

Current Diagnosis, Treatment And Follow-up Procedures of Paratesticular Masses

Paratestiküler Kitlelerin Güncel Tanı, Tedavi ve Takip Prosedürleri

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Özet

Amaç: Paratestiküler kitlelerin testis tümörleri ile pre-op ayrımı net olarak yapılamamaktadır. Genellikle testis tümörü ön tanısıyla radikal orşiektomi yapıp nihai patoloji sonucuna göre klinik yaklaşım planlanmaktadır. Bu noktadan hareketle, klinisyenler skrotal kitle ile karşılaştıklarında, bu kitlenin testis tümörü dışında; epididimidis, tunika vaginalis, spermatic kord, yağ-kas-bağ dokusu gibi destek dokular ve embriyonel kalıntılardan da orijin alabileceğini akılda tutmalı ve tedavi yönetimini buna göre belirlemelidir.

Gereç ve Yöntemler: Merkezimize 2008-2018 yılları arasında skrotal kitle ile başvurmış, testis tümörü kabul edilerek ingüinal radikal orşiektomi yapılan 140 hastanın patoloji sonucu geriye dönük olarak incelendi. Patoloji sonucu paratestiküler kitle rapor edilen olguların preoperatif ve postoperatif verileri, klinik seyri, tedavi yönetimi literatür eşliğinde tartışıldı.

Bulgular: Retrospektif olarak incelenen serimizde 13 olguda paratestiküler kitle saptandı. Bunlardan 10 hastada Adenomatoid tümör, 2 tanesinde Rabdomiyosarkom, 1 hastada Anjiomiksoma saptandı.

Sonuç: İntraskrotal yerleşimli kitlelerin köken aldığı dokunun testis kaynaklı ya da paratestiküler yapılardan mı kaynaklandığının ayırıcı tanısı sıklıkla yapılamamaktadır. Bu nedenle genelde testis tümörleri ile benzer şekilde radikal orşiektomi yapıp kesin tanı patolojik inceleme ile konulabilmektedir. Tüm skrotal kitlelerin %2-3 'ünü oluşturan paratestiküler kitleler, skrotal kitle ile başvuran ve tedavi planlanması yapılan hastaların ayırıcı tanısında akla gelmesi gereken tanılar arasında kendine yer bulmalıdır. Bu konuda daha fazla sayıda hastayla ileri düzey çalışmalara ihtiyaç vardır.

Anahtar Kelimeler: Paratestiküler kitle, Testis tümörü, Radikal orşiektomi, Rabdomiyosarkom

Abstract

Objective: The pre-op differentiation of testicular tumors from paratesticular masses cannot be made clearly. Generally, radical orchiectomy is performed with pre-diagnosis of testicular tumor and a clinical approach is planned according to the final pathological result. When clinicians diagnose a scrotal mass, they should consider that this mass may be originated from epididymidis, tunica vaginalis, spermatic cord, fat-muscle-connective tissue, and embryonic remnants and should determine the treatment management accordingly.

Material and Methods: The pathology results of 140 patients, who were admitted to our center between 2008 and 2018 presenting scrotal mass and underwent inguinal radical orchiectomy considering as testicular tumor, were retrospectively analyzed. Preoperative and postoperative data, clinical course, and treatment management of the cases reported to be paratesticular mass by pathology were discussed in the light of the literature.

Results: Paratesticular mass was detected in 13 cases of our retrospectively analyzed series. Of these, 10 patients were found to have adenomatoid tumors 2 patients have rhabdomyosarcoma and 1 patient angiomyxoma.

Conclusion: The differential diagnosis of whether intrascrotal masses are originating from testicular tissues or paratesticular structures is usually cannot be made. Therefore, radical orchiectomy is performed in the same way as testicular tumors and a definite diagnosis can be made by pathological examination. The paratesticular masses, 2-3% of the scrotal masses, should be considered in the differential diagnosis. In this regard, there is a need for studies with a higher number of series.

Keywords: Paratesticular mass, Testicular tumor, Radical orchiectomy, Rhabdomyosarcoma,

INTRODUCTION

In a patient with a scrotal mass, the underlying pathology may be acute scrotum (testicular torsion, epididymo-orchitis epididymitis, orchitis), which is among the urological emergencies, as well as an extensive clinicopathological condition consisting of hydrocele, varicocele, testicular tumor, epididymal cyst/mass, cyst-mass in the spermatic cord, and inguinal hernia. Although the majority of testicular masses have a malignant characteristic, approximately 70% of paratesticular masses have a benign characteristic.(1) The first diagnostic method in the differential diagnosis is the ultrasound(US) following anamnesis and physical examination. Besides the US can identify the characteristics of a mass such as solid, cystic, it can show whether it is testicular or paratesticular. It has a sensitivity close to 100% in the diagnosis of testicular tumor. (2) Magnetic Resonance Imaging(MRI) may provide more accurate information in terms of localization, association with surrounding tissues and invasion.(3,4)

The majority of the paratesticular masses, 2-3% of scrotal masses, are benign. With regard testis sparing surgery can be applied in paratesticular masses.(5) However, standard inguinal radical orchiectomy is performed in testicular masses except for special cases (Solitary testis, bilateral multiple testicular masses).(6)

In this retrospective study, we aimed to evaluate the cases operated in our center and diagnosed with paratesticular mass within the context of the literature and to identify the diagnosis, treatment and follow-up procedures.

MATERIAL AND METHODS

140 patients who underwent radical orchiectomy between 2008 and 2018 were identified retrospectively by reviewing the hospital records in our center's database. 13 paratesticular mass cases were found. In the first stage, a high-ligation inguinal radical orchiectomy had been performed for all cases. When the pathology results were reviewed, 10 Adenomatoid tumor(AT), 1 Aggressive angiomyxoma (AAM) (1) and 2 Rhabdomyosarcoma (RMS) cases were seen.

RESULTS

Benign Masses

In paratesticular benign masses, patients diagnosed with AT and AAM are followed up without any further intervention, since additional treatment is not required after inguinal radical orchiectomy with negative surgical margin. (7)

Malignant Masses

In both cases with RMS, time between the onset of symptoms and the duration of admission is remarkable and 7 and 10 days respectively. Alpha Fetoprotein, Human chorionic gonadotropin and lactate dehydrogenase were normal in both cases. The physical examination of the 15 years old patient, who was admitted with the complaint of gradually increasing pain and growth in the left hemiscrotum after the scrotal trauma occurred about one week ago, revealed an increase in the size of the left hemiscrotum, edematous appearance and tenderness by palpitation (Image 1). In the US examination, a 85x42 mm hypervascular solid lesion with lobular contour adhered to the testicle and thought to be originated from the testicle was visualized in the left scrotum. The subsequently performed scrotal MRI showed a massive lesion of 113x68 mm in size with cystic-necrotic components which involved the left hemiscrotum nearly total and showed a heterogeneous contrast uptake. The pathology result of the patient underwent left inguinal radical orchiectomy was Stage IV embryonal type RMS.(Image 2,3) All of the abdominal computed tomography(CT) and F-18 fluorodeoxyglucose positron emission tomography(FDG-PET) examinations showed a diffuse intraabdominal lymphadenopathy (LAP) and bone marrow involvement. Although bone marrow aspiration and biopsy revealed hypocellular bone marrow appearance, RMS infiltration was not detected. VAC (Vincristine, Actinomycin-D, Cyclophosphamide) and VC (Vincristine) combination treatment was initiated for the patient. This chemotherapy treatment continued with 40 cycles. Granulocyte colony-stimulating factor(G-CSF) treatment for cellular support was given intermittently.

Before the radiotherapy (RT), the elevation and fixation of the right testicle onto the external oblique fascia was performed by surgical intervention. Orchiopexy was again performed after chemotherapy plus radiotherapy (24 sessions of RT in total). In the last FDG-PET, the patient is followed up with no residual and recurrent mass.

In the other 18-year-old RMS case, the patient presented with complaints of growth and mild pain start-

ing 10 days before the left hemiscrotum. There was no trauma and additional risk factors. On physical examination mass lesion was found in the left caudal junction. In US, a heterogeneous mass of 46x40 mm in size was visualized in the inferior pole of the left testicle. The MRI showed that the mass was extratesticular, solid lesion with epididymal origin. During the exploration with inguinal approach, inguinal radical orchiectomy plus high cord ligation was performed



Figure 1. Before operation

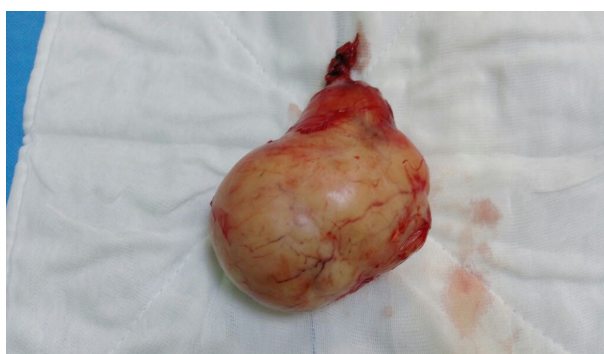


Figure 2. The mass after orchiectomy

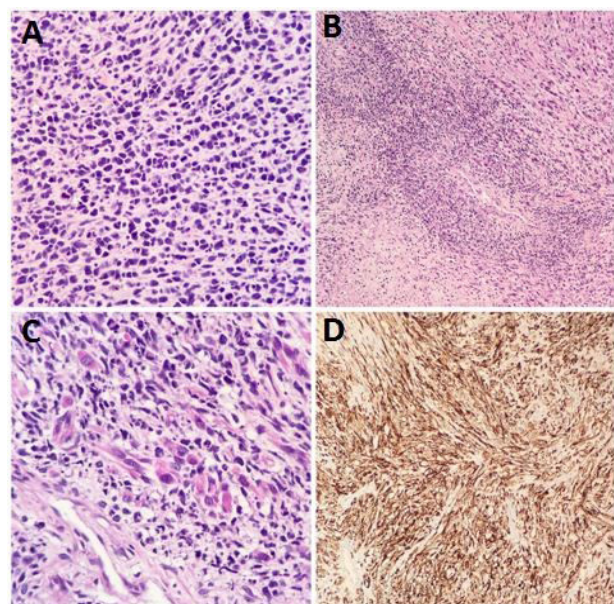


Figure 3. Embryonal Rhabdomyosarcoma

- 3A: The majority of the neoplastic cells have oval-round nucleus and primitive blastic appearance. There are too many mitoses and apoptosis.
- 3B-C: Rhabdomyoblasts that show an increased cellularity around the vessels and intrastoplasmic striae at the periphery are visualized.
- 3-D: Desmine

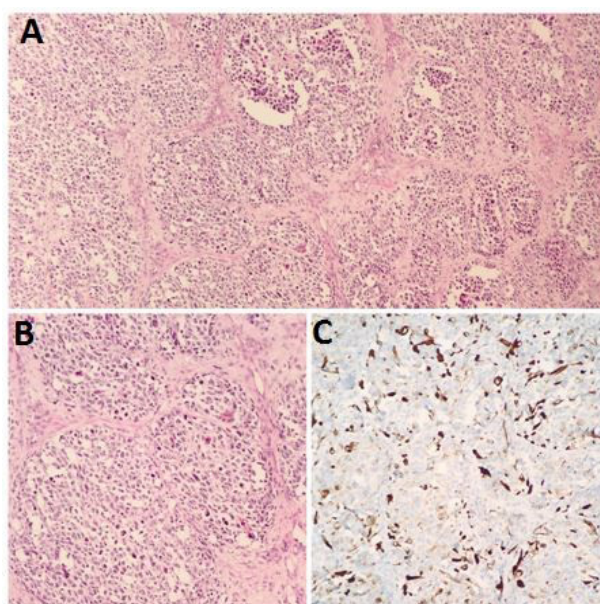


Figure 4. Alveolar Rhabdomyosarcoma

- 4-A-B-C: Small, round or oval cells are visualized in the form of neoplastic islands separated from each other by connective tissue.
- 4-C: Desmine

since the frozen result showed a malignant characteristic. The pathological examination revealed Alveolar type RMS. The CT and FDG-PET for staging showed multiple intraabdominal LAP, the largest of which was 4,5 cm in the left infrarenal area, and activity uptake in the left scrotum.(Figure4) After that retroperitoneal lymph node dissection(RPLND), left scrotal skin excision, right testicle elevation were performed and then chemotherapy(VAC) and RT were started. He received 12 cycles of chemotherapy and 24 sessions of RT in total. The scrotal skin excision pathology was reported as reactive granulation tissue and the RPLND pathology was reported as reactive lymph node. The patient who underwent right orchiopexy after chemotherapy plus radiotherapy has been followed up without recurrence.

DISCUSSION

Lipomas are the most common paratesticular benign tumors and constitute about 90% of spermatic cord tumors.(7). It may sometimes be difficult to make differential diagnosis from liposarcomas which show a more aggressive growth, infiltrate peripheral tissues with irregular borders and is seen at later ages. Well-defined slow-growing yellowish structure and similar US echogenicity with normal fatty tissue are helpful in differential diagnosis. (8,9,10)

AT is the second most common, constitute about 30% of all paratesticular masses and 60-65% of benign tumors.(11,12,13) Non-hormone-dependent AT, also referred to as benign mesothelioma, is often located on the head and tail part of the epididymis, it may also be originated from tunica vaginalis, tunica albuginea, rete testicle, spermatic cord. Even though it can be incidentally detected in epididymo-orchietomy material or autopsy, it is usually presented with a palpable and painless mass.(14) An exploration with a scrotal approach can be performed if it is definitely thought to be an extratesticular mass with benign appearance in the preoperative evaluation.(7) If a benign character is detected per-operatively in the frozen sampling, the excision if possible enucleation of the mass should be performed and the testicle should be conserved. (15) Surgical excision or radical orchiectomy provides a curative treatment for adenomatoid tumors and no additional treatment is required.

Angiomyxoma, another benign tumor, is the mass that usually progress slowly, generally without distant metastasis, but with local infiltration. The best imaging method for diagnosis is MRI.(16) Histopathologically, it is divided into three subgroups: aggressive angiomyxoma, angiomyofibroblastoma, and superficial angiomyxoma. In the case of these tumors, the aim is to provide surgical margin negativity. Despite the non-metastatic characteristics in general, systemic imaging should be performed because of case reports reporting lung metastases. Angiomyxoma can also be seen in the female urogenital system and gonadotropin-releasing hormone analogues are used in cases with positive estrogen, progesterone receptor and surgical margin positivity or in cases where a complete resection cannot be obtained. There is no similar treatment in male patients in the literature. The high infiltration capacity of the tumor in terms of recurrence and the surgical margin positivity are the most important predictive parameters. Therefore, especially patients in the risk group should be closely followed up and surgical excision should be performed again if a recurrent mass develops. In our patient with pathologic aggressive angiomyxoma no tumor was detected at the surgical margin. Metastases were not detected in systemic cross-sectional views. Routine and close follow-up did not reveal any additional pathology.

Leiomyoma, papillary cystadenoma, angioma, dermoid cyst, fibroma, hamartoma, teratoma, choletoma, rhabdomyoma can be regarded as other benign tumors. In such cases, total excision of the mass is generally curative. However, in cases with papillary cystadenoma is detected, a systemic evaluation should be performed in terms of Von-Hippel Lindau (VHL). Especially in 17% of bilateral cases, VHL coexist.(17) Surgical excision of the mass is also curative in papillary cystadenoma.

Paratesticular malignant tumors constitute about 2-3% of all intrascrotal tumors and sarcomas 2-3% of these. Following liposarcoma and leiomyosarcoma, RMS is the third most common paratesticular sarcoma of childhood seen at later ages such as at the 6th and 7th decade and has a bimodal age distribution of 4 and 18 years of age.(18) Approximately 80% of it is seen

under 21 years of age. The most common subtype is embryonal RMS. Other subtypes include alveolar, botryoid and pleomorphic RMS.(19) One of our patients was embryonal RMS and the other was alveolar type RMS. Malignant paratesticular tumors, which are usually painless and rarely painful, can reach large sizes in a short time. It is reported in the literature that 92% of the tumor is localized at the time of diagnosis.

Treatment planning is carried out with multidisciplinary approach. The basic treatment is to provide adjuvant chemotherapy and radiotherapy after performing high-ligation inguinal radical orchiectomy, pelvic, ipsilateral or bilateral RPLND in which surgical margin negativity is obtained. An intact testis elevation before radiotherapy should be done to protect the fertility. In the case of scrotal involvement, hemiscrotectomy should be performed. If inguinal lymph node involvement is present, inguinal lymph adenectomy should also be added.(20)

In order to evaluate post-operative false negative or positive results that may be caused due to surgery, pre-operative abdomen and lung CT and FDG-PET CT should be performed and used as a guide in surgical planning. If lymph node involvement is radiologically positive in pre-op imaging, RPLND can be performed with orchiectomy. Although RMS, which can demonstrate hematogenous invasion to local peripheral tissues and lymphatic spread, primarily metastasize to lung but may also to all systems. Bone marrow aspiration and biopsy should be performed. Because pancytopenia can be seen both secondary to bone marrow involvement and chemotherapy, G-CSF are used in the treatment. In children, the 5-year progression-free survival in localized disease after primary surgery and chemotherapy plus radiotherapy reached up to 94%, while this rate drops to 40% in the case of metastatic disease. The 5-year progression-free survival FS increased from 68% in patients without RPNLD to 90% in patients with RPNLD (21).

In accordance with the literature, we performed radical orchiectomy in both patients and then applied chemoradiotherapy. Bone marrow biopsy and testicular elevation before RT were performed in both of our patients. One patient underwent RPLND and hemis-

crotoectomy. The patient underwent orchiopexy at the end of chemotherapy plus radiotherapy. We continue to coordinate the treatment of the other patient with pediatric oncology and radiation oncology.(10)

Other malignant paratesticular tumors include liposarcoma, malignant mesothelioma other than leiomyosarcoma, ovarian-type müllerian epithelial tumors, epididymal adenocarcinoma, and very rarely malignant fibrous histiocytoma. The common treatment is high-ligation inguinal radical orchiectomy similar to RMS. Although there is no consensus in terms of RPLND and chemoradiotherapy, they are generally not carried out.(22)

CONCLUSION

Although paratesticular masses are rarely seen among scrotal masses with a ratio of 1-2%, they are rapidly progressive pathologies that may be fatal if they are malignant tumors, and the mortality and morbidity can be significantly reduced by early diagnosis and treatment. It should be remembered that scrotal masses may also be a paratesticular mass and organ loss can be avoided with organ sparing surgeons. Malignant tumors should be close followed-up because of their aggressive nature and frequent recurrence potentials. As a result, there is a need for studies involving diagnosis and treatment outcomes of large patient series on diagnosis and treatment.

Abbreviations

AAM: Aggressive angiomyxoma

AT: Adenomatoid tumor

CT: Computed tomography

FDG-PET: F18-fluorodeoxyglucose positron emission tomography

G-CSF: Granulocyte colony-stimulating factor

LAP: Lymphadenopathy

MRI: Magnetic resonance imaging

RMS: Rhabdomyosarcoma

RPLND: Retroperitoneal lymph node dissection

RT: Radiotherapy

US: Ultrasound

VAC: Vincristine, Actinomycin-D, Cyclophosphamide

VHL: Von-Hippel Lindau

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