THE FIRST REPORTED CASE OF A PALPABLE AND HYPOECHOIC BURNED OUT TESTICULAR TUMOR

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ABSTRACT

Primary extra gonadal germ cell tumors are rare.In case of the diagnosis of a midline retroperitoneal mass in a male, germ cell tumors should be taken into consideration. It is commonly accepted to consider retroperitoneal germ cell tumors as the metastasis of a viable or burned out testicular tumor. In such casesorchiectomy should be performed since the burned out site in the testis could continue to harbor malignancy despite systemic chemotherapy.A 45-year-old man presented as an outpatient with complaints of back pain. He was diagnosed with a retroperitoneal mass and a palpable testis mass. Due to increased serum tumor markers and the testicular mass, he underwent radical orchiectomy. Pathological study revealed a burned out tumor.Followingchemotherapy, retroperitoneal lymph node dissection(RPLND)was performed and despite the residual and enhanced retroperitoneal lymph nodes in his CT scan, none of the resected lymph nodes showed a viable tumor.However and despite the rising number of reports onthis disease, our case had adifferent presentation both at initial diagnosis and during follow up after chemotherapy. This is the first reported case of a palpable and hypoechoic burned out testicular tumor.

KEY WORDS: testis, burned out tumor, lymphadenectomy, tumor

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INTRODUCTION

As previously described in the literature, primary extra gonadal germ cell tumors are rare[1-3]. In case of diagnosing a retroperitoneal midline mass in a male patient, germ cell tumors (GCT) should be considered and due to their rare primary nature, investigation with the aim of finding other primary sites should be performed[1,4,5]. It is commonly accepted to consider germ cell tumors of the retroperitoneum asmetastasis of a viable or burned out testicular tumor[1-4]. Systemic chemotherapy is the most widely used treatment for extra gonadal tumors[3, 4]. Burned out testicularmass or Azzopardi tumor is a known condition which has retoperitonealmetastasis but the primary tumor regresses and only small foci of calcification remain as signs of its presence[5-8]. Therefore, the diagnosis and treatment of these cases may be delayed due to the nonspecific initial manifestations and regression of the primary lesion.

Orchiectomy is the treatment of choice in such cases since the burned out site in the testiscould continue to harbor malignancy despite systemic chemotherapy[3-5]. Herein, we report a middle-aged man who was diagnosed with a retroperitoneal mass and had evidence of a burned out tumor in ultrasound study. Hepartially responded to chemotherapy and underwent retroperitoneal lymph node dissection(RPLND).

CASE PRESENTATION

A 45-year-old man, married withtwochildren, computer engineer, 40 pack year smoker, with a history of right foot cellulitis since one month before, presented as an outpatient with complaints of back pain over the previous 3 months. The pain was mild, dull, without radiation to other sites, non-colicky and non-positional. He did not mention any other symptoms such as weight loss or urinary symptoms.

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Physical examination revealed a small firm area at the upper pole of the right testis.

In ultrasound study of the kidneys and bladder, a 107*80mm exophytic mass was diagnosed neighboring the middle part of the right kidney and proximal to the right ureter with compression effects on the right pyelocalyceal system, causing mild right hydronephrosis. Inscrotal ultrasound study, a few hypoechoic masses in the right testis with a maximum size of 15*8mm were reported.

Laboratory data revealed normal renal function and his creatinine level was 1.1 mg/dL. Tumor marker tests were abnormal with the following results:â-human chorionic gonadotropin (âHCG) level was 510(normal range in non-pregnant women<20), lactic dehydrogenase (LDH) was 1248(normal range <480) and á-fetoprotein (AFP) was 0.1(normal range <10).

In the next step, pre-operative spiral abdominopelvic CT scan with and without IV contrast was performed. CT showed a 100*80mm enhancing mass between the aorta and the right kidney with compression effect on the right kidney. Thrombosis inside the IVC, below the mass and the perinephric lymph node was also reported (Figure 1).

He underwent radical orchiectomy and right DJ insertion. Pathology showed two foci of scar of tumor without evidence of viable tumoral cells (Burned out tumor/Azzopardi tumor);and Intratubular germ cell neoplasia(ITGCN)was not identified. One of the scar tissues was indicative of a rete testis.

Tumor markers were rechecked after surgery and the results were as follows: LDH: 1180, âHCG: 277 and AFP: 0.6. Chest CT scan was also performed which was normal.

According to the pathology lab result confirming the diagnosis of a metastatic germ cell tumor, chemotherapy was initiated and 4 cycles of BEP (bleomycin, etoposide, cisplatin) regimen, 6 sessions at each cycle was given. By the end of chemotherapy, the abdomenopelvicCT scan with IV contrast showed a 58*40mm mass with enhancement, IVC invasion and compression effect on the right renal artery and right renal pelvis. CT also showed thrombosis inside the IVC which was extended to both the common iliac veins (Figure 2).

Color Dopplerultrasoundof the veins showed complete thrombosis of the IVC up to the level of the hepatic vein entrance and thrombosis of the right renal vein.

At the end of the chemotherapy course, tumor markers became normal:âHCG: 0.9, AFP: 3.1 and LDH: 327.At this stage, he was admitted and anticoagulation therapy with IV heparin was started.

On the ninth admission day he underwent RPLND at our center. Duringsurgery multiple adhesive lymph nodes were dissected from the paraaortic and paracaval areas. Pathology reported extensive necrosis with no evidence of malignancy.

The retroperitoneal fluid was sent for cytologic examination and the result was negative for malignant cells. Today, the patient is in well health and is living with no complaints or complications.

DISCUSSION

Primary extra gonadal GCT is rare and accounts for approximately 5% of germ cell tumors (3).Patients with metastatic GCT in there troperitoneum are commonly divided into 3 categories. Two third have evidence of a testicular tumor in the form of ITGCN in the testis or evidence of burned out tumor at imaging whereasone third of such cases truly and primarily arise from the retroperitoneal origin(3).

When a retroperitoneal mass is diagnosed in adults or adolescents, we should always consider GCT as an important differential diagnosis, especially in the absence of an imaging clue for its origin(3-5), in order to avoid misdiagnosis and to apply the best treatment schedule for the patient.

However, in the presence of elevated tumor markers like âHCG and AFP, treatment by chemotherapy can be initiated and no further evaluation is warranted as the diagnosis of extra gonadal GCT is definite(3).Furthermore, in the presence of ultrasound evidence of a burned out tumor or when the pattern of lymph node involvement is consistent with a right or left side testicular tumor, inguinal orchiectomy should be performed during the treatment course (3).

Nevertheless, our case had a different presentation both at initial diagnosis and during the follow up after chemotherapy. The presence of a palpable and hypoechoic burned out tumor has not been reported in previous case reports. This finding could be a sign of a previous huge tumor which was burned out, but an area of hypoechoic lesion with a diameter of 2 cm had remained. It shows that irrespective of the size of the primary tumor, in case of a metastatic burned out mass, the original tumor cells will regress and die. This once again brings up the hypothesis that these kinds of testicular masses may be associated with different genetic abnormalities which can lead to their different behaviors. Further investigation in this field might lead

to better understanding of such tumor cells biology besides enlightening the path tonovel treatment options.

Another interesting finding in our case was related to the imaging findings after chemotherapy and the surgical pathology report. Although CT scan with IV contrast is not the standard imaging for evaluating post chemotherapy activity of retroperitoneal lymph nodes, but enhancement of these nodes would be suggestive of tumor viability. As we described, the pathologic report of the resected lymph node, revealed a necrotic and nonviable tumor. Our explanation for this finding would be the fact that previous neovascularization in theretroperitoneum might have remained intact after chemotherapy while the tumor cells all died during treatment, resulting in the enhanced tumoral residue.

CONCLUSION

Burned out tumor or azzopardi tumor is still one of the challenging cases in the field of germ cell tumors. The exact histopathology of the tumor is unknown in some cases and we are unable to explain the reason for the differences in their behavior. However, with the rising number of reports about this disease, further research Is anticipated so that with determining the exactetiology of tumor regression, we might be able to trigger tumor suppression at metastatic sites.

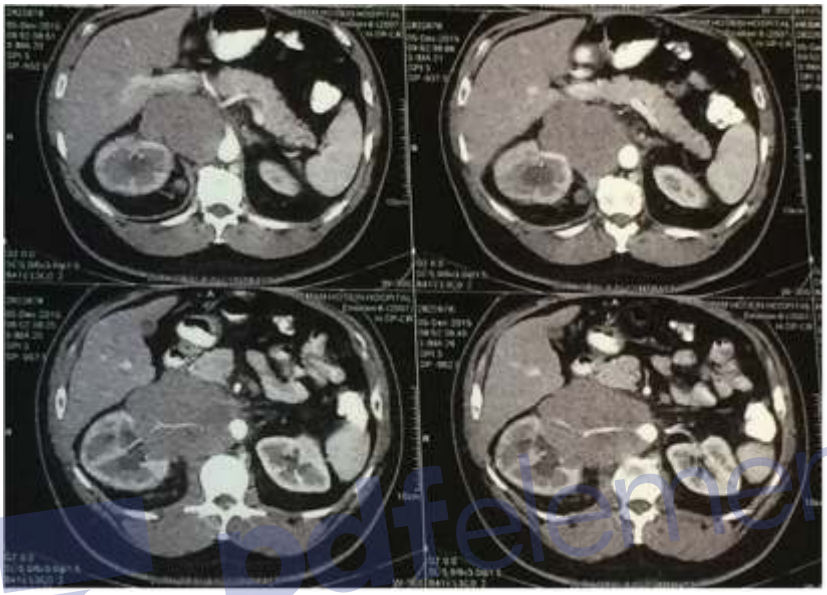

Figure1: CT scan images before chemotherapy show thrombosis inside the IVC, below the mass and perinephric lymph node

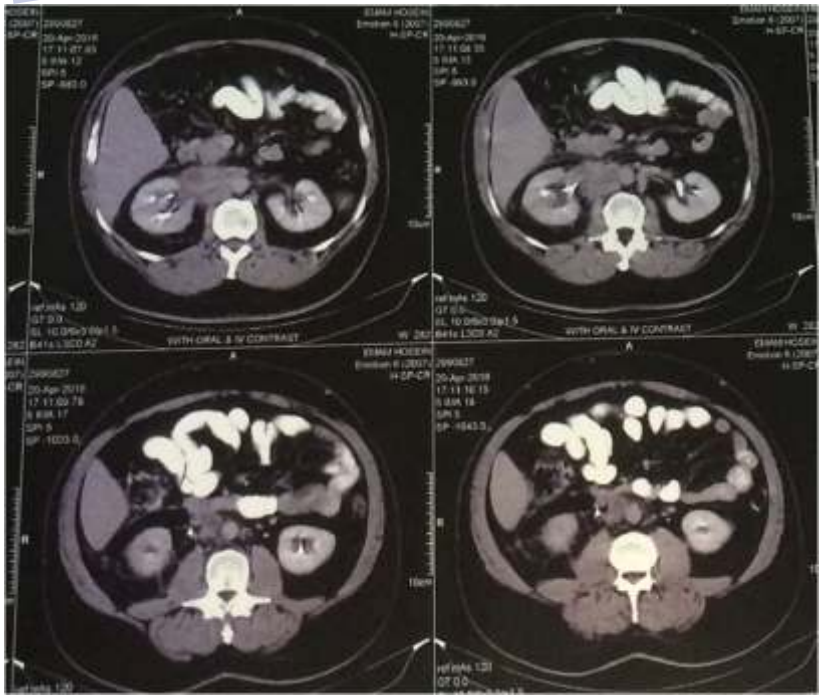

Figure 2: CT scan after chemotherapy also showed thrombosis inside the IVC, extending to both the common iliac veins

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