Retroperitoneal Seminoma with the 'Burned out' Phenomenon in the Testis

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The rare 'burned out' phenomenon in germ cell tumors is known as the presence of an extragonadal germ cell tumor with a spontaneously regressed testicular tumor found in common metastatic sites, including the retroperitoneal, mediastinal, supraclavicular, cervical, and axillary lymph nodes; lung; and liver. We report a patient who presented with a retroperitoneal extragonadal germ cell tumor with a spontaneously regressed testicular tumor. **(Korean J Urol 2009;50:516-519)**

Key Words: Seminoma, Retroperitoneal neoplasms, Testicular neoplasms

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The 'burned out' phenomenon in germ cell tumors refers to a germ cell tumor in extragonadal tissues with spontaneous regression of an intragonadal tumor. Extragonadal germ cell tumors are usually found in the retroperitoneal, supraclavicular, cervical, and axillary lymph nodes and occasionally are found in the lung and liver. There are 2 theories to explain this phenomenon: one is spontaneous regression of a primary germ cell tumor after metastasis of the germ cell tumor, and the other is the arising of a primary germ cell tumor in extragonadal tissues. However, no consensus exists about the origin of extragonadal germ cell tumors.

We have experienced no intragonadal germ cell tumors except for metastatic retroperitoneal germ cell tumors in patients with testicular atrophy.

CASE REOPRT

A 23-year-old Russian man visited our hospital complaining

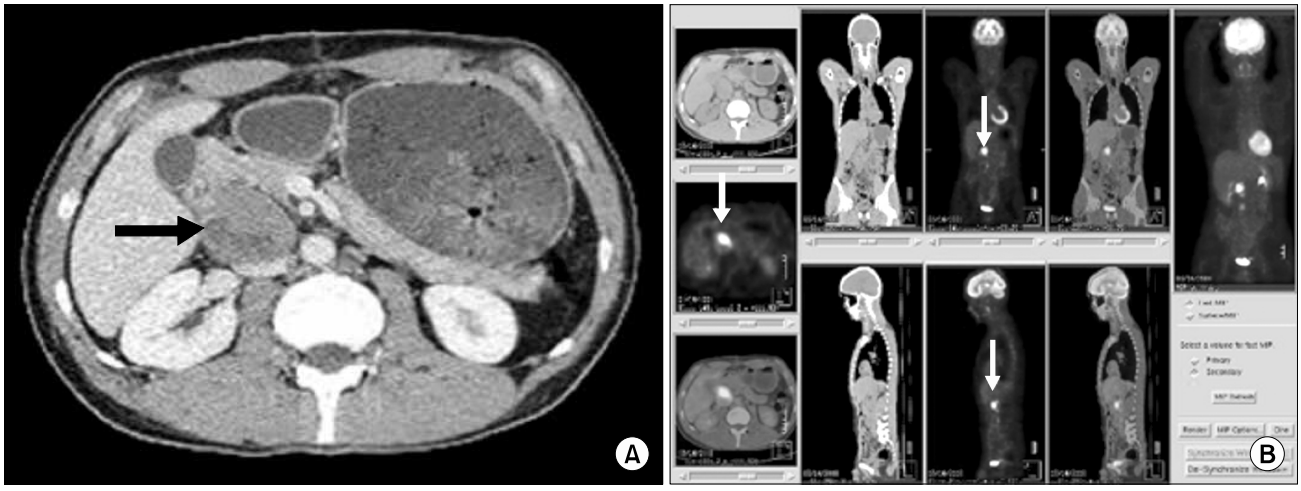

Fig. 1. (A) Computed tomography (CT) of the retroperitoneum shows an enhanced mass (black arrow) with a lobulated margin and necrotic portion located between the pancreatic head and the inferior vena cava. (B) Proton emission tomography (PET) shows 2 hypermetabolic masses between the duodenum and the inferior vena cava (white arrow).

of abdominal pain. A contrast-enhanced mass sized 37x46 mm with a leaflike margin was found on abdominal computed tomography (CT) between the pancreatic head and inferior vena cava, with a suspected necrotic lesion at the posterior part of the tumor and no other abnormalities. Another contrast-enhanced mass sized 38x26 mm that included a necrotic lesion was found anterior to the inferior vena cava between the origin of the inferior mesenteric artery and the bifurcation of the abdominal aorta. There were no abnormal findings on a physical examination and photon emission tomography (PET) except that the tumor that was found in the prior CT appeared to be a hypermetabolic mass (Fig. 1). Complete excision of the tumor, which was located between the abdominal inferior vena cava and the aorta, was done without adhesion. A metastatic seminoma was revealed pathologically (Fig. 2). We then examined testis tumor markers, performed testis ultrasonography, and performed pelvic magnetic resonance imaging (MRI). The results of tests such as serum human chorionic gonadotropin (hCG), lactate dehydrogenase (LDH), and alpha-fetoprotein were in the normal range, and ultrasonography showed a localized inhomogeneous mass with microlithiasis and atrophy in the right testis. T2-weighted MRI showed a 33 mm mass with an ill-defined margin, and this mass was revealed as an inhomogeneous, unenhanced low signal on contrast-enhanced MRI (Fig. 3). In the MRI images, the testicular mass and the surrounding tissue were well differentiated without adhesion, hyaline fibrous tissue with calcification replaced normal testis, and there was atrophied testis tissue around the mass. In microscopy, intraepithelial malignant cell neoplasia was found (Fig. 4). We finally diagnosed T0N2M0S0 with stage IIB. We scheduled postoperative chemotherapy for the patient, but he had returned to his country and we did not carry out the schedule.

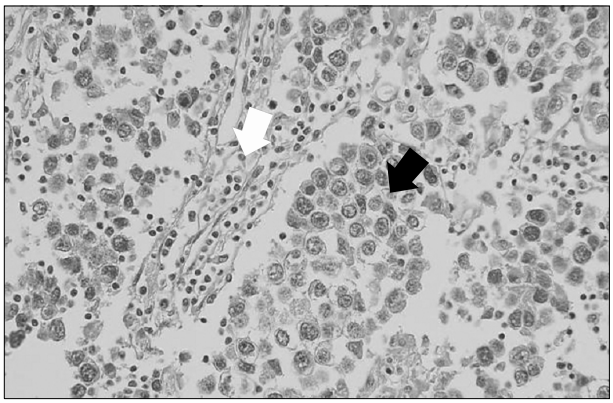

Fig. 2. Microscopic finding of a retroperitoneal mass shows seminoma cells with enlarged nucleoli in mitosis (black arrow) and many lymphocytes in the lobular septum (white arrow) (H&E, x400).

DISCUSSION

Extragonadal chorionic carcinoma with scar reaction in the testis was reported in 1927 for the first time by Prym.[1] Since then, extragonadal germ cell tumors with the burned out phenomenon have only rarely been reported throughout the world, and until now, no case had been reported in Korea. Of 250 testis cancer patients, Fabre et al[2] reported that 5 patients, which is 2%, had this phenomenon and 2 patients had a

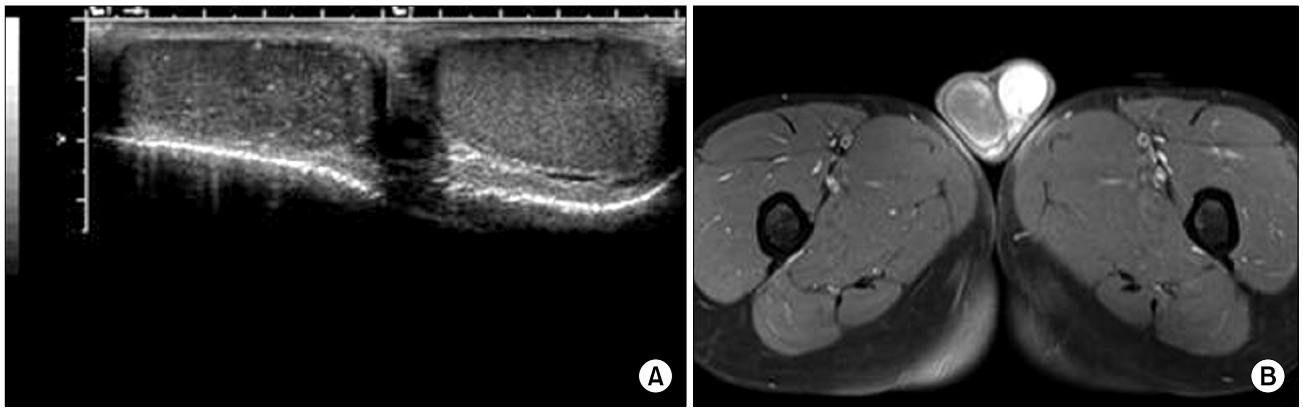

Fig. 3. (A) Ultrasonography of testis shows a segmental inhomogeneous lesion containing calcification in the atrophied right testis. (B) Magnetic resonance imaging (MRI) of testis shows a 33 mm ill-defined mass within the right testis that was not enhanced well and had heterogeneous low signal intensity on T2-weighted MRI.

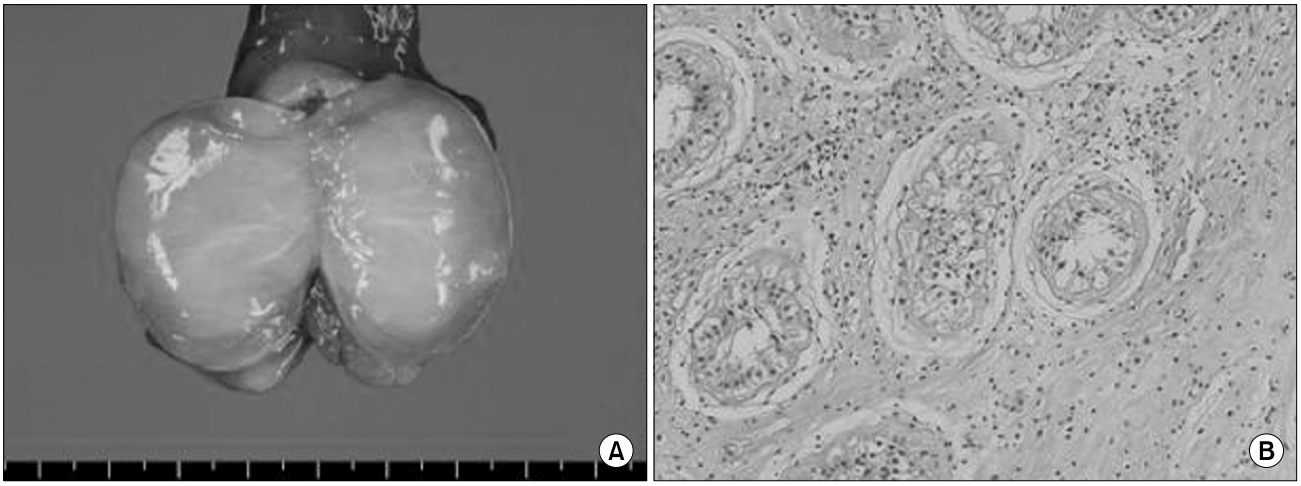

Fig. 4. (A) Gross finding of the right testis shows a well-defined, light yellow, solid scar-like mass, measuring 3.9x2.8 cm. The mass has replaced most of the testis and was abutting the tunica albuginea. (B) Microscopic finding of the right testis shows a tubular hyalinization dominant pattern with intratubular germ cell neoplasia (H&E, x400).

seminoma. The prevalence of testicular tumors is 0.65 out of 100,000 pediatric patients,[3] but there are no reports on this phenomenon. Baek et al[4] reported that a patient admitted complaining of a testicular mass underwent surgery under suspicion of a testicular tumor, but there was only hemorrhagic necrosis with no testicular tissue. However, this case differed from the burned out phenomenon.

The theory of 'metastasis of primary germ cell tumor of testis' and 'extragonadal germ cell tumor' are the main theories about the origin of retroperitoneal extragonadal germ cell tumors. Metastasis of a primary germ cell tumor is the spread of testicular tumor cells via lymphatic channels. Hailemariam et al,[5] however, suggested the theory of an extragonadal germ cell tumor, in which the tumor cell is usually originated from a primitive germ cell located in the midline of the body that failed to migrate to the scrotum during the development of these cells. In addition, it has been reported that the tumor cell origins from primitive totipotential cells remain from the morula and blastemal stages, which are later cell differentiation periods.

The mechanism for the regression of a primary gonadal tumor is unclear, but it has been suggested that the regression results from immunologic or ischemic change, as reported in patients with renal cell carcinoma, breast cancer, lymphoma, and malignant melanoma. It has been reported in 25% of patients with regression of malignant melanoma. It was suggested that the reason this phenomenon results from an immunologic mechanism is the local infiltration of T lymphocytes causing tissue fibrosis during regression of melanoma. Saleh et al[6] reported that the appearances of primary malignant melanoma disappeared completely after repeatedly disposing tumor antigen to the immune system in 17 malignant melanoma patients. The mechanisms of this theory will need to be studied provided that interstitial hyaline degeneration and fibrosis are considered to be part of the tumor regression reaction.

Patients with extragonadal germ cell tumors with the burned out phenomenon usually complain of many vague symptoms, such as flank pain, abdominal mass, night sweating, or scrotal pain, and occasionally reveal an elevated testicular tumor marker. In this case, the patient only complained of abdominal pain without elevation of tumor markers (hCG, LDH, aFP). Hence, it was not sufficient to diagnose the phenomenon through clinical features and tumor markers. Tasu et al[7] also reported that it was difficult to diagnose this phenomenon through clinical features in 5 patients and that ultrasonography was valuable for the diagnosis because there was microlithiasis in all patients. Because these patients usually present with variable clinical features and it is easy to pass over this diagnosis, in patients with testicular microlithiasis, we should consider a testicular tumor with the burned out phenomenon.

There was an agreement to radical orchiectomy because of the existence of an intratubular germ cell tumor in the patient with the burned out phenomenon. Primary extragonadal germ cell tumors resist chemotherapeutic agents more than do

metastatic extragonadal germ cell tumors, so we should differentiate between primary and metastatic tumors.[8] We usually treat metastatic extragonadal germ cell tumors with combined chemotherapeutic agents, but primary extragonadal germ cell tumors usually resist the chemotherapy. This resistance seems to be caused by a junctional complex between Sertoli cells, which act as a blood-testis barrier to chemotherapeutic agents. There was a report that the primary tumor still remained in more than half of patients after chemotherapy.[9] Therefore, orchiectomy was recommended before chemotherapy. We diagnosed the retroperitoneal mass as an extragonadal germ cell tumor and found an intratubular germ cell tumor surrounded by fibrotic tissues in the orchiectomy specimen.

Extragonadal germ cell tumors are more aggressive than primary testicular cancer, and resection of this tumor is difficult because of invasion to surrounding tissues. Because nonseminomatous extragonadal germ cell tumors are usually resistant to chemotherapy, the 5-year survival rate is reported to be 0-67%. On the other hand, extragonadal seminoma is sensitive to chemotherapy and the 5-year survival rate is more than 90%.[10]

Patients with the burned out phenomenon, a manifestation of germ cell tumors, differ in diagnosis and management from patients with other testicular cancer. We suggest a more extensive survey about the origin and the mechanism of this phenomenon.

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