a mass of 9 cm in size was found in the right testicle with a firm-to-hard inconsistent mass. The patient pre-operative clinical condition is shown in Fig. 1.

All of the laboratory parameter for this case were within normal limit (LDH serum: 133 U/L; alfa-fetoprotein: 1,85 ng/mL; and β-hCG: <1,2 mIU/ml). The ultrasound evaluation found a solid mass in the scrotum and right inguinal that compressed the right hemitesticle. However, no abnormality was found on the left testicle and surrounding structure (Figs. 2 and 3).

A radical orchidectomy based on MRI results was further performed on the patient before taking the sample for biopsy. On June 23rd, a senior urology oncologist performed surgery on the right inguinal exploration and radical orchiectomy without significant complication. This enables the easy identification of the tumor and its surrounding structure. High ligation orchidectomy was performed because the right testicle adhered to the tumor. Right Retroperitoneal Lymph Node Dissection was not performed on this patient due to insufficient data regarding its benefit. The wound was closed in layers (Fig. 4).

In addition, the specimen of paratesticular mass was sent to Pathology, the result is cell proliferation in spindle form, and stellate with matrix myxoid in the background., it was concluded that the scrotal mass is paratesticular myxofibrosarcoma. FNCLCC Grading System was used to assess this tumor as low-grade paratesticular myxofibrosarcoma (Fig. 5).

The patient attended two weeks post-operative evaluation process at the clinic without any adjuvant chemotherapy or radiation therapy. The wound showed no sign of infection or excessive inflammation, and after two months, the incision scar had healed perfectly without complaints (Fig. 6).

3. Clinical discussion

Paratesticular myxofibrosarcoma is a very rare tumor found in the paratesticular region, with only 8 cases historically recorded in Japan [10]. Myxofibrosarcoma tumor is commonly found in the trunk, head, neck, hands, and feet of the elderly, between the ages of 60 to 80, and rarely found in children under 20 years. Tumors of the retroperitoneum and the abdominal cavity are extremely rare, while the paratesticular ones are painless [8]. Despite of that, A case of paratesticular myxofibrosarcoma was reported in Turkey, with the patient experiencing enlarging pain in his scrotum [11]. The characteristics are pathologically related with spermatic cord origin (high position of the lesion within the scrotum), absence of testicular infiltration (definite testicular borders), neoplastic lesions within the spermatic cord (thick and edematous appearance), and infiltration of the vessel (distention within the cord) [11].

Ultrasound evaluation is unlikely to define the spatial relationship of a paratesticular or extratesticular mass to the testis in terms of mass. However, it is still widely used as the first diagnostic tool to assess this condition [12]. MRI demonstrates the enhancement pattern of scrotal adenomatoid tumors and its effect on the signal intensity of the adjacent testicular parenchyma. The morphologic range of prominent myxoid areas is similar to those reported in other soft tissue sites, such as inflammatory leiomyosarcoma and a neoplasm with epithelioid foci [13]. Myxofibrosarcoma was found on the multinodular tumor with variable cellular spindle cell proliferation on a myxoid background. Furthermore, focal cellular atypia with nuclear pleomorphism and enlarged hyperchromatic nuclei was observed. The characteristic feature is associated with the presence of delicate curvilinear vessels [13].

Radical Orchiectomy is the primary treatment for paratesticular myxofibrosarcoma, due to its invasiveness. After ligation of the vas and spermatic vessels at the internal ring, all cord structures, including the cremaster muscle and fascia, were removed. The testis was delivered with the scrotum's tunica intact to avoid the risk of metastasis. Previous studies found that the recurrence of this tumor after orchidectomy alone was frequent, which led to the recommendation of the adjuvant radiation process to reduce locoregional failure [7]. In cases of intratesticular mass with normal values of tumor markers, malignant pathologies cannot be excluded with safety [14]. The imaging tests in this situation were unable to distinguish the benign nature of the lesion. In the reported case, the right inguinal exploration and radical orchiectomy approaches were used due to the patient's feeling of discomfort with the condition and pre-operative diagnosis of extratesticular mass confirmed by MRI [12]. Therefore, an inguinal approach is recommended with

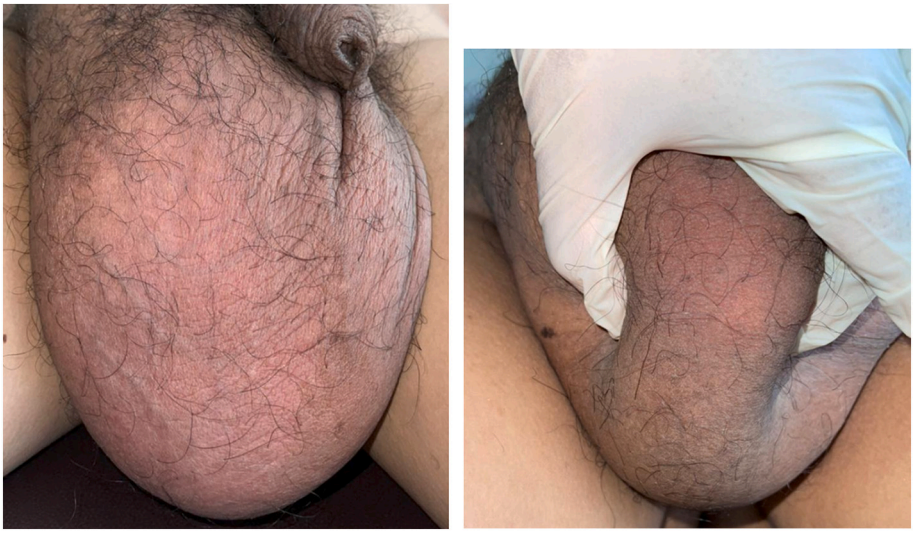

Fig. 1. Clinical picture of the right paratesticular tumor.

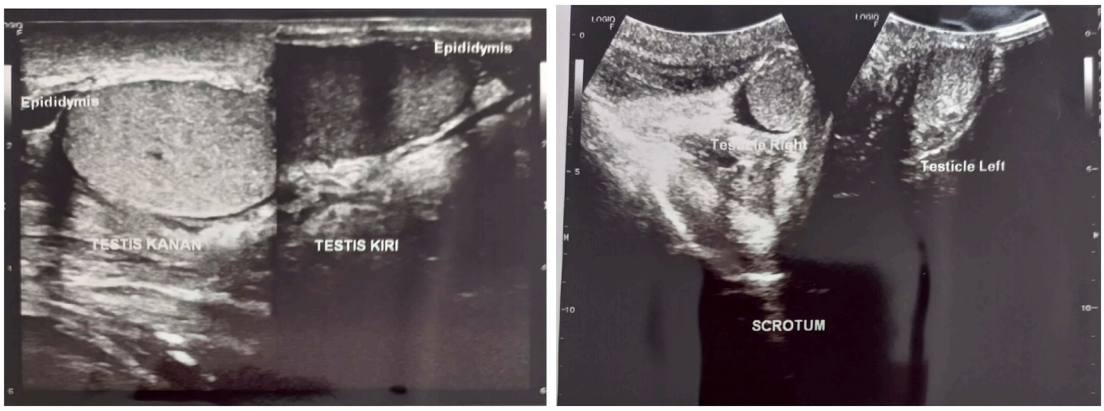

Fig. 2. Ultrasonography evaluation of the testis.

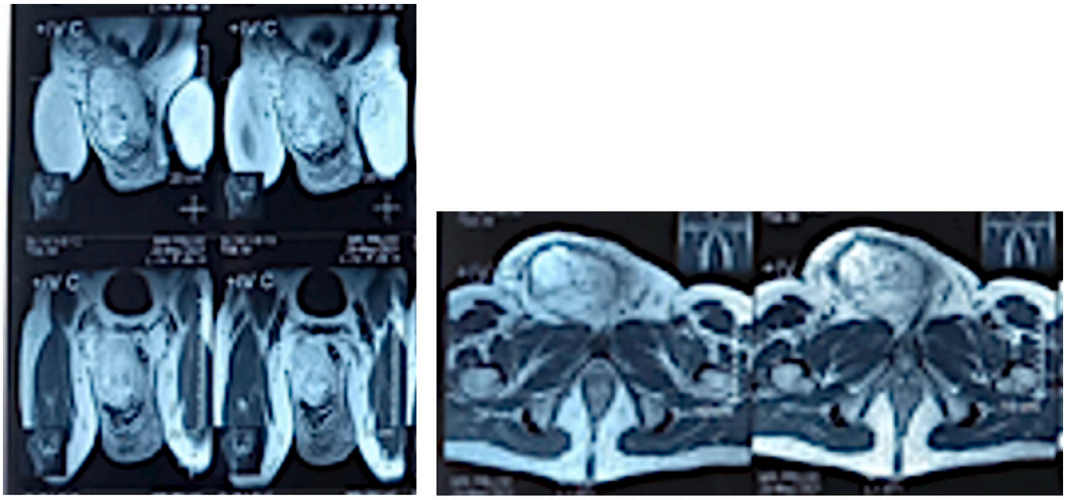

Fig. 3. Imaging of Magnetic Resonance Imaging with the contrast of the lower abdomen. (a) frontal view (b) sagittal view. A right intrascrotal mass was found with a clear margin sized 7,5 × 9,4 × 14,9 cm that compressed the testicle inferiorly.

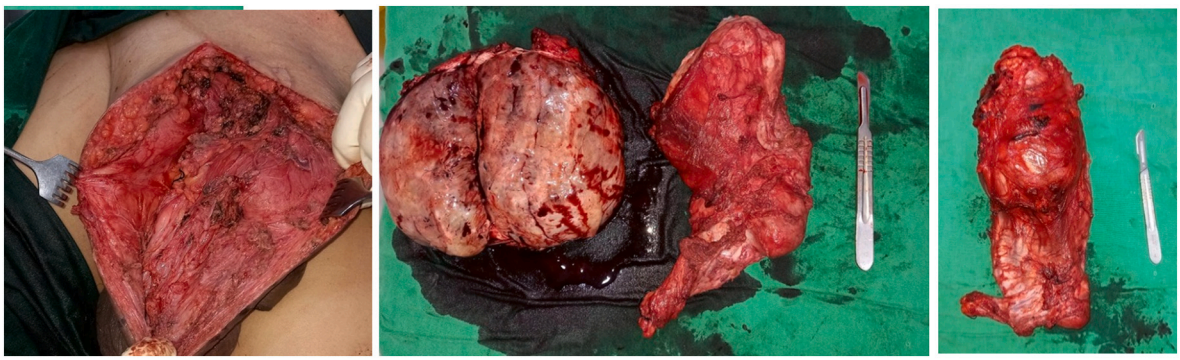

Fig. 4. Intraoperative documentation. (a) intraoperative surgery, (b-c) Macroscopic view of the resected tumor.

isolation of the operative field's testis before the tunica's opening. Histological findings of this case show that the paratesticular mass consists of cell proliferation in the spindle form and stellate with matrix myxoid in the background. Cells with enlarged nuclei, are in the spindle form, and consists of oval, hyperchromatic, and eosinophilic cytoplasm myxofibrosarcoma. Based on this case, it was found that adjuvant chemotherapy was not needed as long as the radical orchidectomy surgery was executed excellently. Furthermore, no reported case of myxofibrosarcoma treated with chemotherapy has been recently reported.

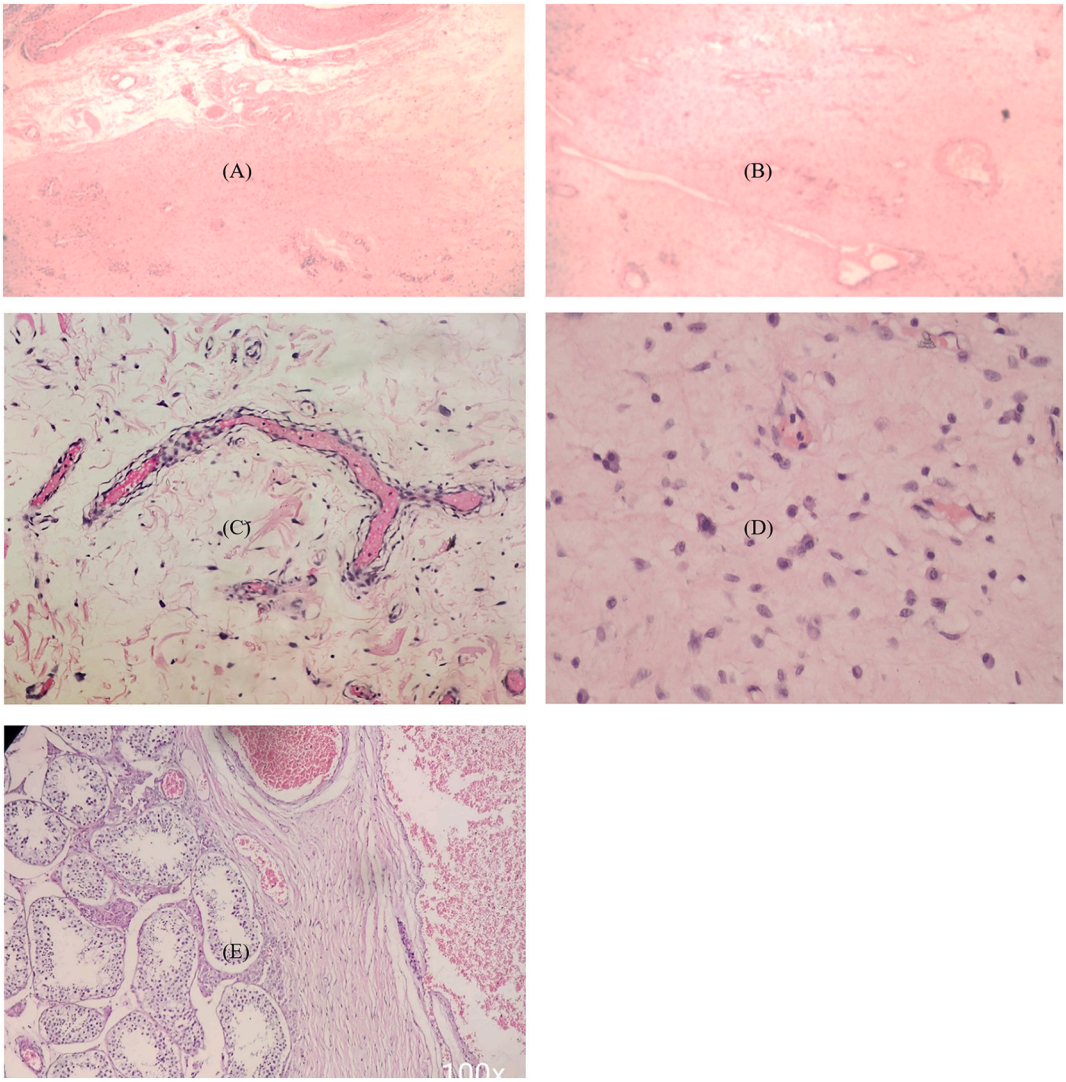

Fig. 5. Histopathological evaluation in this patient. (A) & (B) shown densely packed tumor mass with infiltrative margins (**HE 40**×). (C) Blood vessels proliferate and mostly form a curvilinear pattern (**HE 100**×). (D) a proliferation of spindle- and stellate-shaped cells diffusely dispersed between the loose myxoid matrix. Tumor cells with mucin-filled vacuolated cytoplasm (pseudolipoblasts) were also seen (**HE 400**×). (E) the structure and configuration of the seminiferous tubules were within normal limits. (**HE 100**×).

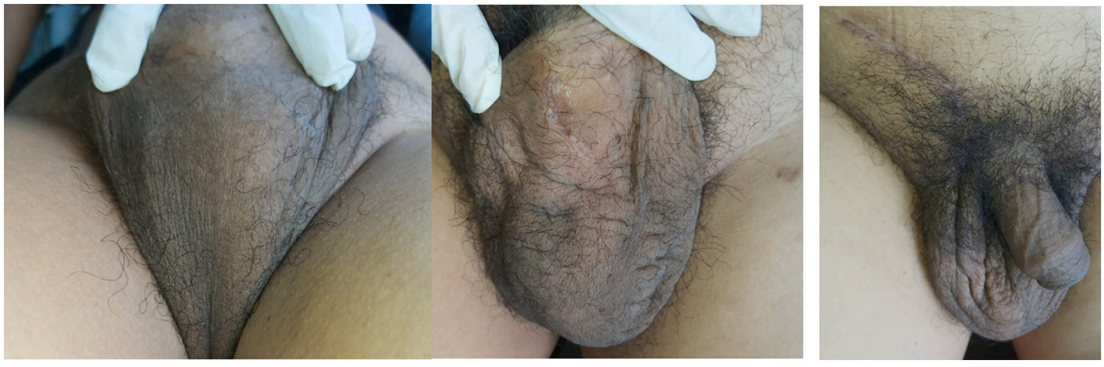

Fig. 6. Clinical picture of 2-month post-operative.

4. Conclusion

This report is the first adult paratesticular myxofibrosarcoma case in Indonesia. Radical orchiectomy with high ligation was performed due to its invasiveness, excluding the use of adjuvant chemotherapy because the benefit is still inconclusive. The patient showed excellent recovery, indicating that this approach gives excellent results without relapse.

Sources of funding

No received funding.

Ethical approval

This study is exempted from obtaining ethical approval from our institution.

Consent

Written informed consent was obtained from the patient to publish this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registration

Not applicable.

Guarantor

Syah Mirsya Warli

Provenance and peer review

Not commissioned, externally peer-reviewed.

CRediT authorship contribution statement

Eggi Respati: Conceptualization, Methodology, Writing Original-Draft, Investigation, Resources.

Fauriski F Prapiska: Supervision, Methodology, Validation, Writing-Review, and editing, Resources.

Bungaran Sihombing: Supervision, Writing-Review and editing, Resources.

Syah Mirsya Warli: Writing-Review and editing, Resources, Guarantor.

Lely Hartati: Writing-Review and editing, Resources.

Sahat Matondang: Writing-Review and editing, Resources.

Declaration of competing interest

The web report has no declarations of interest.

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