

Burned-out germ cell tumor presenting with acute abdomen

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ABSTRACT

Acute abdomen is a serious condition frequently encountered in the emergency departments (ED). There are various etiologies causing acute abdomen, most common being acute appendicitis; however, there are rare causes of acute abdomen as well and one should keep them in mind while handling a patient with unusual clinical features. We herein present a 26-year-old male, with no past medical or surgical history, presenting with acute abdominal pain and distension to the ED. He was found to have a large vascular retroperitoneal mass on computed tomography which had invaded and perforated the duodenum thus causing the acute presentation. Repair of the duodenal perforation and sampling of the mass were performed in terms of surgical management. Pathology results revealed the mass originating from a burned out testis yolk sac tumor with embryonal carcinoma component. Although the original tumor had regressed at the testis, its metastasis at the retroperitoneal area had caused the clinical condition. This entity is described as the retroperitoneal metastasis of a burned-out testicular tumor. Few cases presenting with gastrointestinal bleeding secondary to invasion of the retroperitoneal metastasis have been reported. However, this is the first case in the literature presenting with duodenal perforation and acute abdomen. Sampling of the retroperitoneal tumor for histopathological diagnosis during the immediate surgical intervention facilitates the diagnostic management in these cases. Although scrotal examination combined with testis tumor marker assessments is essential for optimal patient management, the possibility of a burned-out testicular tumor with normal scrotal examination should always be kept in mind.

Keywords: Acute abdomen; intestinal perforation; neoplasm invasion; retroperitoneal neoplasms; testicular neoplasms.

INTRODUCTION

Testicular tumor is the most common solid tumor in men aged between 15 and 35 years and accounts for approximately 1% of tumors detected in men.^[1] The typical finding of testicular tumors is a solid testicular mass with hard consistency. Apart from this, there may be complaints related to metastasis.^[1] Testicular cancers usually metastasize through hematogenous and lymphatic drainage.^[2] Although retroperitoneal metastasis and gastrointestinal involvement of testicular cancers are rare, they may lead to complications such as intestinal obstruction, intestinal perforation, and hemorrhage.^[3] Acute abdomen has many causes; however, metastatic testicular

cancer presenting as an acute abdomen is very rare.^[4] In this study, we present a case of duodenal perforation secondary to duodenal invasion of a burned-out testis tumor.

CASE REPORT

A 26-year-old male patient presented to the emergency department (ED) with a 2-day history of abdominal distension, pain, nausea, and vomiting. He had no history of recent travel or infectious disease. Physical examination revealed 4-quadrant abdominal rigidity with rebound tenderness and minimal abdominal distension. Bowel sounds were absent. There were no abdominal signs of previous surgery nor evidence

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for abdominal wall hernias. Scrotal examination was unremarkable. The complete blood count revealed a hemoglobin value of 8.3 g/dL and normal white blood cell count. Blood biochemistry results were within normal ranges. Abdominal ultrasound revealed a 5×4 cm para-aortic solid lesion located at the level of the iliac bifurcation. Computed tomography (CT) subsequently revealed a 5×6 cm mass occupying the area between the inferior border of the pancreas and the iliac bifurcation in the midline. There was long segmental wall thickening of the duodenal and proximal jejunal loops. Periduodenal inflammation and edema of the adjacent peritoneal surfaces were observed as pockets of dense fluid collections and contrast extravasation at infra-hepatic recesses. There was abdominal free air suggestive of a viscus perforation. The center of the mass was hypodense with a necrotic core (Fig. 1). Emergency laparotomy was performed with a preliminary diagnosis of retroperitoneal mass and viscus perforation.

A nasogastric tube and Foley catheter was inserted, followed by empiric intravenous antibiotics. Midline laparotomy revealed purulent free fluid in the abdomen and pelvis. There were retroperitoneal and para-aortic necrotic masses. The

dominant mass in the retroperitoneum invaded the third portion of the duodenum, encircling the superior mesenteric artery. Invasion of this mass led to duodenal perforation. Necrotic and infected tissues at the perforation site were debrided and excised. The excised and tumor free edges of the duodenum were dissected free from the retroperitoneum, and the 3 cm defect was primarily repaired using 2–0 Vicryl sutures in an interrupted fashion. Since there was a regional invasion of abdominal aorta at the level of iliac bifurcation, a vascular surgeon was consulted intraoperatively and the mass was deemed unresectable. The tumor was sampled, an abdominal drain was placed and the abdominal layers were closed leaving the large para-aortic mass *in situ*.

The post-operative course was uneventful and the patient was discharged home on the 9th post-operative day.

Pathology of the tumor biopsies revealed a mixed germ cell tumor which comprised for 90% of yolk sac and 10% embryonal carcinoma (Fig. 2). The tumor cells expressed alpha-fetoprotein (AFP) within the yolk sac tumor cells, whereas the embryonal carcinoma component was identified with cytok-

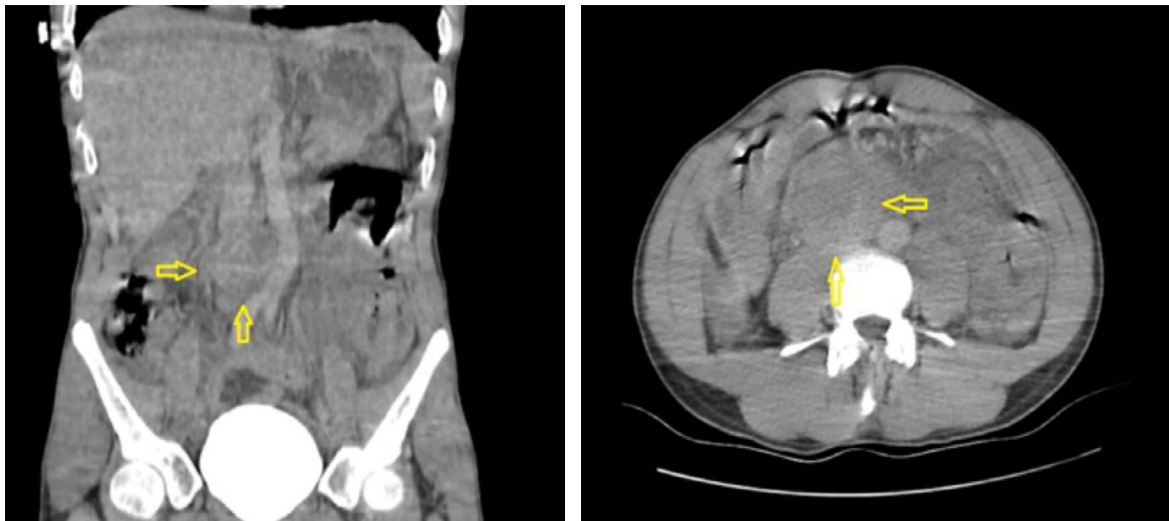


Figure 1. Pre-operative computed tomography images of the abdominal mass. Arrows: Borders of the tumoral mass.

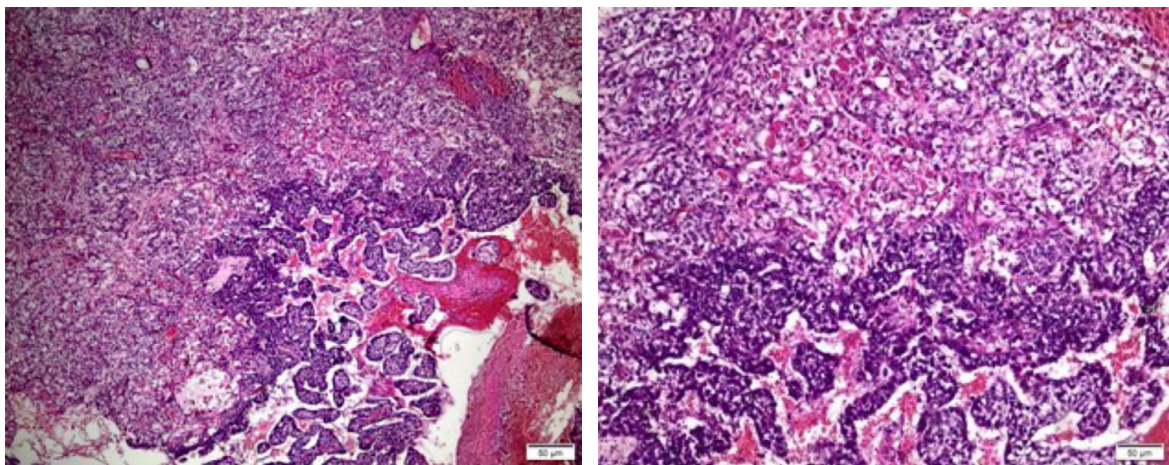


Figure 2. Embryonal carcinoma and Yolk Sac tumor cells observed in the retroperitoneal mass (Hematoxylin and Eosin, ×40).

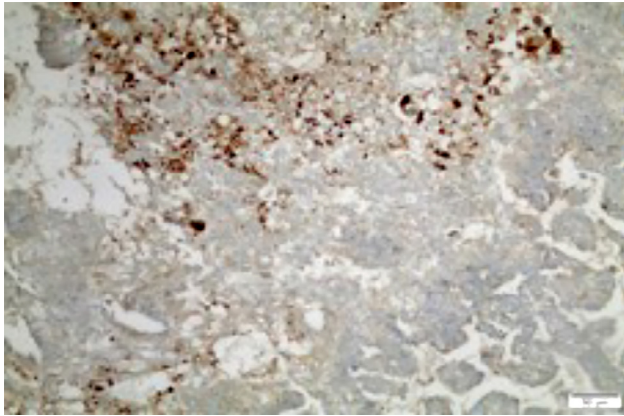


Figure 3. AFP expression of Yolk Sac tumor cells in the retroperitoneal mass (magnification $\times 100$).

eratin 7 and CD30 expressions (Figs. 3 and 4). It was stated that the tumor invaded the serosa and muscular layers of the duodenum leaving the mucosal layer intact.

Once the pathological diagnosis was confirmed, scrotal physical examination was repeated and was found to be

Table I. Staging of testis tumors by the American Joint Committee on cancer

Stage	Tumor	Node	Metastasis	Serum Markers
0	pT _{is}	N ₀	M ₀	S ₀
1	pT ^I - pT ₄	N ₀	M ₀	S ₀
2	Any T	N ₁₋₃	M ₀	S ₁
3				
3A	Any T	Any N	M _{1a}	S ₀₋₁
3B	Any T	Any N	M _{0-1a}	S ₂
3C	Any T	Any N	M _{1a-1b}	S ₃

S: Serum tumor marker levels; M_{1a}: Nonregional nodal or pulmonary metastasis; M_{1b}: Nonpulmonary visceral metastasis.

normal again. The patient's (AFP) level was 2544 (reference value: 0–9 $\mu\text{g/dL}$), human chorionic gonadotropin (hCG) level was 45.37 (reference value: 0–5 U/L) and lactate dehydrogenase (LDH) value was 347 (reference value: 25–248 U/L). Scrotal ultrasonography did not demonstrate any tes-

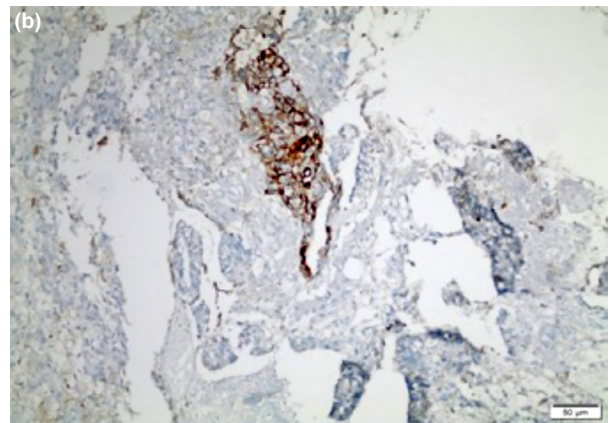
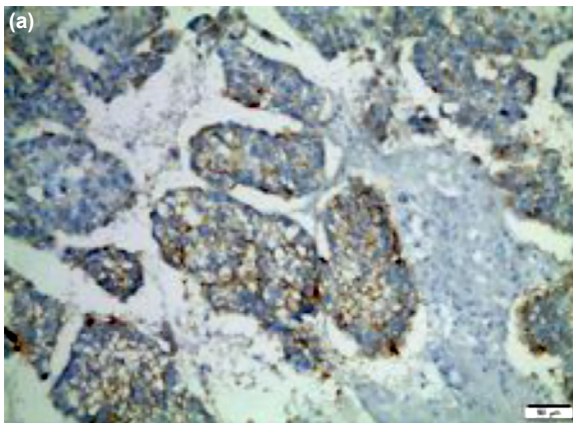


Figure 4. (a) CD30 expression within the embryonal carcinoma component of the retroperitoneal mass (magnification $\times 100$). (b) Cytokeratin 7 expression within the embryonal carcinoma component of the retroperitoneal mass (magnification $\times 100$).

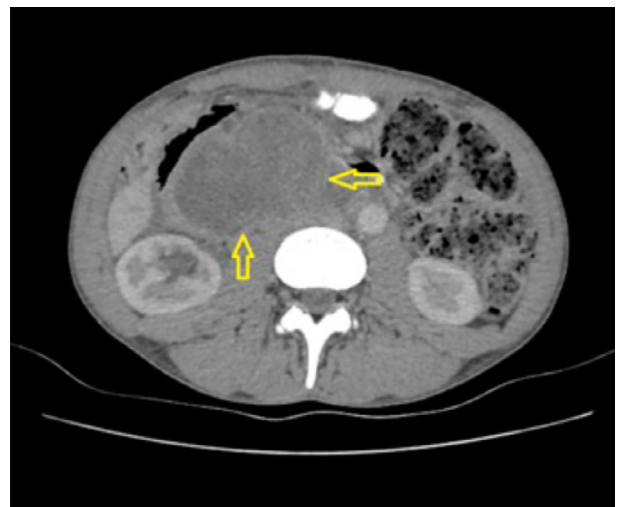
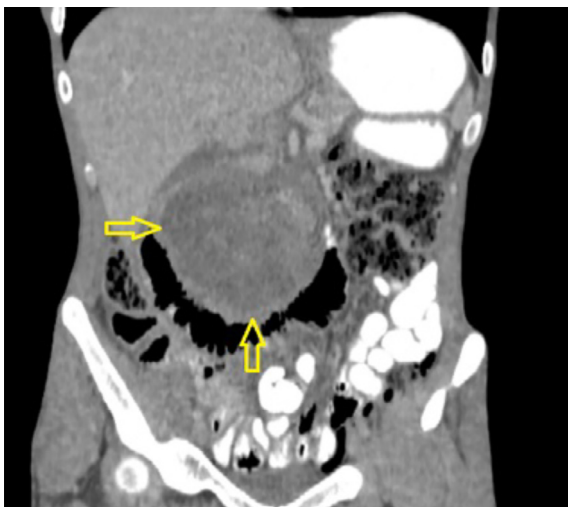


Figure 5. Computed tomography images of the rapidly growing tumor immediately before the BEP treatment. Arrows: Borders of the tumoral mass.

ticular lesions, but coarse calcifications were observed in the right testis.

The patient refused a biopsy from the right testis and was subsequently classified as Stage I in terms of serum tumor markers (SI) and MIA for the non-regional metastasis. According to the staging of testis tumors by the American Joint Committee on Cancer, classified as stage 3B testicular cancer (Table 1).^[5] Four cycles of Bleomycin, Etoposide, and Cisplatin (BEP) chemotherapy protocol were planned.

Immediately before initiating chemotherapy, thorax and abdomen CT scan was performed. This revealed a massive increase in the dimensions of the retroperitoneal mass in the para-aortic area reaching up to 11×7 cm (Fig. 5). In addition, hydronephrosis of the right kidney was detected secondary to entrapment of the right ureter by the mass. No additional metastases were observed.

Patient received four cycles of BEP chemotherapy on which complete regression of the tumor and resolution of the right-sided hydronephrosis was observed on follow-up CT. The AFP, hCG, and LDH values returned to normal ranges. Further follow-up with CT and tumor markers at 3-month intervals did not reveal any tumor recurrence at 1 year. Periodic follow-up of the patient with tumor marker measurements and thoracoabdominopelvic CT scans were planned. A template retroperitoneal lymph node dissection will be planned in the case that marker levels increase and/or there is radiological suspicion of recurrence.

DISCUSSION

Testicular cancer is encountered in 1% of men and is frequently encountered between the ages of 15 and 35.^[1] Non-seminomatous testicular tumors are less common than seminomatous testicular tumors and mostly contain more than one cell type. The prognosis of mixed germ cell tumors depends on the structural components of the tumor. Patients are divided into three different groups according to tumor markers and metastasis in terms of prognosis. Treatment is determined according to these groups. Our patient's current findings are in the medium prognosis group and the 5-year progression-free survival is 75%.^[6]

Germ cell tumors of the testis have the potential to metastasize to various sites in the body.^[7] In an autopsy study conducted by Johnson et al., the frequency order of testicular cancer metastasis sites was the lung, followed by retroperitoneal lymph nodes, liver, mediastinal lymph nodes, brain, kidneys, gastrointestinal system, bone, adrenal gland, peritoneum, and spleen.^[7] In a study by Chait et al.,^[7] germ cell testicular tumors were observed to metastasize to the gastrointestinal tract at a rate of 5%, and similar to the presented case, these tumors were histopathologically non-seminomatous germ cell tumors. Metastasis to the gastroin-

testinal tract mostly occurs as a direct invasion from the retroperitoneal lymph nodes where lymphatic drainage of the testicle occurs.^[2-7] Jejunum and ileum are more commonly invaded by retroperitoneal metastatic masses due to their retroperitoneal location.^[2] However, isolated duodenal involvement is extremely rare.

Testicular tumors presenting with gastrointestinal involvement may exhibit symptoms such as low back pain, abdominal pain, and gastrointestinal hemorrhage. Furthermore, they can present with symptoms associated with anemia, intestinal obstruction, or viscus perforation.^[3-11] While the patients presenting with pain or anemia are diagnosed during the search regarding the underlying causes of these symptoms, patients presenting with intestinal obstruction or perforation are typically diagnosed at emergency surgical exploration as in our case.

Moicinovic and Abaza^[9] reported a 45-year-old male with 14 cm seminomatous tumor of the right testis who presented to the ED with acute abdomen. This patient, similar to ours, underwent emergent surgical exploration and found to have intestinal perforation due to retroperitoneal lymph node invasion to the duodenum. Senadhi and Dutta,^[2] on the other hand, reported a patient who presented with intestinal hemorrhage and underwent surgical treatment after failed endoscopic interventions. However, when these cases are reviewed that these patients had a diagnosis of testicular tumor in their past medical history and/or complained about palpable scrotal masses during their initial presentation. In our case, as a rare clinical condition, a testicular solid mass was not detected. This condition is called burned-out testicular cancer and refers to complete regression of testicular tumor without any treatment. It is known that burned-out testicular cancer may reveal itself with metastases.^[12] Focal areas with increased echogenicity due to calcifications and fibrosis can be observed in scrotal ultrasonography of these patients. In the case that, a testicular malignancy is suspected in these patients, testicular biopsies should be performed.^[9] Unfortunately, we could not perform a testicular biopsy in our case for lack of consent. However, we continued to follow our patient with periodic imaging and tumor markers.

The success of chemotherapy regimens in the treatment of metastatic non-seminomatous germ cell testicular tumors has been demonstrated.^[9] However, it should be noted that emergent surgery may be necessary for treatment of complications such as intestinal perforation and obstruction in patients with gastrointestinal involvement. In case of an urgent surgery, the intervention should focus on treating the surgical emergency and sampling for histopathological diagnosis rather than resecting the entire tumor in cases with unresectable masses. Definitive treatment can be planned postoperatively based on pathology reports and tumor stage.

Conclusion

Gastrointestinal system involvement is rare in testicular tumors and symptoms such as abdominal pain and gastrointestinal hemorrhage may develop. In extremely rare cases, gastrointestinal perforation may occur due to mass invasion. In young male patients undergoing surgical exploration due to intestinal perforation by invasion of a tumor, retroperitoneal metastasis of a testicular tumor should be considered. Sampling of the retroperitoneal tumor for histopathological diagnosis during the immediate surgical intervention for primary repair facilitates the diagnostic management in these cases. Although scrotal examination combined with testis tumor marker assessments is essential for optimal patient management, the possibility of a burned-out testicular tumor should always be kept in mind.

Patient Perspective

“It was shocking for me to figure out that my severe belly pain was due to a tumor stemming from my testis. I am happy with the results of the surgery and chemotherapy. I hope my case will assist the surgeons and physicians in the world.”

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OLGU SUNUMU - ÖZ

Akut batın ile başvuran burned-out germ hücreli tümör olgusu

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Akut karın, acil serviste sıklıkla karşılaşılan ciddi bir durumdur. Akut karına neden olan çeşitli etiyolojiler vardır, en sık görülen akut apandisitir, ancak nadir akut karın nedenleri de vardır ve olağandışı klinik özelliklere sahip bir hastayla ilgilenirken bunları akılda tutmak gerekir. Bu yazıda, akut karın ağrısı ve distansiyon ile acil servise başvuran, 26 yaşında bir erkek hastayı sunuyoruz. Bilgisayarlı tomografide duodenumu invaze ederek perforasyon etmiş ve akut karına neden olan büyük bir vasküler retroperitoneal kitlesi olduğu bulundu. Cerrahi tedavi olarak, duodenal perforasyon onarımı ve kitle örneklenmesi uygulandı. Patoloji sonucu testis kaynaklı embriyonal karsinom bileşenli yolk sac tümörü olarak raporlandı. Orijinal tümör testiste tamamen gerilemiş olmasına rağmen, retroperitoneal bölgedeki metastaz klinik duruma neden olmuştu. Bu antite, regrese olmuş bir testis tümörünün retroperitoneal metastazı olarak tanımlanır. Retroperitoneal metastaz invazyonuna sekonder gastrointestinal kanama ile başvuran az sayıda olgu bildirilmiştir. Bildirdiğimiz olgu literatürde duodenal perforasyon ve akut karın ile prezente olan ilk olgudur. Acil cerrahi müdahale sırasında histopatolojik tanı için retroperitoneal tümörün örneklenmesi, bu olgularda tanıyı kolaylaştırır. Testis tümörü belirteçleri ile kombine skrotal muayene hasta yönetimi için gerekli olmasına rağmen, normal skrotal muayene mevcudiyetinde testis tümörünün tam regrese olma olasılığı akılda tutulmalıdır.

Anahtar sözcükler: Akut karın; intestinal perforasyon; retroperitoneal tümörler; testis tümörleri.

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