

Primary extragonadal germ cell tumor of the retroperitoneum: case presentation and review

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Abstract

Primary extragonadal germ cell tumors of the retroperitoneum are extremely rare. During embryonic development germ cells are first seen outside of the embryo, inside the yolk sac. At about four to six weeks of development, these cells migrate into the embryo where they populate the developing testes or ovaries. When these cells miss their destination, they tend to remain in one of the midline sites of the body. These are usually one of three locations: the anterior mediastinum, the retroperitoneum or the pineal gland. This case is that of an extragonadal germ cell tumor of the retroperitoneum, to the present, still of primary origin. Differentiating between primary and secondary tumors of this type can be difficult, as these tumors can regress or develop slowly. Ultrasound and computed tomography play a crucial role in the detection of these tumors. The treatment of extragonadal germ cell tumors involves surgery, radiation and chemotherapy.

Key words

Gonads, extragonadal germ cell tumors (EGCT), retroperitoneum, primary, yolk sac, ultrasound, computed tomography

Resumen

Los tumores primarios extragonadales de células germinales en el retroperitoneo son extremadamente raros. Durante el desarrollo embrionario, las células germinales aparecen primero fuera del embrión, dentro del saco vitelino. Entre las cuatro y seis semanas de desarrollo, estas células migran hacia dentro del embrión donde pueblan los ovarios o testículos en desarrollo. Cuando estas células no llegan a su destino, tienden a quedar en sitios de la línea media del cuerpo; generalmente en una de tres localizaciones: el mediastino anterior, el retroperitoneo o la glándula pineal. En este caso es un tumor extragonadal de células germinales en el retroperitoneo, hasta el momento de origen primario. Diferenciar entre estos tumores primarios y secundarios puede resultar difícil, ya que estos tumores pueden remitir o desarrollarse lentamente. El ultrasonido y la tomografía computarizada tienen un papel esencial en la detección de estos tumores. El tratamiento de los tumores extragonadales de células germinales incluye cirugía, radiación y quimioterapia.

Palabras clave

Gónadas, tumores germinales extragonadales, retroperitoneo, primario, saco vitelino, ultrasonido, tomografía computarizada

■ INTRODUCTION

Germ cell tumors are one of the most common tumors among young adult men. An extragonadal germ cell tumor (EGCT) is a germ cell neoplasm having one of the histolo-

gies associated with gonadal origin, but located outside the gonads. These tumors are relatively uncommon but represent 1 to 5% of all germ cell tumors. Seminomas account for 30 to 40% of these tumors. And non seminomatous tumors account for 60 to 70%. The most common site for EGCT is the mediastinum (50 to 70%) followed by the retroperito-

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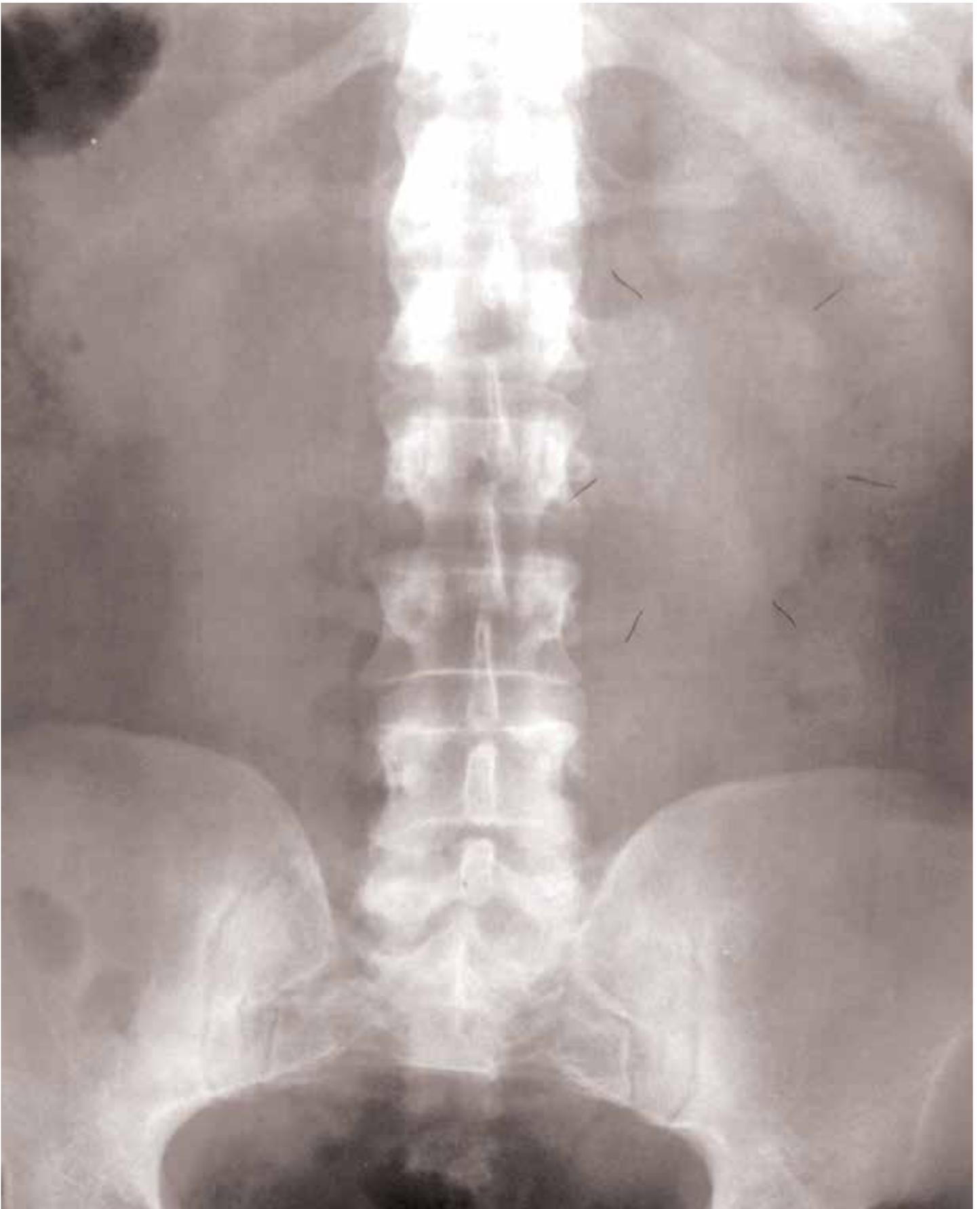


Figure 1. Simple abdominal film revealing a soft tissue mass occupying the left upper quadrant, located medial to the left kidney and parallel from L2 to L4

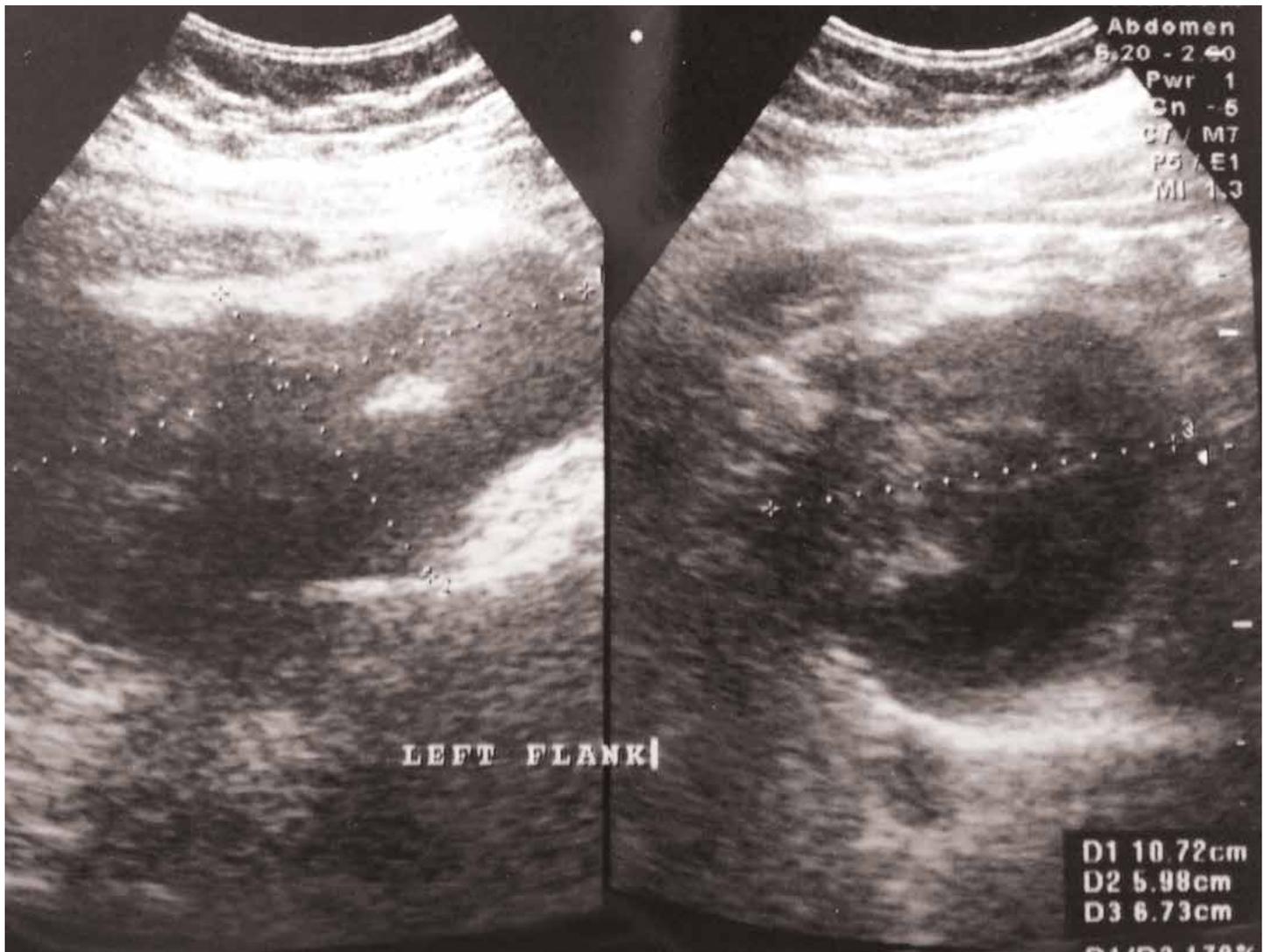


Figure 2. Ultrasound findings revealing a large solid mass with calcifications in the left flank arising from the retroperitoneum. It measured: 10.72cm x 5.98cm x 6.73cm and presented no vascularity with color Doppler interrogation.

neum (30 to 40%), pineal gland (5%), and sacrococcygeal area (less than 5%).

The treatment and prognosis of the disease depends on a variety of factors including the type of cancer, the tumor location and the size of the tumor.

■ CASE PRESENTATION

A 41 year old male was consulted at the Radiology Department for left lower abdominal pain that radiated to the left renal fossa. These symptoms had been occurring for some months but had become more severe recently (Figure 1).

INVESTIGATIONS AND DIAGNOSIS

A panabdominal ultrasound was initially requested, which reported a large hypoechoic solid appearing mass in the left flank (Figure 2). A contrasted abdominal CT done a few days later confirmed the presence of a large capsulated hypodense solid retroperitoneal mass, medial to the left kidney and producing slight displacement of the renal artery and medial displacement of the abdominal aorta. It measured 11.4cm x 12cm x 8cm. The presence of a capsule and mild to

moderate stranding of the fat surrounding the mass, as well as a leukocytosis of 14,600 with 86% neutrophilia led to the diagnosis of retroperitoneal abscess (Figures 3 and 4).

Note that the testes were sonographically normal.

TREATMENT

The patient was later admitted and surgically intervened to drain and remove this lesion. The post-operative diagnosis was encapsulated retroperitoneal abscess. The gross description was a tan to brown radiated mass measuring 11.5cm x 7.5cm x 5.5cms. The pathology report revealed a diagnosis of anaplastic malignant neoplasm consistent with seminoma.

The patient has since received 24 sessions of radiation therapy. He admits to have also received alternative medicine treatments for cancer from Tijuana. This patient has been doing bi-annual checkups with CT and ultrasound. So far controls have revealed the persistence of a small retroperitoneal mass adjacent to the left psoas muscle (Figure 5). This mass has not increased in size over the past 18 months. Testicular ultrasound so far remains normal.

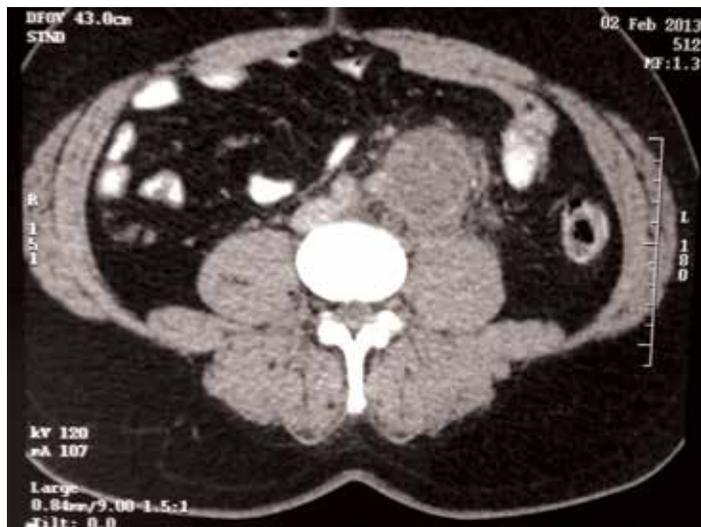


Figure 3 and 4. Computed tomographic scan(axial and reformatted views) revealed a large capsulated hypodense retroperitoneal mass, medial to the left kidney and producing mild superior displacement of the renal artery and medial displacement of the abdominal aorta. The mass measures: 11.4cm x 12cm x 8cm. The capsule enhanced discretely with IV contrast. A small amount of gas is detected in its center and a calcification is seen in the capsule.

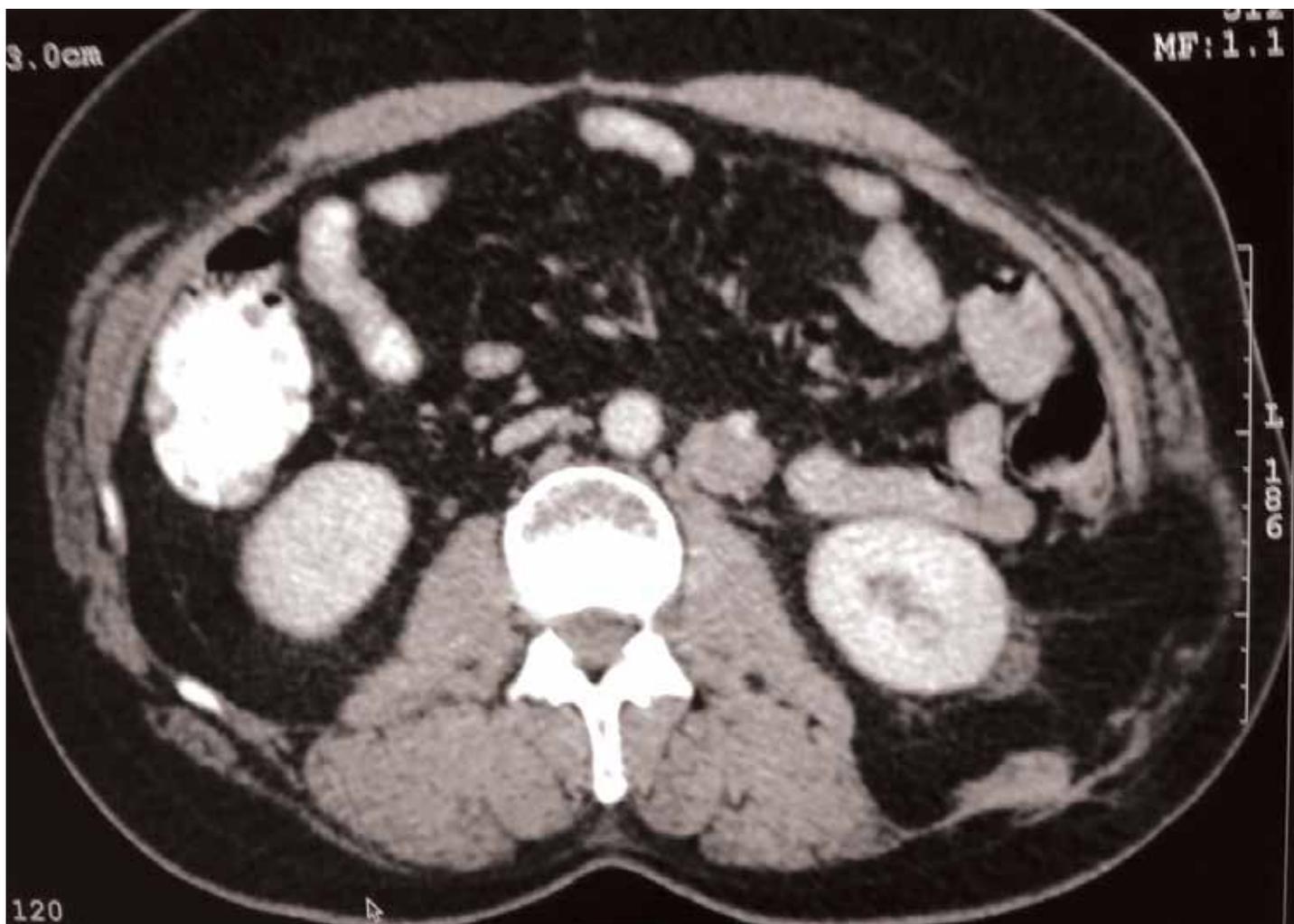


Figure 5. One year post operative CT control shows a small hypodense solid mass measuring 3.0cm x 2.3cm x 2.5cm in the retroperitoneum, in the left paraaortic area, anterior and adjacent to the left psoas muscle.

REVIEW OF EGCT

Pathogenesis

There is sometimes confusion in the diagnosis of primary EGCT. The delayed appearance of secondary tumors is sometimes mistaken for primary tumors. The mediastinum is the most common site for primary EGCT, followed by the retroperitoneum. Usually retroperitoneal germ cell tumors are metastases from primary germ cell tumors of the testicle. The testicle is the most common site for germ cell tumors, occurring in a frequency of 90% of all testicular tumors. Thirty percent of these men initially present with retroperitoneal metastases, making this the most common cause of retroperitoneal germ cell tumors. Indisputable evidence of primary retroperitoneal extragonadal germ cell tumor is often lacking, as incontrovertible evidence can only be ascertained by bilateral orchiectomy and careful sectioning. Our patient's diagnosis is then very reserved in spite of the fact that this patient's testicles are still normal.

All germ cell tumors arise from malignant undifferentiated germ cells. Malignant transformation can occur in any location where germ cells are found. These tumors develop from primordial germ cells located in or near the midline. These germ cells failed to migrate during embryonic development. The primordial germ cells travel from the yolk sac to the genital ridge which extends from T6 to S2. By the end of the 8th week, the gonads extend from the diaphragm to the inguinal canal. Later on they migrate to the scrotum and pelvis. Abnormal germ cells at times remain along this path. It is these remaining cells that can undergo at times neoplastic development.

Due to the occurrence of subclinical testicular abnormalities accompanying retroperitoneal EGCT, other mechanisms have been proposed to explain the occurrence of primary retroperitoneal EGCT. These are: failure of the germ cells to migrate during embryonic development and subsequent transformation into an EGCT; occult gonadal germ cell tumor (burned out tumor) with metastasis or delayed metastasis (asynchronous) or metachronous appearance of a primary testicular germ cell tumor and a primary retroperitoneal EGCT.

Small intratesticular scars and testicular atrophy have been found in some patients with EGCT with no histologic evidence of gonadal neoplasm. The scar represents a "burned out" or regressed germ cell tumor. Hematoxyphilic bodies, hemosiderin pigment and calcium phosphate deposits are present. These indicate the former presence of a neoplasm with subsequent cell death. EGCT may also arise from regressed cryptorchid testicular tumors. The entire testicle may be fibrosed.

In many cases of retroperitoneal EGCT, no abnormality can be found at initial presentation. It may take as much as 8 to 18 years after the removal of the retroperitoneal mass for the testicular primary lesion to manifest.

Clinical features

The average age of EGCT detection is 47 years. Retroperitoneal EGCTs occur later than testicular germ cell tumors (32

years) due to the lack of specific symptoms of retroperitoneal processes in general and this explains why these lesions are detected only after they have reached a certain size.

Abdominal (53%) and low back symptoms (43%) account for most symptoms. This pain may mimic low back pain. Other symptoms and signs include: palpable mass, weight loss, constipation, hip and back pain, dyspnea, leg edema, fever, varicocele, and urinary retention. As previously mentioned the testes may be unilaterally or bilaterally atrophic or contain a scar.

Radiology findings

RADIOGRAPHY

Plain abdominal film findings of EGCT are not different from any other retroperitoneal mass. Findings may include: displaced bowel loops, effacement of psoas muscle margin, displacement of the kidney, and sometimes calcifications. Local bone destruction or bowel obstruction may rarely occur.

CONTRASTED STUDIES

On IVPs (intravenous pyelogram), the ureters and kidneys may be displaced laterally due to a mass. Gastrointestinal studies may reveal displacement or encasement of the bowels by the tumor without evidence of mucosal or mural invasion.

ULTRASONOGRAPHY

In the case of a possible primary retroperitoneal GCT, it is very important to perform ultrasound of the ovaries and testes. Primary tumors may not be palpable, but still visible sonographically. Findings of regressed or burned out primary tumors may range from subtle small echogenic foci, representing scars to moderate sized hypoechoic lesions, representing fibrosis. If testicular microlithiasis is seen, this may be associated with growing tumors or non developing tumors and warrants keen attention and monitoring when seen in a patient with EGCT.

COMPUTED TOMOGRAPHY (CT)

Computed tomography is considered the primary method of choice for detection and staging of retroperitoneal EGCT. These studies should be done with oral and IV contrast material to permit assessment of tumor size, extent, and adjacent organ involvement. CT is also helpful in defining the primary site of metastases to the lungs or lymphatic vessels. A midline mass is more suggestive of a primary malignancy than a metastatic lesion and is the most helpful for achieving the diagnosis. CT is also used to guide biopsy and surgery. CT is used for follow-up of the retroperitoneal masses once treated. Persistent masses, even after therapy, usually represent necrotic or fibrotic reaction to chemotherapy. Quarterly or semi-annual scanning is usually sufficient in most cases even if the mass does not completely resolve.

MAGNETIC RESONANCE IMAGING (MRI)

Very few cases of EGCT have been reported by MRI to permit assessment of its role. MR provides no more of a specific

diagnosis of histological features than any other radiologic technique.

Laboratory findings

The most common markers are human chorionic gonadotropin (hCG) and serum alpha fetoprotein (AFP). hCG is produced by syncytiotrophoblasts occurring as a component of choriocarcinoma or in association with other types of germ cell tumors such as seminomas, embryonal carcinoma or yolk sac tumor. AFP is produced by yolk sac tumors. It can also be produced in association with teratomas.

Therapy

Surgical excision is attempted in resectable lesions. Positive margins or adenopathy often remain. The mainstay of treatment is chemotherapy which may include: vincristine, cyclophosphamide, dactinomycin, bleomycin, cisplatin, and etoposide.

Radiation is employed for irradiation of nodes and surgical margins.

DISCUSSION

Primary germ cell tumors of the retroperitoneum are very uncommon neoplasms that occur without gonadal primary lesion. They account for 1 to 2.5% of all germ cell tumors. The majority of retroperitoneal germ cell tumors in males are really metastases from a testicular primary lesion. To date this case seems to represent a primary EGCT since the patient's testicles remain unaffected.

CONCLUDING REMARK

Seminomas account for approximately one half of all testic-

ular germ cell tumors and are most frequent in the fourth decade. Primary extragonadal germ cell tumors of the retroperitoneum however are extremely rare. When a potential primary EGCT of the retroperitoneum is suspected, it is crucial to do ultrasound of the testes in addition to control abdominal CTs. This is important even if the initial testicular study results are normal, biopsy findings are negative and treatment has been administered, since studies have shown that occult primary testicular seminoma can appear even 8 to 18 years after retroperitoneal and mediastinal tumors were detected at initial presentation.

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