A LARGE SEMINOMA ARISING IN ONE OF BILATERAL UNDESCENDED TESTES: A CASE REPORT

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ABSTRACT

A large seminoma arising in an undescended testis was diagnosed in a 53-year-old infertile man who had bilateral undescended testes and presented with progressive enlargement of abdominal mass. Plain KUB revealed evidence of a large soft tissue mass without calcification in left-sided and mid lower abdomen. CT scan showed a large left-sided retroperitoneal mass, extending from the level of left renal hilum to the left inguinal canal. On CT scan, hypodense areas and small calcifications inside the seminoma were noted. The right -sided undescended testis was atrophy and was located in the right inguinal canal.

PIG = Par infravaginalis gubernaculi
β-HCG = β-human chorionic gonadotropin

INTRODUCTION

Undescended testis is usually within the inguinal canal but can be anywhere along the path of descent from the retroperitoneum. The two most important complications of undescended testis are infertility and testicular cancer. The most common malignancy of the undescended testes is seminoma. A case presenting with a large retroperitoneal mass that proved to be a seminoma in one of bilateral undescended testes is presented with a review of the radiological features and current literatures.

CASE REPORT

A 53-year-old man presented with progressive enlargement of abdominal mass and weight loss 7 kg in approximately 2 months. He was in good health until this time, and he had not previously undergone surgery. He was HIV negative. He was infertile but had no medical work up.

The initial physical examination revealed body temperature 36.5 °C, BP 120/80 mmHg, pulse rate 80 beats/min, and respiratory rate 22 /min. The patient was cachectic, mild pale, and no jaundice. The heart and lungs appear normal. The abdominal examination revealed a large palpable mass in the left-upper and mid lower abdomen, size approximately 15x20x20 cm. It was not movable and was firm in consistency. The testes were not palpable in the scrotal sac. The rest of physical examination including neurological examination was within normal limits.

Plain KUB revealed a large soft tissue mass in left-sided and mid lower abdomen. The mass had neither calcification nor fat density (Figure 1). Chest radiograph appeared normal.

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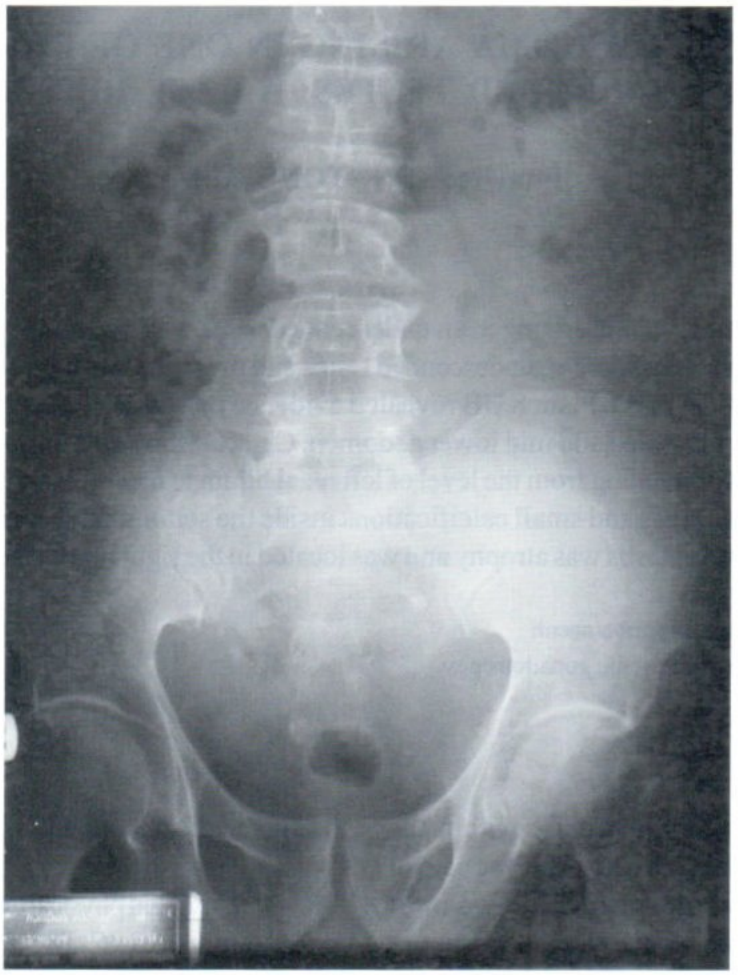

Fig. 1 Plain KUB revealed a large soft tissue mass in left -sided and mid lower abdomen. The mass had neither calcification nor fat density.

Abdominal CT scan showed a large, well circumscribed soft tissue mass extending from deep retroperitoneum space to anterior abdominal wall, left side. Hypodense areas and small calcifications inside the mass were noted. It measured approximately 13x20x22 cm in size. Its most cephalic portion was located at the level of left renal hilum. Its most caudal portion was located in the left inguinal canal. The aorta, IVC, urinary bladder, and sigmoid colon were displaced to the right. The left psoas muscle was compressed. Dilatation of the left pelvio-calyceal system and left proximal ureter was noted. Right kidney appeared normal. An oval-shaped soft tissue mass with central hypodensity was visualized in right inguinal canal, size 1.5x2x2 cm. It was located superomedially to the right inguinal ligament. There was no other right-sided soft tissue mass along the path of testicular descent. Neither ascites nor enlarged lymph node was observed. By CT scan, no evidence of other associated anomaly of the genitourinary system was demonstrated (Figure 2).

The initial tumor markers were measured. The serum level of α-fetoprotein was 2.9 IU/ml (normal value < 4.6 IU/ml) and β-human chorionic gonadotropin (β-HCG) was less than 1.0 mIU/ml.

Fig. 2 Transverse CT scan obtained after administration of oral contrast material.

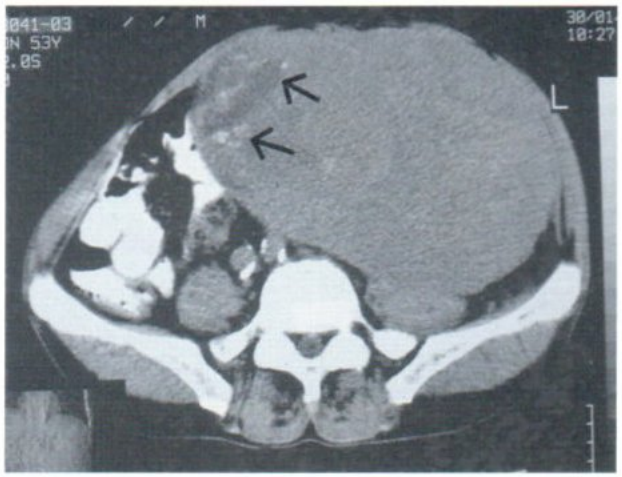

Fig. 2A CT scan obtained before I.V. contrast administration showed a large soft tissue tumor in left-sided retroperitoneum with small calcifications and hypodense areas (arrows).

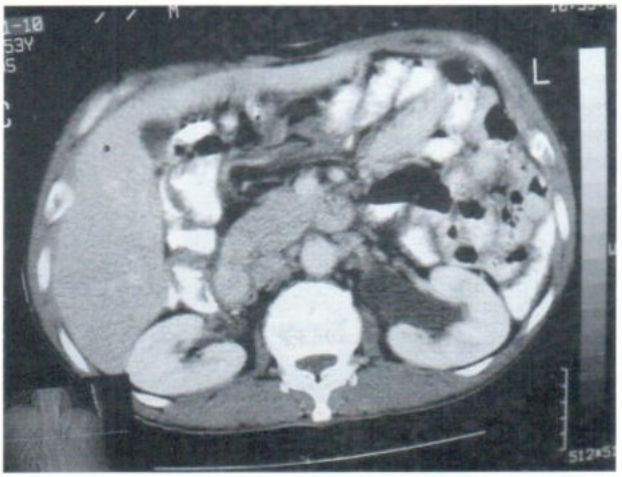

Fig. 2B CT scan obtained after I.V. contrast administration through the level of the kidneys showed hydronephrosis of left kidney.

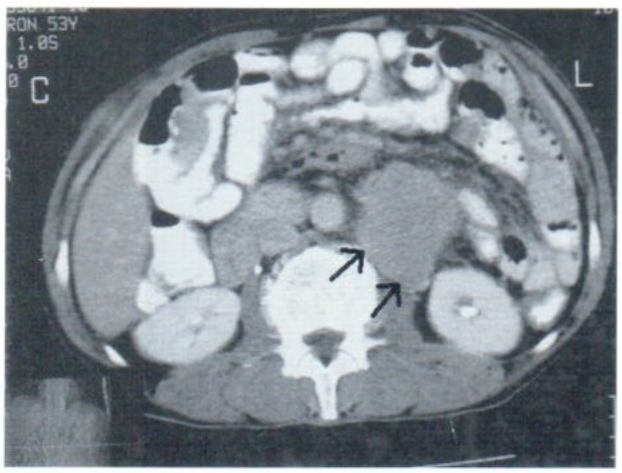

Fig. 2C The CT section caudad to 2B showed a homogeneous retroperitoneal soft tissue mass located between the lower pole of left kidney and aorta (arrows).

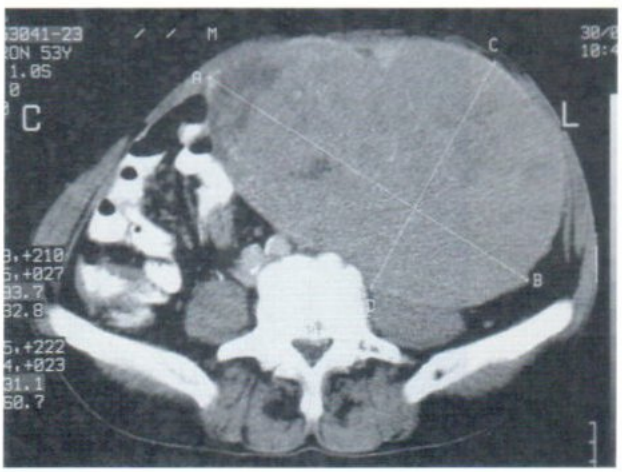

Fig. 2D CT scan post I.V. contrast at the same level of 2A showed that most of the mass enhanced homogeneously except the hypodense areas which could be due to tumor necrosis.

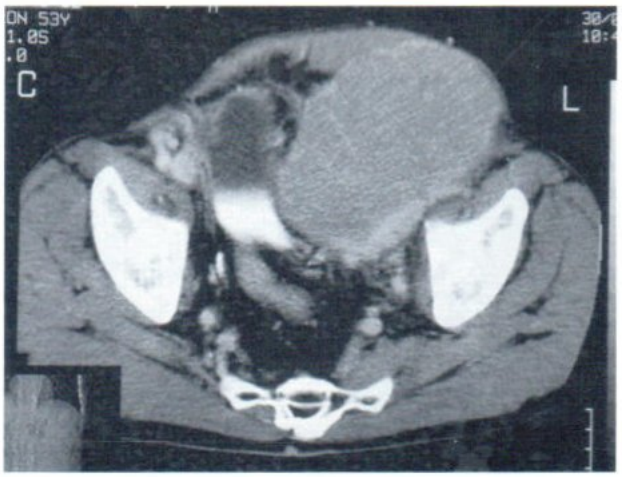

Fig. 2E The mass displaced the urinary bladder and sigmoid colon to the right side.

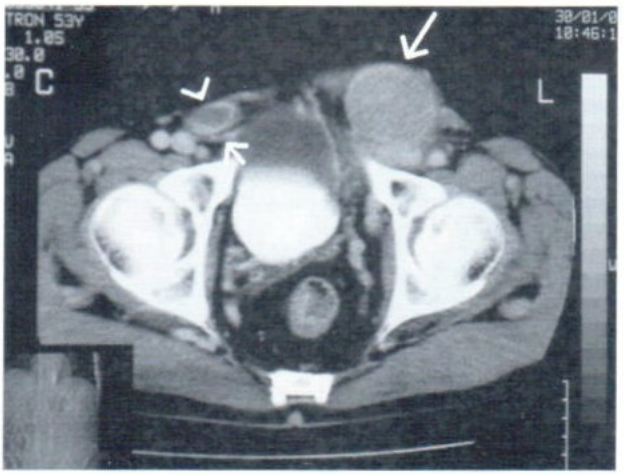

Fig. 2F The tumor extended inferiorly to the left inguinal canal with evidence of left external iliac vein invasion(long arrow). There was a 2 cm mass with central hypodensity in right inguinal canal (arrow head). It was located superomedially to the inguinal ligament (short arrow).

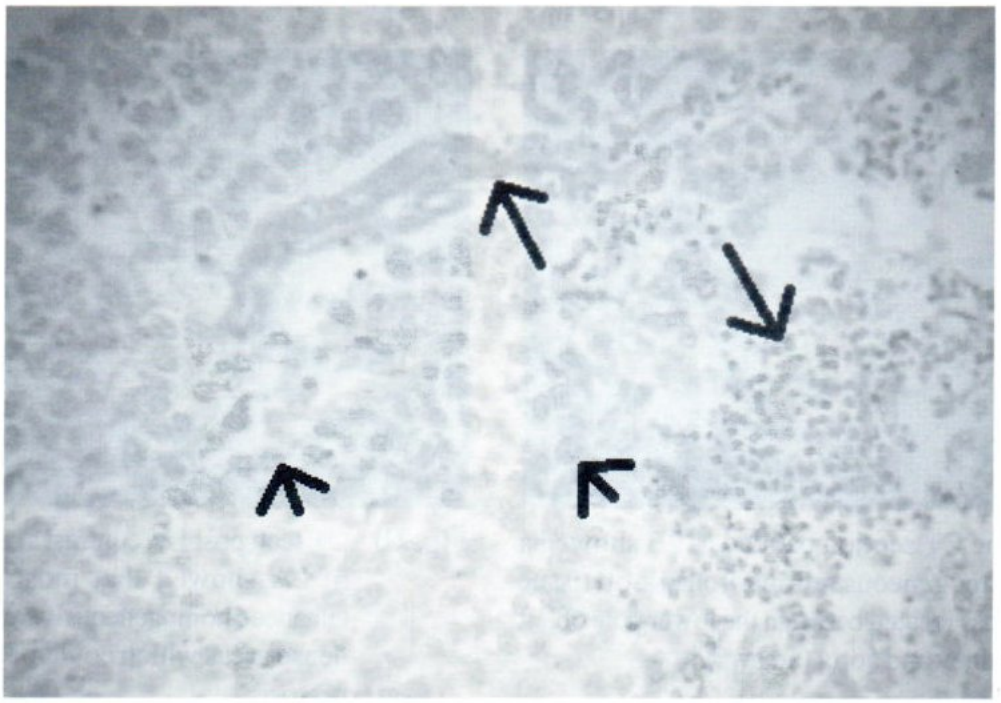

Fig. 3 High-power photomicrograph (Original magnification, x200) revealed lobules of seminoma cells with abundant clear cytoplasm (short arrows) which were separated by fibrous septa that contained numerous lymphocytes (long arrows).

The operation showed a large tumor, measuring 14x17x22 cm. It extended from left renal hilum to the left inguinal canal. The urinary bladder and sigmoid colon were displaced to the right. The left ureter and left external iliac vein were wrapped around by the tumor. External surface was adherent to surrounding tissue. Histological examination of the tumor revealed lobules of seminoma cells with abundant clear cytoplasm. Tumor cells were separated by fibrous septa that contained numerous lymphocytes (Figure 3). The right testis was located in the right inguinal canal near the external inguinal ring. Right orchidectomy was also performed. Histopathology of right testis was atrophic testes.

Seminoma arising in an undescended testis was diagnosed. Postoperative radiotherapy was the additional treatment. Unfortunately, the patient died after incomplete course of radiration due to multiple medical complications.

DISCUSSION AND REVIEW OF LITERATURES

Cryptorchidism results from the abnormal formation and descent of the testes. The testes form from genital ridge, which lie on both sides of the midline and extend from T6 through S2 vertebrae in the developing embryo. Between the 7th and 12th weeks of gestation, the testes contract and become more ovoid as they begin their descent into the pelvis. They remain near the deep inguinal ring until the 7th month of gestation.[1,2] The testes usually descend into the scrotum at approximately 8th month of fetal life.[3-5] About 33% of premature and 3% of full-term male neonates have unilateral undescended testes.[4] It can be bilateral in 10% of patients.[6,7] Most of undescended testes will descend spontaneously around the age of 1 year, but 0.8% of boys continue to have undescended testes. Spontaneous descent after 1 year of age is unlikely.[5]

Factors causing testicular undescended include[1] hormonal factors, especially dihydrotestosterone;[2] the development of the abdominal muscles, which increases the intraabdominal pressure, forcing the testes down (this is the reason that testes are undescended in the absent abdominal muscles syndrome and in some patients with congenital abdominal wall defects;[3] swelling of the gubernaculum, which distends the scrotum and the inguinal canal allows the epididymis, and then the testis, to pass through the inguinal canal.[6]

An incompletely descended testis may be located anywhere from renal hilum to the inguinal canal or along the course of the normal spermatic cord.[8] Sixty-six percent of undescended testes are ectopic (pass through the external ring but not reaching the scrotum), 16% are intracanalicular (within the inguinal canal), 10% are intraabdominal, and 3% are surgically absent.[9]

Because of its association with other urinary tract abnormalities, cryptorchidism is thought to be one manifestation of a generalized defect in genitourinary embryogenesis. Other associated malformations include renal agenesis or ectopias, ureteral duplications, seminal vesical agenesis or cysts, and hypospadias.[10-14]

Undescended testes tend to be more atrophic or dysplastic. They can be associated with significant complications such as cancer, infertility, torsion with infarction, and indirect inguinal hernia.[15-20] Therefore, early diagnosis and management of an undescended testis are required to preserve the patient's fertility and allow early detection of testicular malignancy. Orchiopexy should follow diagnosis of an ectopic testis in a child. Because of the risk of cancer, close observation or surgical removal of the undescended testis is recommended if it is first detected in an adult. Orchiopexy does not prevent the development of cancer; regular self-exmination and medical follow-up are needed.[21]

The pathophysiology of malignant transformation in undescended testes is not completely understood. One hypothesis is that cryptorchidism is not merely incomplete descent of the testis, but that reflects a

generalized defect in embryogenesis and results in bilateral dysgenetic gonads.[14] An embryologic defect in testicular formation is supported by the observation that risk for testicular carcinoma is not limited to the undes- cended testis but extends to the contralateral testis, even if it is normally descended. The defective embryogene- sis hypothesis is further supported by the observation that orchiopexy, even at early age, does not appreciably decrease the risk of developing a tumor.[13]

Malignancy occurs 12-40 times more commonly in the undescended than in the descended testis.[22] The peak age for cancer in undescended testis is similar to that in scrotal testes, generally the third and fourth decades of life. The distribution of histologic types of tumors in undescended testes are similar to that in scrotal testes: pure seminoma, 43% ; embryonal cell carcinoma, 28%; Teratocarcinoma, 27% ; and chorio- carcinoma, 2%.[23] With appropriate management, the prognosis for seminomas arising in undescended testes is similar to that for seminomas arising in scrotal testes, i.e. excellent.[24]

Radiologic methods have been used in the preoperative localization of an undescended testis. The previously available imaging methods, such as testicular arteriography and venography are not only invasive but also technically difficult. They are also associated with high radiation dose and some morbidity.[25,26]

Ultrasonography has been used in the localization of an undescended testis within the inguinal canal but not within the pelvis or abdomen. An undescended testis will appear hypoechoic with ultrasonography. A bright echogenic band inside the testis, the medias- tinum testis must be identified for confident diagnosis. This is necessary because an enlarged lymph node can simulate an undescended testis but lacks this structure.[6] More importantly, agenesis cannot be discriminated from atrophy with US.[6,7]

Computed tomography scanning is easily reproducible and does not depend on operator skill. Detection of an undescended testis by CT is based upon the recognition of a mass, which is of soft tissue density and is oval in shape, along the expected course of testicular descending. In the inguinal area and lower pelvis where normal structures are usually bilaterally symmetrical, an extra soft tissue mass, even as small as 1 cm, can be detected. Higher in the pelvis or lower abdomen where bowel loops, vascular structures, and lymph nodes are more abundant, detection of such an atrophic testis and differentiation from adjacent structures may be more difficult. In young children, examination of the abdomen and upper pelvic struc- tures by CT may also be difficult due to lack of body fat.[8] CT cannot show the mediastinum testis and also lacks the specificity and sensitivity that are needed to diagnose agenesis.[14,27]

Magnetic resonance imaging has advantage of improved soft tissue contrast and can be used to detect undescended testis in both transaxial and coronal planes, but reports have varied as to its usefulness. [28,29] On MR imaging, the testis is typically of low signal on T1-weighted and of high signal on T2-weighted images. Undescended testes that are atrophic may not be of high intensity on T2-weighted scans.[27] The mediastinum testis is a band along the posterior margin of the testis. It is hypointense relative to the testicular parenchyma in both T1-weighted and T2 -weighted images.[30] The results of a study by Lam et al[31] showed that gadolinium-enhanced MR veno- graphy performed in conjunction with routine pelvic MR imaging increased sensitivity for differentiation of agenesis from ectopia.

There is an important potential pitfall that can cause confusion with the undescended testis. The par infravaginalis gubernaculi (PIG) can be similar to the undescended testis on any imaging study. It is a distal bulbous segment of the gubernaculum which is a cordlike structure that extends from the lower end of the testis to the scrotum and guides the testis in its descent. In early fetal development, the gubernaculum is soft and jellylike. When descent is completed, the

gubernaculum and par infravaginalis gubernaculi normally atrophy. If the testis fails to descend to the scrotum, the gubernaculum frequently persists as a fibrotic rather than gelatinous remnant. The PIG is always located distal to the undescended testis, usually in the scrotum but sometimes craniad to the scrotum. It is the latter case that the PIG and undescended testis may be confused. More importantly, cases with the PIG in the high scrotum or in the inguinal canal with surgically absent of the testes had been reported.[27]

Sonographically, the PIG appears as a hypoechoic mass with a cordlike structure of similar echogenicity led into it. The mediastinum testis, which can be identified in the normally descended and in some undescended testes, is absent in the PIG.[27]

The CT appearance of the gubernaculum and PIG is indistinguishable from that of the spermatic cord and testis. In the patient in whom the PIG is present distal to a normal testis, CT identifies two structures, allowing the correct diagnosis. Therefore, if scanning is started at the scrotum, it is important to continue to the renal hila, because the initial structure identified could be the PIG.[27]

On MR imaging, the PIG appears as a mass, which is isointesnse on T1-weighted and hypointense relative to the normal testis on T2-weighted images. These findings could not be reliably distinguished from an atrophic ectopic testis. The spermatic cord leading to the testis and gubernaculum leading to the PIG have identical properties on MR imaging. The mediastinum testis, which sometimes can be seen on MR studies, is the only finding that clearly identifies a structure as a testis.[27]

Diagnosing malignancy in an undescended testis can be difficult. The clinical presentation varies considerably and, when the tumor is in certain locations, often includes secondary symptoms from mass effect. Most commonly, patients seek medical attention for symptoms simulating appendicitis or retroperitoneal mass, urinary frequency or dysuria from mass effect on the bladder, or acute abdominal pain from tortion of the malignant growth.[20]

Tumor markers have a well-established role in the diagnosis of germ cell tumors. The two most clinically useful tumor markers are α-fetoprotein and β-human chorionic gonadotropin (**β-HCG**). α-fetoprotein is a protein produced early in gestation by the fetal liver, gastrointestinal tract, and yolk sac. It is a marker for hepatocellular carcinoma and germ cell tumors (nonseminoma with yolk sac elements). **β-HCG** is a glycoprotein produced by syncytiotrophoblasts of the developing placenta during pregnancy. Its level is elevated in tumors containing the syncytiotrophoblasts such as gestational trophoblastic disease and germ cell tumors, especially teratocarcinomas and choriocarcinomas. Both α-fetoprotein and β-human chorionic gonadotropin play crucial roles in the management of patients with nonseminomatous germ cell tumors. α-fetoprotein or **β-HCG** is elevated in 85% of nonseminomatous tumors. α-fetoprotein does not elevate in seminoma and dysgerminoma. **β-HCG** is occasionally elevated in seminoma and dysgerminoma.[30,32] These two tumor markers were not elevated in this reported case of seminomatous carcinoma.

The imaging findings of cancerous intraabdominal testes have not been well described. Sonography has been advocated to detect malignancy in undescended testes. It can be a solid slightly heterogeneous echoic mass anywhere along the path of descent from renal hilum to the inguinal canal. On CT examination, it can be a well circumscribed, homogeneous soft-tissue density mass without obvious evidence of necrosis or calcification.[20] However, areas of necrosis and small calcifications in a seminoma arising in the intraabdominal testes were also reported.[14,18] MR findings can show the mass as isointense to muscle on T1-weighted images, of relatively high signal intensity on T2-weighted and short inversion time inversion recovery images, and markedly enhanced on T1-weighted images after I.V. gadolinium

administration.[20]

The seminoma of this reported case is a large retroperitoneal mass, extending from the left renal hilum to the inguinal canal. It is located between the aorta and left psoas muscle. It is a well circumscribed mass with CT evidence of small calcifications and hypodense areas, which possible to be necrotic areas. No adjacent enlarged lymph node is noted. Although the differential diagnosis of a retroperitoneal mass might have included retroperitoneal fibrosis, lymphoma, metastatic adenopathy, paraganglioma, liposarcoma, leiomyosarcoma, and malignant fibrous histiocytoma. None of the previously mentioned diagnoses could reveal as a large mass extending from renal hilum to the ipsilateral inguinal canal. The only clue that makes the most likely diagnosis carcinoma arising in an undescended testis is a single mass locating within the nearly entire path of testicular descent.

Therefore, when a retroperitoneal or pelvic mass is detected in a male patient, tumor arising in an undescended testis might be included in the differential diagnosis. The helpful information is history taking about the lack of a testis in the scrotum. Past history of infertility is also helpful in the diagnosis, in a patient with bilateral undescended testes. Tumor markers may useful in the diagnosis of nonseminomatous germ cell tumors but they show no benefit in seminoma.

A small mass with central hypodensity in the right groin of this patient was detected by CT scan. This mass was located in the right inguinal canal, just proximal to the external inguinal ring (Fig 3F). It can be distinguished from an enlarged lymph node because an enlarged groin lymph node is located inferolateral to the inguinal ligament or adjacent to the femoral and iliac vessels.[33]

The differential diagnosis of a right-sided inguinal canal mass in this patient might have included atrophic testis and par infravaginalis gubernaculi. These two conditions cannot be distinguished by imaging. Although, there is no soft tissue mass in the more craniad sections along the path of right testicular descent, the PIG is still possible, because this may be due to small atrophic right testis which could not be demonstrated by imaging or due to agenesis of right testis.

CONCLUSION

An undescended testis can be found any where along the path of testicular descent from renal hilum to the upper part of scrotum. Atrophic undescended testis has similar radiologic features with the par infravaginalis gubernaculi. An important CT feature of a seminoma arising in an undescended testis is a retroperitoneal or pelvic soft tissue mass with or without internal calcifications and necrotic areas.

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