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Case Report

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Choriocarcinoma Syndrome Complicating a Mixed Testicular Burned Out Tumor: About a Case

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Abstract

A 32 year old man was admitted to our hospital for refractory back pain despite treatment by usual painkillers. Imaging exams revealed multiple enlarged and necrotic lymph nodes throughout the body and bilateral lung metastases. High beta HCG levels in his blood were suggestive of a testicular germ cell tumor, even though testicular clinical examination and ultrasonography weren't conclusive. Surgical biopsy of an axillary lymphadenopathy revealed the presence of choriocarcinoma cells along with embryonal carcinoma cells, compatible with a mixed germ cell tumor. Before chemotherapy initiation, our patient developed an acute dyspnoea caused by choriocarcinoma syndrome that eventually resulted in his death. We report a rare case of choriocarcinoma syndrome originating from a burned out mixed testicular tumor.

Key words: Mixed burned out tumor; Choriocarcinome syndrome; Factor V deficiency

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Case Report

A 32 year old man was sent to our medical centre by his family doctor, which was consulted for lingering back pain. An abdominal mass was discovered during physical examination.

Anamnesis revealed that, along with back pain, the patient suffered from asthenia as well as weight loss of a couple of kilograms. There was no fever or dyspnoea, nor was there any history of coughing or urinary symptoms.

The patient, originally from Slovakia, arrived in Belgium 4 years ago. He worked as a dustman and lived in a small apartment with his wife and his 3 children.

The patient had no relevant prior medical history and did not consume alcohol, tobacco or drugs. He hadn't had any surgery in the past and took no day to day treatment.

Physical examination revealed only a stony abdominal mass located above the bladder. Testicular palpation was normal, however, bilateral lung metastases and multiple, some necrotic, lymphadenopathies (axillary, tracheal, para-oesophageal, para-aortic and pre-sacral) were found on the chest, abdomen, and pelvis CT-scan (Figure 1).

Ultrasonography of the scrotum showed no obvious mass, but multiple microlithiasis in both testicles and remarkable right testis hypotrophy (7,6 ml), with a small nodular area and a macro-calcification of about 6mm in its lower pole (Figure 2).

The FDG PET-CT showed multiple hypermetabolic lymph nodes above and below the diaphragm, disseminated in both lungs metastases, and left testis aspecific moderate hypermetabolism (Figure 3).

CRP-levels were slightly elevated (33,95 mg/L) and the sedimentation rate was of 69 mm/h. The patient showed a slight anaemia (12,6 g/dl). High levels of lactate dehydrogenase (LDH: 628 UI/L) and total beta-hCG (64980 UI/L) were found while alfa foeto protein-levels (AFP: 4,5 µg/L) was normal.

Considering the multiple retroperitoneal necrotising lymphadenopathies, tuberculosis and neoplastic diseases such as lymphoma and germinal neoplasia had to be excluded, as well as some other infectious diseases such as cat-scratch disease, syphilis, HIV and some other rare causes such as sarcoidosis and Kikuchi Fujimoto disease.

Testing for HIV and syphilis came back negative, and peripheral blood immunophenotyping was normal. A surgical biopsy of an accessible clavicular adenopathy showed cells compatible with a metastatic mixed germinal tumor composed of embryonal carcinoma cells (80% of the tumor volume) and choriocarcinoma (20% of the tumor volume).

Thus the patient's diagnosis was stage IV mixed extra-gonadic germ cell tumor.

He was then referred to our university centre specialized in oncology. Four cycles of chemotherapy based on bleomycin, etoposide and cisplatin were to be started.

Besides his mixed germ cell tumor, congenital factor V deficiency was found (FV activity assay was measured at 55% (normal range; 70-150%)).

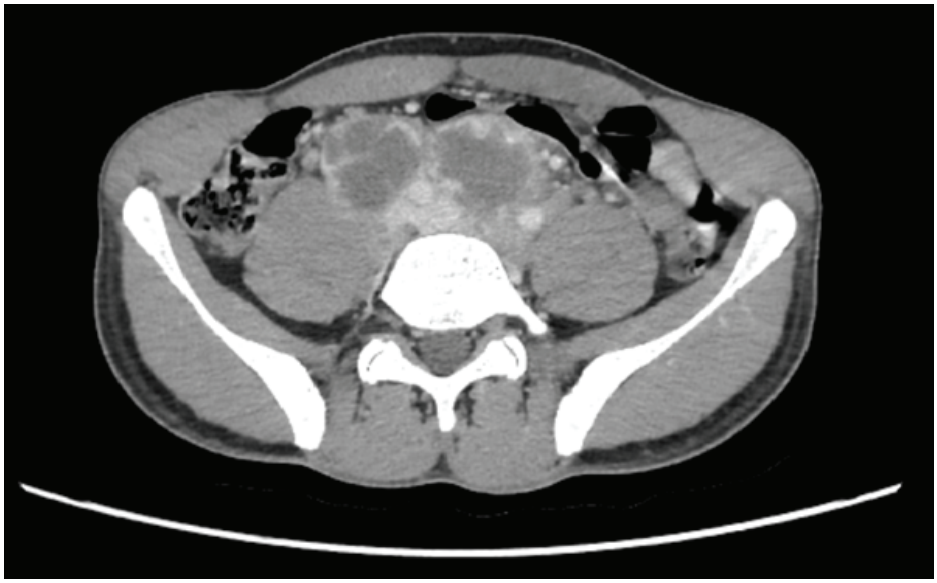

Figure 1: Abdominal CT-scan showing multiple necrotic pelvic lymphadenopathies.

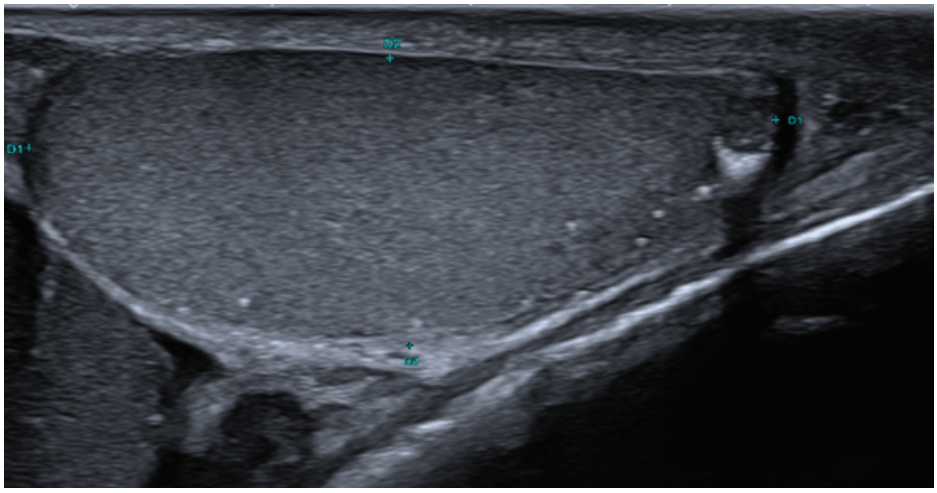

Figure 2: Right testicular ultrasound showing multiple microlithiasis, right testis hypothrophy and a macro-calcification (6 mm).

Sadly, few days later, our patient developed acute dyspnoea. Chest CT-scan performed in emergency showed enlarging known metastases and large alveolar infiltrates disseminated in both fields (Figure 4). Total HCG levels had increased to 999080 UI/L. This clinical presentation associating a sudden respiratory distress, along with a rapid evolution of the pulmonary lesions (as shown by CT-scan) and an image of alveolar haemorrhage, is compatible with choriocarcinoma syndrome (Figure 5).

Unfortunately, despite getting his first BEP chemotherapy while in the intensive care unit, our patient's condition did not improve and resulted in his death.

Discussion

Testicular cancer is a rare neoplasm, representing only 1 to 2% of all cancers among men [1], yet it is the most frequent neoplasm observed in young men. Testicular cancer develops mainly from germ cells, and can therefore be classified as a form of germ cell tumors (GCT) [2].

The vast majority (90%) of GCT have a gonadal origin. Testicular cancers typically show peak incidence, which has been increasing progressively while mortality has been decreasing, during the fourth decade for Seminomatous Germ Cell Tumors (SGCT) and during the third decade for Non Seminomatous Germ Cell Tumors (NSGCT). NSGCT are comprised of teratomas, embryonal carcinoma, and choriocarcinoma and yolk sac tumors. Often, GCT's are not pure tumors but made out of multiple histological types [1].

Histological analysis of our patients' tumor showed a mixed tumor, composed of Embryonal Carcinoma (EC) as well as choriocarcinoma.

These two types of tumors particularly are known to show partial or complete testicular tumor regression, a phenomenon commonly called "burned-out" tumor. At this stage, the patients' testis are left with scar tissue, and extra-gonadal metastasis are present [3,4].

These metastasis are often located in the retro peritoneum, lymph nodes and the lungs, as seen in our patient, as well as in the mediastinum or the liver [5,6].

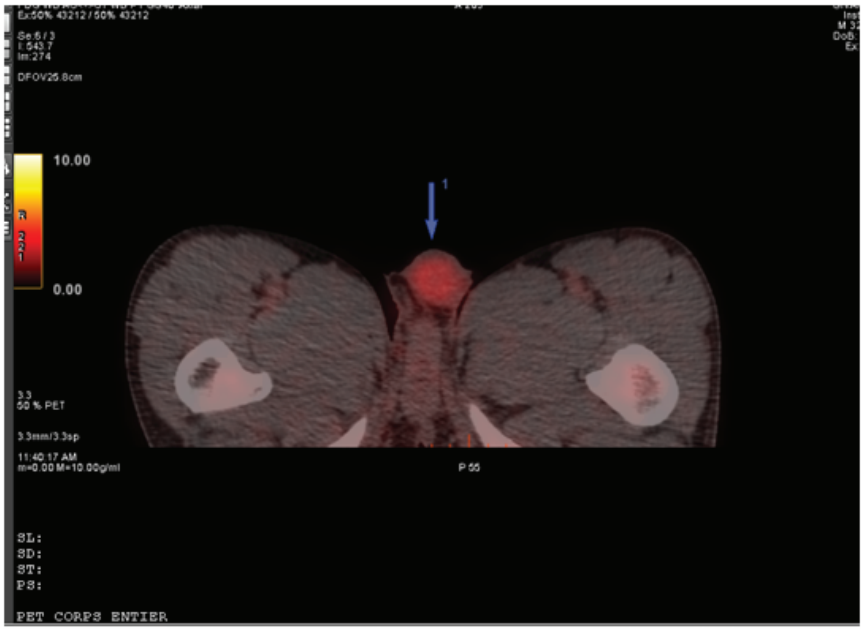

Figure 3: FDG PET-CT showing aspecific moderate left testicular hypermetabolism.

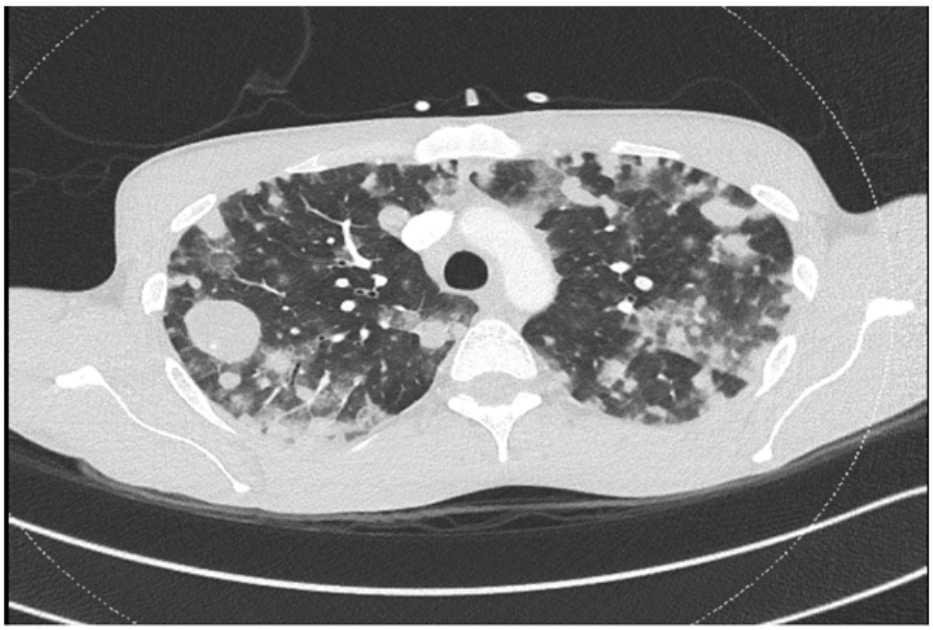

Figure 4: Chest CT-scan showing large alveolar infiltrates of both fields and enlarging pre-existing lymphadenopathies.

EC in its pure form is quite rare; however it is the second most frequent GCT that can be encountered as such. Metastasis at diagnosis are frequent, due to its early vascular spread, and is directly correlated with the proportion of EC in the tumor. Total HCG secretion can occur in EC, however, elevation of that marker is typically associated with choriocarcinoma [7].

Choriocarcinoma is a rare form of NSGCT, and accounts only for 1% to 3% of all GCT. Pure forms of choriocarcinoma are very rare (0, 3% of all GCT) [8].

βHCG is always secreted by choriocarcinoma and plays an important role in prognosis. Metastasis at diagnosis is frequent, yet not systematic, and due to lymphatic and vascular spreading [9].

Choriocarcinoma Syndrome (CS) is a complication in 10% of cases. CS is a clinical entity characterized by rapidly progressive and hemorrhagic tumors. Its pathogenesis is yet unknown but it has been related to a direct invasion of small vessels.

Typically, choriocarcinoma syndrome appears shortly after the introduction of the chemotherapy, yet pre-treatment onsets have been reported in literature [10].

Diagnosis is made in front of an acute pulmonary haemorrhage along with lungs metastases.

Prognosis is poor in such cases due to its hemodynamic implications, especially when associated with levels of βHCG > 50000 IU/l, directly secreted by the tumor cells [11].

Treatment is Commonly Combined Chemotherapy (BEP). Surgery may be required in case of life-threatening haemorrhage.

Factor V defiency is a rare cause of coagulopathy. Most of the patients are asymptomatic, diagnosis commonly made by a routine blood test. The only treatment is transfusion of fresh frozen plasma, mostly during surgery [12].

In our case, congenital Factor V deficiency might be implicated in our patients' rapidly worsening medical condition. Although the pathogenesis of CS is not yet certain, some authors reported that the tumor itself may participate directly to the haemorrhage, by development of products causing damage to the blood vessels [13].

In conclusion, we report a case of rapidly evolving choriocarcinoma syndrome secondary to a burned out testicular tumor (consisting of Embryonal Carcinoma (EC) as well as choriocarcinoma) which eventually resulted in our patients' death. We suspect that his congenital factor V deficiency could partly explain his unfavorable evolution.

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