SAS Journal of Surgery ISSN 2454-5104
SAS J. Surg., Volume-2; Issue-3 (May-Jun, 2016); p-90-93
Available online at http://sassociety.com/sasjs/

Case Report

Burned-Out Phenomenon Presented a Multiple Retroperitoneal Masses: A Case Report

Hakan Türk[1]*, Mehmet Yoldaş[1], Ebru Çakir[2], Sümeyye Ekmekçi[2], Oğuz Mertoğlu[3], Ferruh Zorlu[3]
[1]Dumlupınar University, Evliya Celebi Training and Research Hospital, Department of Urology, Kutahya, Turkey
[2]Tepecik Training and Research Hospital Deparment of Pathology, Izmir, Turkey
[3]Tepecik Training and Research Hospital Deparment of Urology, Izmir, Turkey

***Corresponding author**
Hakan Türk
Email: hkntrk000@hotmail.com

Abstract: "Burned-out" testicular tumor is a rare clinical entity. The term "burned-out" actually represents a spontaneously and completely regressed testicular tumor that manifests itself by metastasis to retroperitoneal region and mediastinal, supraclavicular, cervical and axillary lymph nodes. This is a different entity than primary extragonadal testicular tumors and is seen more rarely. Here we present a case diagnosed with "burned-out" testicular tumor and we discuss his clinical, radiological and histopathological features.
Keywords: Burned-out, testicular tumor, retroperitoneal, metastasis, extragonadal

INTRODUCTION

"Burned-out" testicular tumor represents a germ cell testicular tumor that has regressed spontaneously and completely, but metastasized to retroperitoneal region more commonly. It is often confused with primary extragonadal germ cell tumors. The first case was presented by Prime in 1927 [1]. In the autopsy of a patient who died of metastatic germ cell tumor but having no mass in the testes, Azzopardi et al. showed the signs of spontaneous regression in the testes [2]. It is also shown that sex cord-stromal tumors may exist in the retroperitoneal area [3]. Some scientists have tried to explain the etiopathogenesis of the disease with immunologic and ischemic causes but it hasn't been fully elucidated yet [4]. In this study we aimed to present a case diagnosed with "burned-out" testicular tumor, by supporting with relevant literature.

CASE PRESENTATION

Forty-two-years-old male patient admitted to our clinic with complaints of left flank pain for two years, and his physical examination revealed no abnormal findings. Computed tomography of abdomen showed a 6.5 x 4.5 cm mass in the retroperitoneal region, around para-aortic and para-caval area (Figure 1). A 12 x 18 mm hypoechoic, heterogeneous mass lesion was observed in doppler ultrasound (US) which caused irregularities in the left testicle capsule. Thereupon, ultrasound guided tru-cut biopsy was performed from retroperitoneal region on the patient. Macroscopic examination of the biopsy material obtained from retroperitoneal region revealed 1.3 cm sized fragmented tissue. Small round neoplastic cells showing nuclear pleomorphism were observed microscopically. The neoplastic islands of tumor cells were accompanied by lymphoid cells. Immunohistochemically, tumor cells were positive with vimentine, CD117, D2-40, placental alkaline phosphatase (PLAP) and negative with desmin, melanA, HMB-45, S100, pancytokeratin, CK7, CD34. The proliferation index was as high as 85% with Ki 67. The final diagnosis was malignant tumor consistent with germ cell tumor. Laboratory investigations were within normal ranges, with α-fetoprotein (AFP):1.86 IU l^{-1} (0–9 IU l^{-1}), β-human chorionic gonadotropin (HCG):2.59 IU l^{-1} (<0.1 IU l^{-1}), and lactate dehydrogenase (LDH): 183 IU l^{-1}(100–190 IU l^{-1}). There was no lung metastasis observed on the chest X-ray. The patient underwent left inguinal orchiectomy because of hypoechoic heterogeneous mass lesion detected in the left testis.

Regarding orchiectomy pathology**,** macroscopic examination of the left total orchiectomy material revealed a 2.3X1.7X1.5 cm nodulary lesion in the parenchyma of testis.

Microscopic examination showed a well-demarcated nodular fibro-hyalinized scar associated with hyalinized and atrophic seminiferous tubules (Figure 2).

There was no evidence of testicular germ cell tumor and intratubular germ cell neoplasia histomorphologically and immunohistochemically with PLAP, CD117, D2-40. The diagnosis was consistent

with burned-out testicular germ cell tumor (Figure 3). The patient followed a chemotherapy protocol of four cycles with bleomycin, etoposide, and cisplatin (BEP). Follow-up CT after chemotherapy revealed that the para-aortic lymph nodes had decreased in size to 3.4 cm × 2 cm × 1.6 cm. The patient then underwent a retroperitoneal lymph node dissection. The para-aortic mass was determined completely as necrosis in pathological examination. No pathology was detected at 12-month follow-up control.

Fig-1: Computed tomography of abdomen showed mass in the retroperitoneal region, around paraaortic and paracaval area

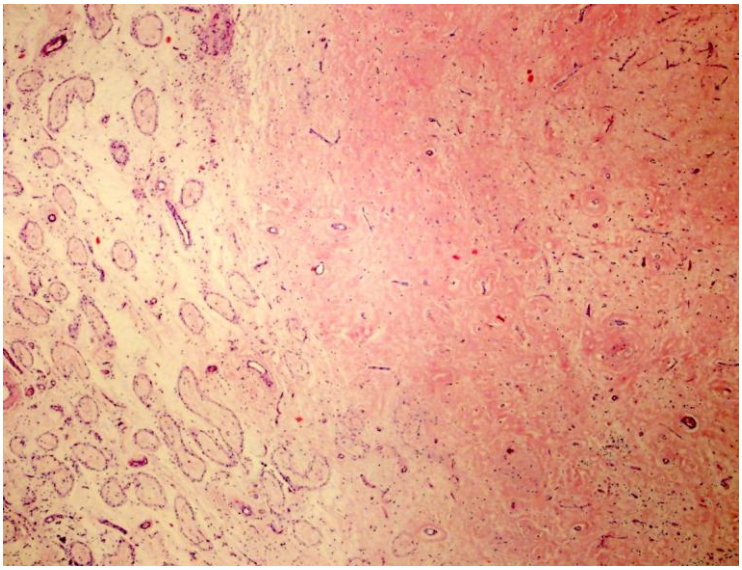

Fig-2: Well demarcated nodular fibrohyalinized scar associated with hyalinized and atrophic seminifer tubules (H&E, X40)

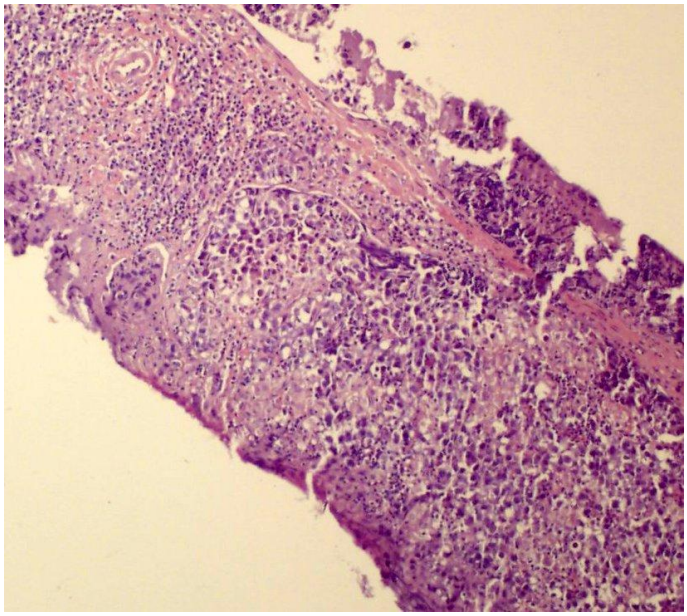

Fig-3: Small round neoplastic germ cell islands accompanied by lymphoid cells (H&E, X100)

DISCUSSION

It is quite difficult to distinguish "burned out" tumors of the testis from primary extragonadal germ cell tumors. Any patient with retroperitoneal or mediastinal germ cell tumor should be evaluated in detail to make sure whether it is primary testicular germ cell tumor or not.

Primary extragonadal germ cell tumors usually occur in retroperitoneal or mediastinal regions and they constitute 5-10% of the germ cell tumors [5]. The most widely accepted theory is that these tumors develop from primordial (embryonal) germ cell tumors settled in the midline of the body [6]. However, it is not yet clear whether these are primary extragonadal tumors indeed or simultaneous presentations of testicular and extragonadal tumors or a metastases of undetectable and regressed (burned-out) primary testis tumors. It has been reported that some of the patients which were initially thought to have primary extragonadal germ cell tumors, actually had primary testicular tumor metastases. However this could not be diagnosed because the primary tumor was regressed [7]. In our case, the tumor was not possibly identified in the testis and the diagnosis was established through retroperitoneal biopsy.

The first case of "burned-out" testicular tumor reported by Prim *et al* [1] and several subsequent series [4, 8, 9] have demonstrated its features in detail. Azzopardi *et al.* observed fibrous scar in the testis and seminomatous and non-seminomatous residual testicular tumor in the autopsy of a case having no palpable mass in the testes with diffuse germ cell tumor metastases [2].

There are mainly two mechanisms explaining the phenomenon of burn-out. First is the regression of primary germ cell tumor after metastasis. This is presumed to occur due to the immune system response and ischemia induced by metabolic activity of the tumor. Another mechanism is speculated as the development of a new tumor in extragonadal tissues [10]. Although rare, spontaneous regression without extragonadal metastatic lesions were reported as well [11, 12]. Histopathological examination of the testes may help to dspecify some facts indicating tumor regression. Possible findings include scar formation, intratubular calcification, lenfoplasmositic infiltration, haemosiderin containing macrophages and testicular atrophy [13]. Nevertheless, it should be kept in mind that, these findings may develop secondary to ischemia, trauma, and infection. Presence of intratubular germ cell neoplasia of the unclassified type (IGCNU) concurrently with testicular scarring is one of the important indicators of regression. Even co-occurrence of scarring and IGCNU without any known metastatic lesion can be interpreted as a sign of regression. In addition, the presence of numerous small blood vessels in the scar tissue indicates angiogenesis of a regressed tumor [7].

In fact, it is entirely up to a comprehensive testiculary evaluation to distinguish primary extragonadal germ cell tumor from primary testis tumor that has metastasized to retroperitoneal region. In the study by Scholz et al. suspected lesions were detected only in 58% of the patients by palpation, while in approximately 10% of the remaining on ultrasonography [14]. Testis USG displaying hyperechoic foci without any hypoechoic lesion in patients having extragonadal germ cell tumors and normal testes palpation should suggest the burned-out testicular tumor. These foci represent hematoxyphilic bodies or psammoma bodies [9]. Nevertheless, in cases with extragonadal germ cell tumors, testes are generally

found normal in the ultrasonography. In our case too, there were hypoechoic foci in the ultrasound examination of the testes. For this very reason, the patient was considered as "burned-out" phenomenon and performed inguinal orchiectomy.

Research data show that primary tumor generally regress after metastasis developes. Surgical removal of the primary tumor is esential. Because persistent testicular malignancy is seen in 50% of patients despite systemic chemotherapy [3, 8].

CONCLUSION

Primary extragonadal germ cell tumors in the retroperitoneum are extremely rare. "Burned-out" testicular tumor should be considered and orchiectomy should be performed in those patients with normal findings in physical examination of testes and having retroperitoneal lymph node metastases as well as germ cell tumors in histological examination.

REFERENCES

1. Prim P; Spontanheilung eines bösartigen wahrscheinlich chorion-epitheliomatosen. Gewaches im Hoden. Virchows Arch Path Anat., 1927; 265:239-58.
2. Azzopardi JG, Mostofi FK, Theiss EA; Lesion of testes observed in certain patients with widespread choriocarcinoma and related tumors. The significance and genesis of hematoxylinstaining bodies in the human testis. Am J Pathol., 1961; 38:207-25.
3. Golombos D, Brison D, Nejad HS; Malignant sertoli cell tumor of the testis with a large retroperitoneal mass in an elderly man. Urol J., 2010; 7:281-3.
4. Böhle A, Studer UE, Sonntag RW, Scheidegger JR; Primary or secondary extragonadal germ cell tumors?. The Journal of urology, 1986; 135(5):939-43.
5. Schmoll HJ; Extragonadal germ cell tumors. Ann Oncol., 2002; 13:265-72.
6. McLeod DG, Taylor HG, Skoog SJ, Knight RD, Dawson NA, Waxman JA; Extragonadal germ cell tumors. Cancer, 1988; 61:1187.
7. Balzer BL, Ulbright TM; Spontaneous regression of testicular germ cell tumors: An analysis of 42 cases. Am J Surg Pathol., 2006; 30:858-65.
8. Lopez JI, Angulo JC; Burned-out tumor of the testis presenting as retroperitoneal choriocarcinoma. Int Urol Nephrol., 1994; 26:549-53.
9. Comiter CV, Renshaw AA, Benson CB, Loughlin KR; Burned-out primary testicular cancer: sonographic and pathological characteristics. The Journal of urology, 1996; 156(1):85-8.
10. Hainsworth JD, Greco FA; Extragonadal germ cell tumors and unrecognized germ cell tumors. Semin Oncol., 1992; 19:119-27.
11. Minamida S, Irie A, Ishii J, Minei S, Kimura M, Iwamura M, Morinaga S; Case of seminoma with possible spontaneous regression without extragonadal metastatic lesion. Urology, 2007; 70(6):1222-e1.
12. Fabre E, Jira H, Izard V, Ferlicot S, Hammoudi Y, Theodore C, Di Palma M, Benoit G, Droupy S; 'Burned-out'primary testicular cancer. BJU international, 2004; 94(1):74-8.
13. Balalaa N, Selman M, Hassen W; Burned-out testicular tumor: a case report. Case Rep Oncol., 2011; 4:12-5.
14. Scholz M, Zehender M, Thalmann GN, Borner M, Thöni H, Studer UE; Extragonadal retroperitoneal germ cell tumor: evidence of origin in the testis. Annals of Oncology, 2002; 13(1):121-4.