INCIDENTAL DISCOVERY OF TESTICULAR MICROLITHIASIS: WHAT IS THE IMPORTANCE OF ULTRASOUND SURVEILLANCE? REPORT OF TWO CASES.

Antonio De Fiores, Clinica Guarnieri, Rome, Italy

Valentina De Marco, Department of Radiology, Policlinico Umberto I, Sapienza Univerity of Rome, Italy

Flavio Barchetti, Department of Radiology, Policlinico Umberto I, Sapienza Univerity of Rome, Italy

Giovanni Barchetti, Department of Radiology, Policlinico Umberto I, Sapienza Univerity of Rome, Italy

Corresponding Author: Flavio Barchetti, flavio.barchetti@live.it, Viale del Policlinico 155, 00161, Telephone Number 3498110106, Fax Number +39064456695

INTRODUCTION

Primary retroperitoneal germ cell tumors account for approximately 30% of extragonadal germ cell tumors and about 10% of all primary malignant retroperitoneal tumors [1].
Many studies demonstrate the association between diffuse bilateral testicular microlithiasis (TM) and gonadal and extragonadal germ cell tumors [2,3].
Nevertheless, it is still uncertain if ultrasound surveillance is really necessary in patients with TM in absence of other risk factors such as previous testicular cancer, a history of cryptorchidism or testicular atrophy [4].
We report two cases of a 33-year-old and 39 year-old men respectively, presenting with a retroperitoneal extragonadal tumor and bilateral testicular microlithiasis, without a focal testicular mass found on ultrasonography.

CASE 1

A 39 year-old man with a six month history of lumbar pain came to our hospital to perform a MRI in order to rule out a lumbosacral hernia. The MRI images showed no slipped discs, but unfortunately detected a voluminous retroperitoneal solid mass (Fig. 1). Therefore we decided to perform a total-body CT to better characterize the mass and its relationship with adjacent structures. CT images showed a large heterogeneous retroperitoneal mass with curvilinear calcification and a marked inhomogeneous ehancement after intravenous contrast-medium injection, due to the presence of necrotic-colliquatives areas. This lesion displaced the left renal vein cranially, the abdominal aorta anteriorly and toward the right, and infiltratated the inferior vena cava, the left renal vein and the left psoas muscle (Fig. 2,3).

The patient's α-fetoprotein, lactate dehydrogenase and the beta subunit of human chorionic gonadotropin levels were 2680 IU/L (normal, 90–180 IU/L), 279 ng/mL (normal, 0–7.5 ng/mL) and 4 mUI/ml (normal < 5 mUI/ml). We performed a scrotal ultrasonography to rule out that this mass was a retroperitoneal metastasis of primary testicular tumor: the imaging revealed bilateral classic testicular microlithiasis (defined as more than 5 calcifications scattered throughout the testicle), without a focal lesion. (Fig. 4); the patient recalled that he underwent a scrotal ultrasonography when he was 25 years-old because of a testicular trauma. Comparing the current ultrasound images with previous images, we noticed that microcalcifications pattern was very similar.

The patient underwent CT-guided biopsy and histological examination was diagnostic of immature teratoma.

CASE 2

A 33 years-old man came to our emergency department coplaining of abdominal pain, vomiting, weight loss and mild jaundice. The ultrasound study detected a large ill-defined heterogenous abdominal mass.

Patient's serum α-fetoprotein, lactate dehydrogenase and the beta subunit of human chorionic gonadotropin levels were 2470 IU/L (normal, 90–180 IU/L), 232 ng/mL (normal, 0–7.5 ng/mL) and 3 mUI/ml (normal < 5 mUI/ml). Besides, the serum markers of cholestasis were high: conjugated bilirubin was 2 mg/100ml (normal, < 0.2 mg/100 ml), gamma-glutammiltranspeptidase 70 UI/l (normal, 1 - 30 UI/l) and alkaline-phosphates 300 UI/l (normal, <170 UI/l).

CT and MRI examinations showed a giant retroperitoneal mass composed by multiple necrotic-colliquative fluid areas with a multilocular aspect, which dislocated anteriorly the inferior vena cava with possible infiltrating signs, compressed the portal vein and the common bile duct with moderate dilatation of the intrahepatic ducts (Fig. 5).

We performed a ultrasonography, in order to rule out the presence of primary testicular tumor, which revealed bilateral testicular microlithiasis without a focal hypoechoic lesion; the microcalcifications pattern was quite similar to that of a past ultrasound exam performed when the patient was 22 years-old because of suspected varicocele.

The patient underwent a CT-guided biopsy and histological examination was diagnostic of yolk sac tumor.

DISCUSSION

Extragonadal germ cell tumors (EGCT) account for 1–5% of all germ cell tumors [1]. An extragonadal germ cell tumor is by definition a germ cell neoplasm displaying one of the histologic types associated with gonadal origin but located outside the gonads [5]. The most widely accepted theory suggests that extragonadal germ cell tumors arise from primordial germ cells misplaced during their migration to gonads [1].

It remains uncertain, however, whether such tumors develop primarily at extragonadal sites or represent metastases of a primary testicular tumor [2].

Regarding the latter case, EGCT may have developed from ‘burned out testicular tumors’ or they may just be metastatic lesions from primary testicular tumors that were not detected at the time of the diagnosis of EGCT [6]. A burned out gonadal primary tumor is a regressed tumor which is seen as an echogenic scar or a hypoechoic tissue on testicular ultrasound and that clinically presents with metastasis.

Histologically, extragonadal germ cell tumors comprise seminomas (30–40%) and nonseminomatous tumors (60–70%) in men and dysgerminomas and nondysgerminomas in women. Nonseminomatous germ cell tumors (NSGCTs) include teratoma, embryonal carcinoma, endodermal sinus tumor (yolk sac tumor), choriocarcinoma, and tumors with mixed histology. NSGCTs usually have a more aggressive course than do seminomatous tumors [1]. Extragonadal nonseminomatous tumors present with an elevated α-fetoprotein or human chorionic gonadotropin level [5]. α-fetoprotein is produced by endodermal sinus tumors either alone or in

association with other types of germ cell tumors. Human chorionic gonadotropin is only produced by syncytiotrophoblasts occurring as a component of choriocarcinoma. These are useful serum markers in the diagnosis, prognosis, and follow-up of patients with germ cell tumors [5].

The majority of the extragonadal germ cell tumors occur in men, except benign mature teratoma, which occurs with equal frequency in men and women . Extragonadal germ cell tumors are usually seen in children or young adults and typically arise in midline locations. In adults, the most common sites of primary extragonadal germ cell tumors are, in descending order, the mediastinum, retroperitoneum, and cranium. In children, the cranium and sacrococcygeal region are the common sites [1].

Primary retroperitoneal germ cell tumors account for about 10% of all primary malignant retroperitoneal tumors and about 30–40% of extragonadal germ cell tumors [1].

Extragonadal germ cell tumor is often seen in or near the midline, especially between the T6 and S2 vertebrae. A midline mass is more suggestive of a primary extragonadal germ cell tumor than of metastasis [7]. These tumors are usually large at presentation. Encasement, displacement, and compression of the abdominal vessels are common. Patients may present with metastases: brain, liver, lungs, and bones are the common sites of metastases [1].

The radiologic findings for primary extragonadal germ cell tumors are non specific. The imaging appearances are similar to those of gonadal germ cell tumors. Seminoma is rare in the retroperitoneum and is seen as a large, lobulated, well-defined homogeneous solid mass with fibrous septa and ringlike or speckled calcifications. Nonseminomatous germ cell tumors are depicted as heterogeneous tumors with areas of hemorrhage, necrosis, and heterogeneous enhancement. Flow voids that are due to hypervascularity may be seen, as well as invasion of adjacent structures [7].

Primary testicular malignancy and extragonadal germ cell tumors are often associated with testicular microlithiasis [2,8]. This is an uncommon pathologic condition that is detected by scrotal ultrasonography and is defined as the precence within the substance of the testis of five or more speckled bright foci, 1 to 2 mm in diameter, with little or no acoustic shadowing; the microcalcifications usually affect both testes but may be unilateral, and can be focal or diffuse [3,4].

J. Richenberg and N. Brejt affirm that ultrasound surveillance is unlikely to benefit patients with TM in the absence of other risk factors, on the

contrary in the presence of additional risk factors (previous testicular cancer, a history of maldescent or testicular atrophy) patients are likely to be under clinical and ultrasound surveillance [4].
Nevertheless, it is still controversial whether performing sonographic survellaince is better than regular testicular self-examination in adult patients with classic testicular microlithiasis and absence of any known testicular tumor.
In this setting, on the basis of our direct experience, we highlight the importance of annually ultrasonographic surveillance of testis and retroperitonel space in patients with occasionally detected testicular microlithiasis. Besides, taking into account the increased risk of metachronous testicular malignancy in patients with previous extragonadal germ cell tumor [6,9], we recommend yearly testicular ultrasound follow-up after surgical removal of retroperitoneal gonadic tumor.

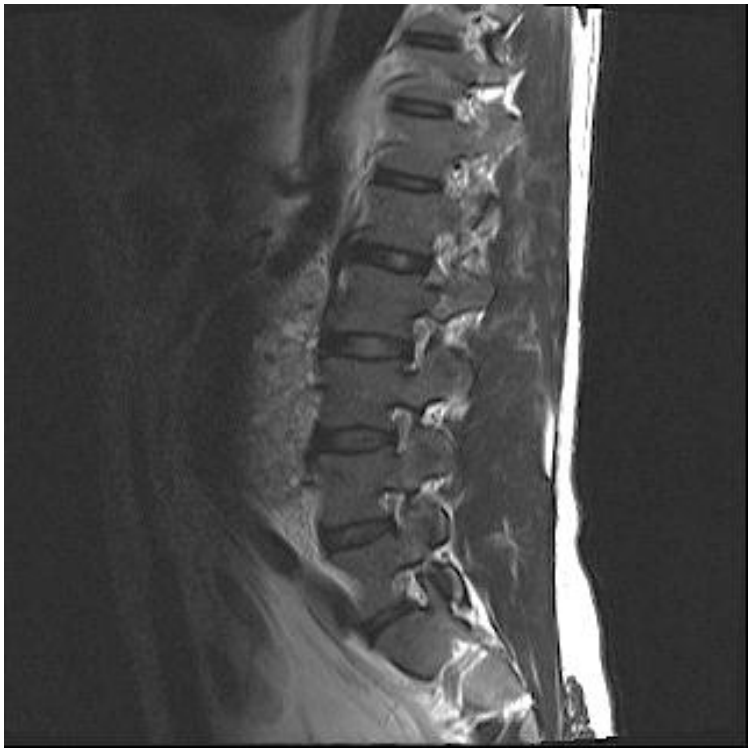

Fig. 1 Sagittal T2-weighted MR image shows a voluminous heterogeneous solid mass in the retroperitoneal space, displacing anteriorly the addominal aorta.

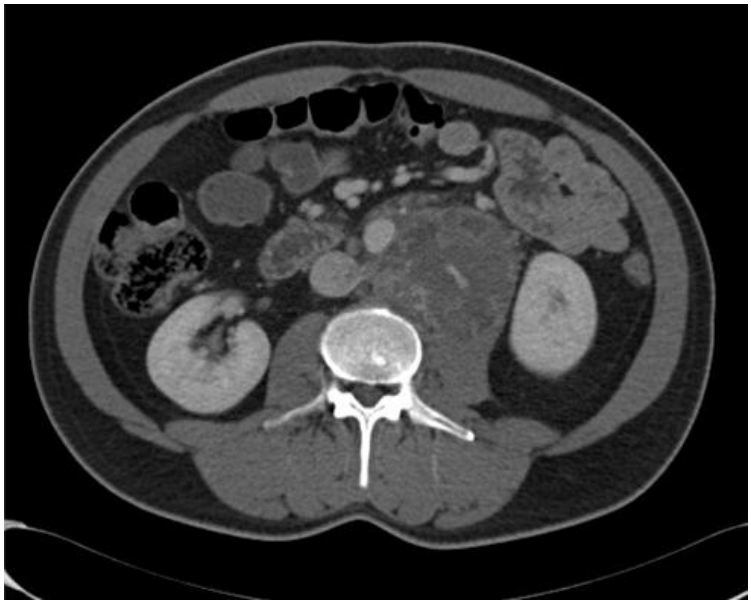

Fig. 2 Axial CT image obtained during venous phase demonstrates a retroperitoneal heterogeneous mass infiltrating the left psoas muscle.

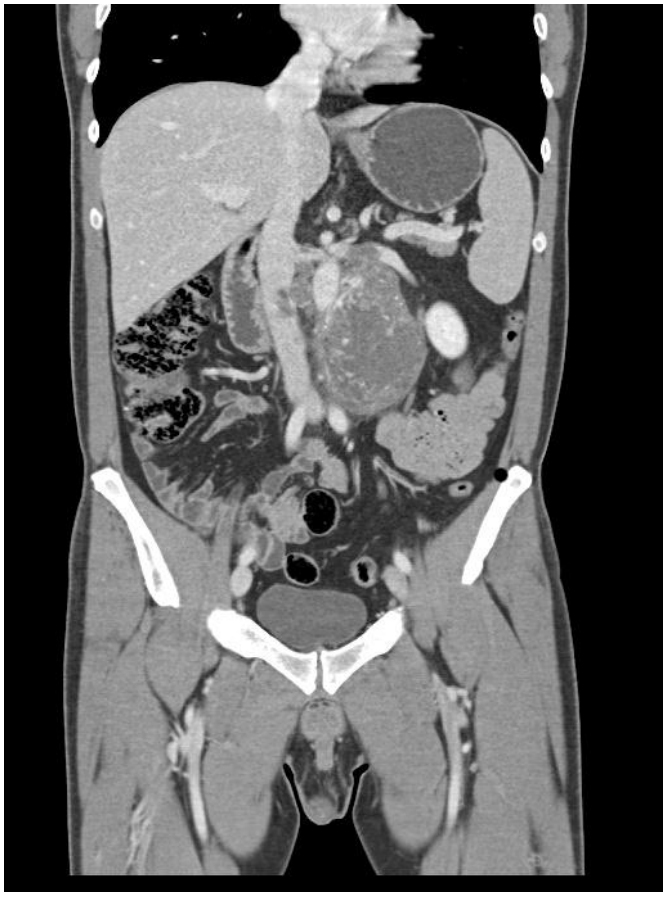

Fig. 3 Coronal CT image shows the mass infiltrating the inferior vena cava and the left renal vein.

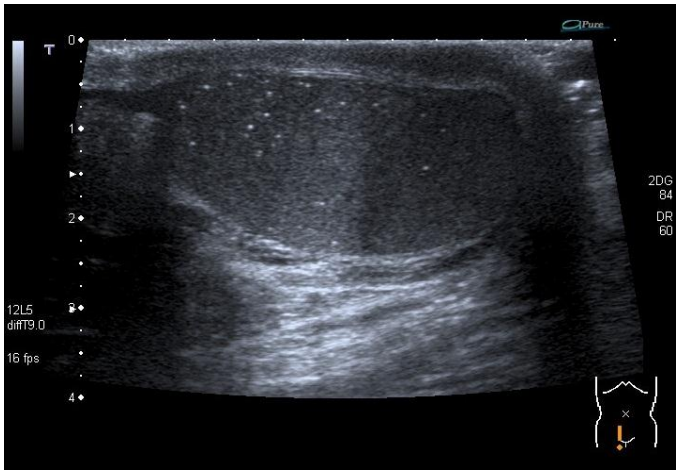

Fig. 4 Ultrasound longitudinal image show classic testicular microlithiasis of right testis without focal lesions.

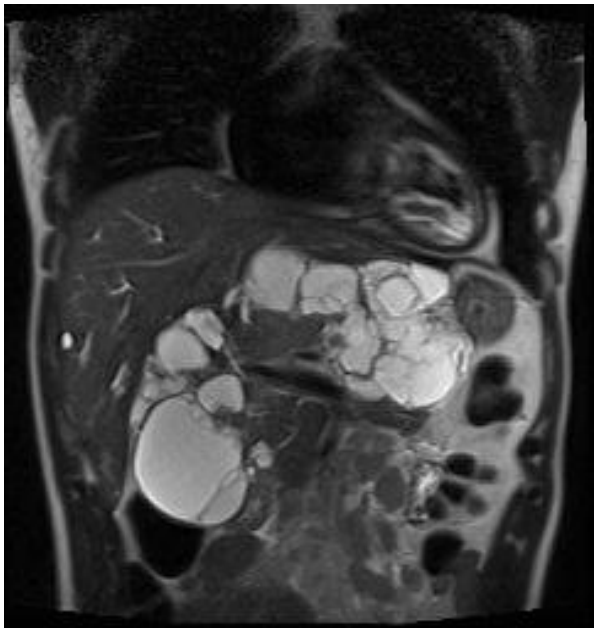

Fig. 5 Coronal T2 weighted MR image shows a large retroperitoneal mass composed by multiple necrotic-colliquative fluid areas with a multilocular appearance.

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