Olgu Sunumu / Case Report
Aegean J Med Sci 2019;2:77-80

'Burned-out' Seminomatous Testicular Tumor Diagnosed as Retroperitoneal Metastasis, A Rare Case

Retroperitoneal Metastaz ile Bulgu Veren Burned-Out Seminomatöz Testiküler Tümör, Nadir Bir Olgu

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ÖZET

"Burned-out testis tümörü" nadir görülen bir tümör türüdür. Burned out fenomeni, testisteki germ hücreli tümörün parsiyel ya da tamamen regrese olup, mediastinal, servikal, aksiller, retroperitoneal lenf nodları veya retroperitonal bölgeye metastaz ile bulgu vermesi olarak tanımlanır. Yapılan fizik muayenede testis muayenesi olağandır. 31 yaşında erkek hasta sırt ve karın ağrısı yakınması ile başvurdu. Yapılan fizik muayenesinde patolojik bulguya rastlanılmadı. Çekilen abdomen tomografide retroperitoneal kitle saptandı. Retroperitoneal kitle eksizyonu, paraaortik, parakaval ve retroperitoneal lenf nodu diseksiyonu uygulandı. Hastanın postoperatif dönemi komplikasyonsuz olup, postoperatif beşinci gün taburcu edilmiştir. Patoloji sonucu, seminom metastazı olarak raporlanmıştır. Retroperitoneal kitle ile karşımıza çıkan genç hastalarda burned out tümörlerin metastazları olabileceği akılda tutulmalıdır. Burned-out tümörlerin tedavi seçenekleri zamanla değişebilir. Farklı risk faktörlerini tanımlamak ve tedavinin optimize edilmesine yardımcı olabilmek için daha geniş olgu serileri yayınlanmalıdır.

Anahtar Kelimeler: burned-out, retroperitoneal kitle, lenf nodu diseksiyonu, orşiektomi

ABSTRACT

"Burned-out testicular tumor" is extremely rare. Burned-out phenomenon describes partially or completely regressed germ cell tumors of the testes diagnosed after metastases in several parts of the body; including the mediastinum, cervical and axillary lymph nodes, retroperitoneal lymph nodes, and the retroperitoneal region. Physical examination of the testicles reveals non-specific findings. A 31-year-old male presented with abdominal and back pain. The physical examination findings were unremarkable. Imaging with computerized tomography showed a retroperitoneal mass. Excision of the retroperitoneal mass and the paraaortic, paracaval, and retroperitoneal lymph nodes was performed. The postoperative period was uneventful and he was discharged on the postoperative 5thday. The postoperative pathological examination of the specimen reported a metastatic seminoma. Investigation of the underlying cause of a retroperitoneal mass in young adults requires including metastasis of burned-out germ cell tumors in the differential diagnosis. Further case reports on this rare type of tumors are required so that all potential risk factors will be defined and treatment protocols will be updated to optimize the management of the patients with burned-out germ cell tumors.

Keywords: burned-out, retroperitoneal mass, lymph node dissection, orchiectomy

Introduction

Burned-out phenomenon describes partially or completely regressed germ cell tumors of the testes diagnosed after metastases in several parts of the body are revealed; including the mediastinum, cervical and axillary lymph nodes, retroperitoneal lymph nodes, and the retroperitoneal region (1). Burned- out testicular tumors are unique and they are rarely encountered compared to primary extragonadal tumors (2). In this article, we aimed to report a 31-year-old male patient with a retroperitoneal mass, of which the postoperative pathological examination revealed a "burned-out" testicular tumor.

Case Presentation

A 34-year-old male patient was admitted to the general surgery department with abdominal and back pain, ongoing for three months. He had a history of left nephrectomy at the age of eight due to nephrolithiasis. He had no history of a chronic illness. A computerized

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Received/Geliş Tarihi: 14.03.2019 || **Accepted/Kabul Tarihi:** 23.04.2019

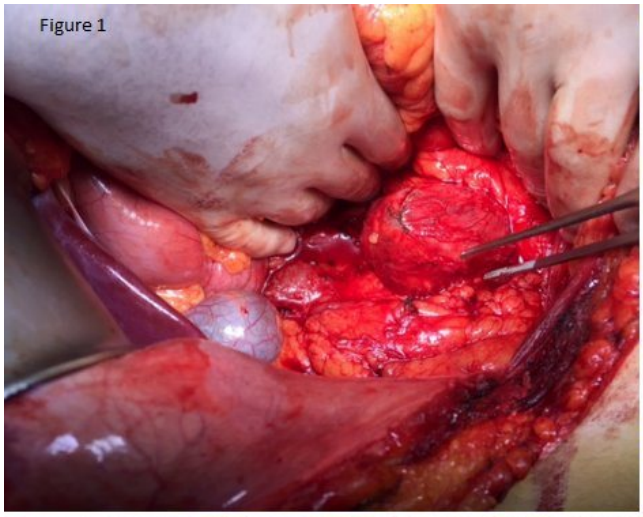
tomography (CT) imaging of the abdomen showed a 4.4x4.0 cm mass in the retroperitoneal region at the level of iliac bifurcation adjacent to the lower pole of the right kidney, vena cava inferior, and duodenum. Extensive lymphadenopathy was observed in the abdomen; in the paracaval, interaortacaval, and the external iliac lymph nodes on the right. A diagnostic biopsy was performed using the current interventional radiology techniques. A fine needle aspiration biopsy was performed, revealing findings compatible with malignancy, but no tumor diagnosis could be made due to the insufficient amount of tissue in the biopsy specimen. Positron emission tomography (PET/CT) was performed to identify the organ of origin of the retroperitoneal mass. The PET-CT imaging (SUVmax: 19.0) revealed similar findings to that obtained with CT. An increased 18-fluorodeoxyglucose uptake was observed in a mass lesion with a size of 4.4x4.0cm (SUVmax: 19.0). Increased uptake was also observed in the areas compatible with the location of the abdominal lymph nodes; including the paracaval, interaortacaval, and the external iliac lymph nodes on the right. An area of 1.8 cm in diameter with increased uptake of 18-fluorodeoxyglucose (FDG) was also observed in the abdomen at the level of the iliac bifurcation (SUVmax: 18.2).

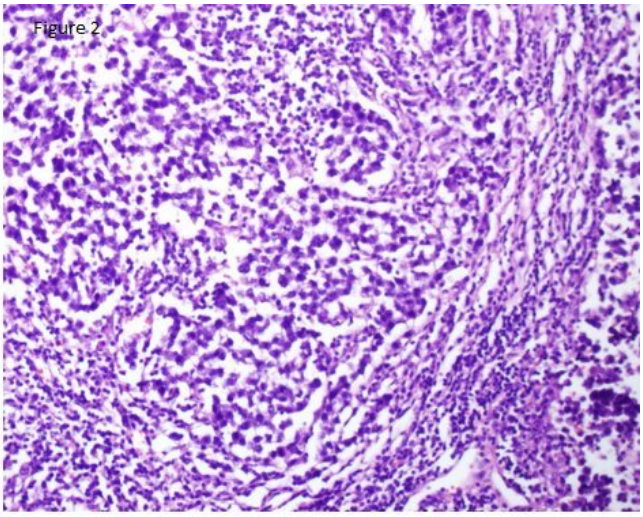
Figure 1a. *Peroperatively image of retroperitoneal mass*

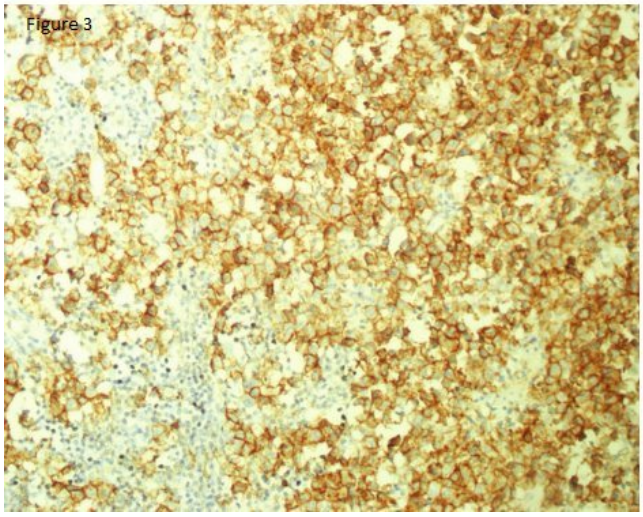
Figure 1b. *Tumor cell foci containing dense lymphocytic infiltrates separated by fibroses (HEx20)*

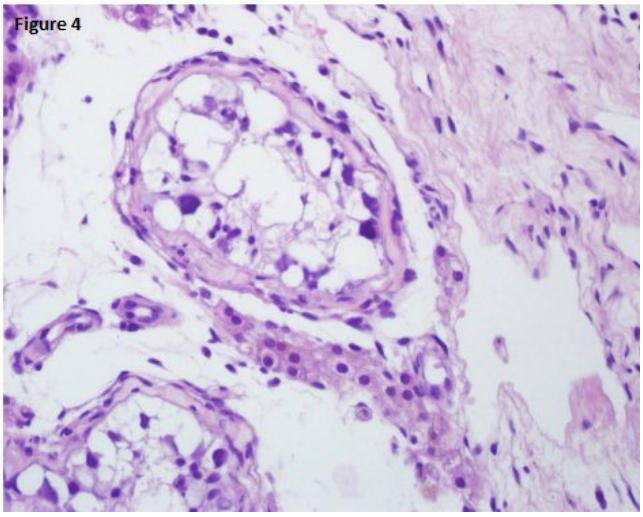
Figure 1c. *Immuhistocehmical examination of retroperitoneal mass, positive staining with CD 117,DABx20*

Figure 1d. *Seminal tubule with hyalinized basal membrane and intratubulergerm cell neoplasm, in orchiectomy material (HEx20)*









Operation was scheduled to identify the primary origin of the tumor so that treatment would be planned. Excision of the retroperitoneal mass and dissection of the paraaortic and paracaval lymph nodes were performed (Figure 1a). The findings in the postoperative period were non-specific.

The patient was discharged on the 6th postoperative day uneventfully.

Pathological examination of the specimen revealed the diagnosis of a seminoma. It was reported that six out of ten lymph nodes in the specimen showed metastases, containing alveolar fibrous septa and solid islands, consisting of atypical cells and tumor tissue (Figure 1b). Immunohistochemical studies on the tumor cells were positive for PLAP and CD117, infrequently positive for PANCK, infrequently positive for HCG, and negative for AFP (Figure 1c).

The patient was referred to the urology clinic and a right radical orchiectomy was performed. The pathological examination of the orchiectomy specimen showed atrophic tubular structures with thick basal membranes in the hyalinised stroma, focal mild lymphoplasmocytic infiltrates, and siderophages in the testicular parenchyma within an irregularly limited area with a diameter of 2.5 cm. Atrophic tubules, Sertoli cell only tubules and Leyding cell groups were observed, as well as tubular structures, showing active spermatogenesis in the testicular parenchyma. A few focal areas showed intratubular germ cell neoplasia. The patient was diagnosed with a 'seminoma' in the previous pathological report of the retroperitoneal mass, and when the previous and current reports of the pathological examination were evaluated together, it was reported that the findings were compatible with a regressed germ cell tumor of the testis (Figure 1d).

The postoperative course was unremarkable. The patient was discharged and adjuvant chemotherapy was recommended for treatment. Written informed consent was obtained from the patient for the publication of this case report and any associated images.

Discussion

"Burned-out testicular tumor" is a very rare type of tumor and unique with differences from primary gonadal germ cell tumors. “Burned-out” phenomenon indicates a partially or completely regressed germ cell tumor of the testes, diagnosed after metastasising to the mediastinum, cervical and axillary lymph nodes, retroperitoneal lymph nodes, or the retroperitoneal region (1). Studies on the immunological mechanisms suggest that malignant germ cells, evading from the immune system in the testes are detected by the immune system via antigen-presenting cells after metastasising to the lymph nodes. Then, the primary tumor is removed from the organ of origin by an adaptive immunoreaction (3, 4).

Clinically, no symptoms may be available or no findings may be observed in the physical examination of the testicles (2). Patients may have complaints of abdominal pain, back pain, due to the compression by the mass in the retroperitoneal region. Metastatic burned-out extragonadal germ cell tumors of the testes have a better prognosis and higher survival rates compared to primary retroperitoneal malignancies (4, 5). Differential diagnosis of a retroperitoneal mass should include germ cell tumors. The primary tumor is usually located on the same side with a retroperitoneal metastatic lymph node (5, 6). Chemotherapy is more effective for the treatment of ''burned-out'' non-seminomatous germ cell tumors compared to the primary testicular tumors (7). A testicle biopsy may only show intratubular germ cells even in patients with metastasis. It is recommended to remove the testicle when the neoplasm is detected (8). In order to ensure a complete cure for patients, orchiectomy is recommended before the start of chemotherapy (6). In the literature, there are two studies available about this rare disease. Despite the small number of included patients, these studies reported that there was no need to perform surgery if the testes were normal (9, 10).

Conclusion

Although they are rare, “burned-out” germ cell tumours should be remembered in a patient presenting with a retroperitoneal or mediastinal mass. For the treatment, orchiectomy is recommended before starting the systemic chemotherapy, however available studies in the literature reported that there was no need to perform surgery if the testes were normal. However, these studies included only a few patients. Therefore, larger case series are required to develop treatment guidelines and to better address the issues associated with this disease.

Yazarlar arasında çıkar çatışması yoktur.

The author declares no conflict of interest.

Finansal Destek: yoktur / Funding : none

This case report has been presented as a poster presentation in National Surgical Congress, in 2018, Antalya, Turkey

doi: https://doi.org/10.33713/egetbd.539862

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