A Case of Burned-out Testicular Germ Cell Tumor Diagnosed Eleven Years Later

On Bir Yıl Sonra Teşhis Edilen Spontan Regrese Olmuş Testis Germ Hücreli Tümör Olgusu

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ABSTRACT

Testicular tumors are a prevalent solid cancer among young men. The burned-out testicular seminoma phenomenon has been documented in several cases in the literature. Typically, these cases are diagnosed after metastasis, with both the metastatic lesions and the primary tumor exhibiting spontaneous regression. We present the case of a 39-year-old male patient who was previously diagnosed with a malignant teratoma in the left supraclavicular region 11 years ago. The patient was admitted to our clinic with inguinal pain, and further investigations revealed masses in the retroperitoneal and left testicular regions. Surgical resection confirmed these masses as spontaneously regressing teratomas and teratoma metastases. The patient did not receive systemic chemotherapy and remained asymptomatic during the follow-up period**.** This rare phenomenon, for which the underlying pathophysiology remains uncertain, is believed to be associated with the patient's immune response. Further studies are warranted to investigate and understand this unique phenomenon in greater detail.

Keywords: Spontaneous tumor regression, testicular cancer, immune system, lymph node metastasis

Received: 10.25.2022 **Accepted:** 07.13.2023

ÖZET

Testis tümörleri genç erkekler arasında yaygın görülen bir kanserdir. Spontan regrese testis tümörü fenomeni literatürde çeşitli vakalarda belgelenmiştir. Tipik olarak bu vakalara metastazdan sonra teşhis konulmakta; hem metastatik lezyonlar hem de primer tümörde spontan gerileme gözlenmektedir.Onbir yıl önce sol supraklaviküler bölgede malign teratom tanısı alan 39 yaşında erkek hastanın olgusunu sunuyoruz. Kasık ağrısı şikayetiyle kliniğimize başvuran hastanın yapılan tetkiklerinde retroperitoneal ve sol testis bölgesinde kitle tespit edildi. Cerrahi rezeksiyon bu kitlelerin spontan gerileyen teratomlar ve teratom metastazları olduğunu doğruladı. Hasta sistemik kemoterapi almadı ve takip süresince semptomsuz kaldı. Altta yatan patofizyolojinin belirsizliğini koruduğu bu nadir olgunun, hastanın immün yanıtıyla ilişkili olduğuna inanılmaktadır. Bu eşsiz fenomeni daha ayrıntılı olarak araştırmak ve anlamak için daha ileri çalışmalara ihtiyaç vardır.

Anahtar Sözcükler: Spontan tümör regresyonu, testis kanseri, immun sistem, lenf nodu metastazı

Geliş Tarihi: 25.10.2022 **Kabul Tarihi:** 13.07.2023

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doi:http://dx.doi.org/10.12996/gmj.2023.89

INTRODUCTION

Testicular cancer is the most frequently diagnosed solid organ malignancy among young men and remains a leading cause of cancer-related mortality in this population. Seminoma is the most prevalent type of germ cell tumor (GCT), while non-seminomatous tumors are more frequently encountered in a metastatic setting (10% versus 2.5%) (1). A rare clinical entity within testicular tumors is the phenomenon of a "burned-out" primary tumor. Such tumors are characterized by spontaneous and complete regression of the testicular lesion, while metastases are present in the retroperitoneum, distant lymph nodes, lungs, and liver (2). Among germ cell tumors, choriocarcinoma is the most commonly associated with spontaneous regression, followed by embryonal carcinoma (3).

In this case report, we describe a unique case in which the primary testicular tumor completely regressed eleven years after the diagnosis of distant lymph node metastasis.

CASE REPORT

A 39-year-old male patient presented with complaints of left inguinal and abdominal pain. The inguinal pain persisted regardless of rest or exertion. Physical examination revealed a pathological left inguinal lymph node, while no abdominal abnormalities were noted upon examination.

Upon reviewing the patient's medical history, it was discovered that he had previously visited our hospital in 2009 with a palpable mass in the left supraclavicular region. The mass was surgically excised, and pathological evaluation identified an 8-cm lymph node containing a malignant teratoma with mesenchymal and epithelial components, as well as poorly differentiated neuroepithelial tumor components. Scrotal Doppler ultrasonography and systemic imaging were recommended to determine the origin of the tumor. However, the patient declined the recommended radiological imaging and additional laboratory tests. Subsequently, the patient did not seek medical attention for 11 years.

Considering the patient's medical history, a potential association between his current complaints and his previous malignancy was suspected. The patient's complete blood count and biochemical values were within normal limits, and tumor marker levels were also normal (B-hCG <2 mIU/ml, AFP 2.1 ng/mL). Scrotal Doppler ultrasound revealed homogeneous right testicular parenchyma, while the left testicular parenchyma exhibited a heterogeneous appearance with hypo-hyperechoic areas and small calcified foci. Abdominal computed tomography demonstrated a solid lesion measuring approximately 40 x 31 mm in the left paraaortic area, adjacent to the anterior aspect of the left renal vein. Additionally, there were cystic necrotic areas measuring approximately 59x62 mm inferior to this lesion and a solid lesion with amorphous calcifications causing a lateral indentation in the left ureter. Positron emission tomography-computed tomography (PET-CT) was requested to assess the systemic spread and metabolic activity of the tumor. PET-CT revealed cystic lesions with solid components in the abdominal paraaortic area, measuring approximately 5.5 cm in diameter at the level of the aortic bifurcation, along with increased 18F-FDG uptake in the left inguinal lymph node, measuring approximately 9 mm in diameter.

The imaging and laboratory results were discussed by a multidisciplinary tumor board, which decided upon surgical resection of the lesions. The patient subsequently underwent retroperitoneal lymph node dissection and unilateral inguinal orchiectomy. Pathological evaluation revealed a scar area consistent with a regressed germ cell tumor, along with in-situ germ cell tumor areas surrounding it (Figure I). A total of nine lymph nodes were removed from the inguinal, paracaval, and paraaortic regions, with two of the lymph nodes showing postpubertal teratoma metastases.

Given the spontaneous regression of the patient's tumor and the absence of germ cell tumor focus in the pathological evaluation, it was decided to monitor the patient without further treatment. The patient has been under surveillance for approximately 18 months, during which time the pain complaints completely resolved, and there has been no evidence of recurrence in the latest imaging studies.

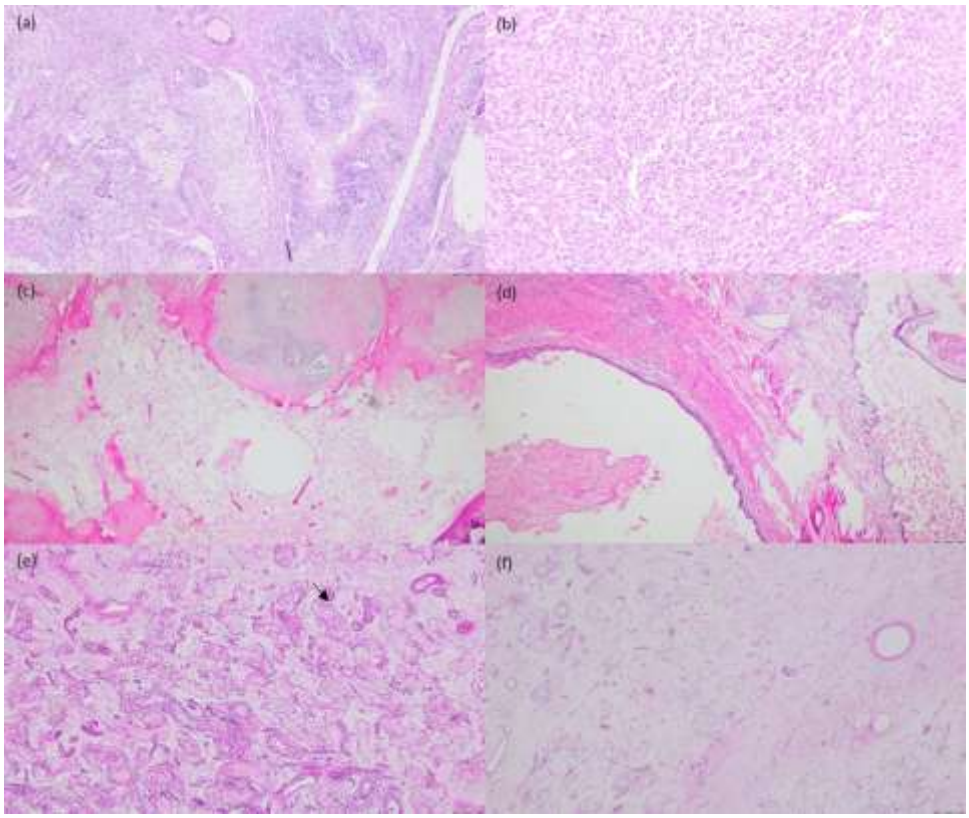

Figure I. Pathological evaluation of excision materials

The cervical mass lesion with teratomatous elements (a) and sarcomatoid differentiation (b). Retroperitoneal mass characterized by mature cartilage, ossification and cysts lined by mature cuboidal and columnar epithelium (c, d). Orchiectomy which reveals atrophic seminiferous tubules (e, arrow) and well defined hyalinised and vascular scar formation (f).

DISCUSSION

The phenomenon of spontaneous regression of cancer has been observed in various tumor types, including testicular tumors, kidney tumors, breast carcinoma, lymphoma, and malignant melanoma. However, the underlying pathophysiology of this phenomenon remains poorly understood (4). In our case, the primary testicular tumor, histologically confirmed as a teratoma, exhibited spontaneous regression eleven years after metastasis. Due to the tumor's spontaneous regression, surgical resection was deemed sufficient, and chemotherapy was not administered.

Extragonadal germ cell tumors account for approximately 5% of all germ cell tumors and are believed to arise from residual embryonic cells. When a retroperitoneal lymph node containing germ cells is identified, the presence of metastatic lesions should be suspected, and testicular ultrasonography (USG) should be performed. While an obvious mass may be visible on testicular USG, it is important to note that findings consistent with fibrosis may also be present in burned-out tumors. Even if imaging findings are indicative of a burned-out or regressed tumor, an orchiectomy must be performed, as microscopic viable tumor foci may still be present. The standard treatment modality for burned-out tumors is cisplatin-based chemotherapy following radical orchiectomy (5). In our case, chemotherapy was not necessary due to the absence of viable invasive tumor areas in the pathological evaluation. The patient continues to be monitored for remission.

The concept of burned-out tumors was initially described in 1900. Although there are cases reported in the literature, it is generally observed that tumors are synchronous with metastases, and treatment is administered following excision. In our literature review, we did not find any cases in which the detection of the primary tumor was delayed by 11 years, eventually revealing a burned-out tumor. The pathophysiological mechanisms underlying this phenomenon remain unconfirmed. One proposed mechanism suggests an immune-related response of the host, where the host's T lymphocytes recognize tumor antigens after prolonged exposure, leading to inflammation and subsequent fibrosis. This hypothesis also explains why primary tumors often regress after metastasis. Another hypothesis posits that tumors reach a size where the blood supply becomes insufficient (6,7).

Additionally, the fact that our case declined imaging tests at the time of cervical lymph node excision makes it difficult to predict the disease's course. We do not have information regarding whether the patient had a testicular mass 11 years ago. However, it is unlikely that the primary origin of an extragonadal germ cell tumor is the cervical lymph node. Therefore, we believe that the burned-out phenomenon best explains the disease's course.

In conclusion, a testicular examination should be conducted in all tumors containing germ cells, regardless of their location, before labeling them as extragonadal. Moreover, there is a lack of studies comparing the prognosis of burned-out tumors with that of non-burned-out tumors. In this disease with an uncertain prognosis, the necessity and intensity of chemotherapy remain controversial, and more patient data are needed to establish standardized treatment approaches.

Conflict of interest
No conflict of interest was declared by the authors.

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