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TESTICULAR SARCOIDOSIS

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We describe a very unusual form of sarcoidosis of the testis, mimicking malignancy at initial presentation. Genitourinary sarcoidosis is rare and this case report emphasizes the importance of meticulous analysis of the patient's clinical history combined with imaging findings and specific pathological criteria to diagnose this granulomatous disorder.

Key-word: Sarcoidosis.

Case report

A 40-year-old Caucasian male patient was referred by his general physician to the hospital for the assessment of a palpable painless mass of the right testis and a dry cough with mild dyspnea for 6 weeks. The pulmonary complaints improved with non-steroidal anti-inflammatory drugs. The patient had no fever and no B-symptoms. There was absence of weight loss. The previous medical history and routine blood examination were unremarkable. Clinical examination confirmed the right testicular mass but also an enlarged mass at the right inguinal region. Scrotal ultrasound showed bilateral, well delineated hypoechoic lesions within the testicular parenchyma (Fig. 1). Color-Doppler showed only subtle intralesional vascularisation. Multiple enlarged inguinal lymph nodes were seen bilaterally. A chest X-Ray showed bilateral hilar lymphadenopathies and multifocal lung nodules (Fig. 2). Computed tomography (CT) showed multiple irregular delineated pulmonary micro- and macronodules with peribronchovascular distribution pattern, irregular septal thickening and multiple mediastinal lymph nodes. On the abdominal series, a hypoattenuating splenic lesion (3 cm) and multiple enlarged mesenteric, retroperitoneal and inguinal lymph nodes were seen (Fig. 3). Laboratory results including serum angiotensin converting enzyme (389 U/L - normal value 115-419 U/L) and specific tumor markers (lactate dehydrogenase (340 U/L - normal value 237-475 U/L), serum alpha-fetoprotein (1,8 µg/L - normal value < 6,1 µg/L) and human chorionic gonadotropin (< 1,0 U/L - normal value < 2,5 U/L)) were within normal ranges. Based on the clinical history and imaging studies, the differential diagnosis included testicular malignancy with metastatic spread to

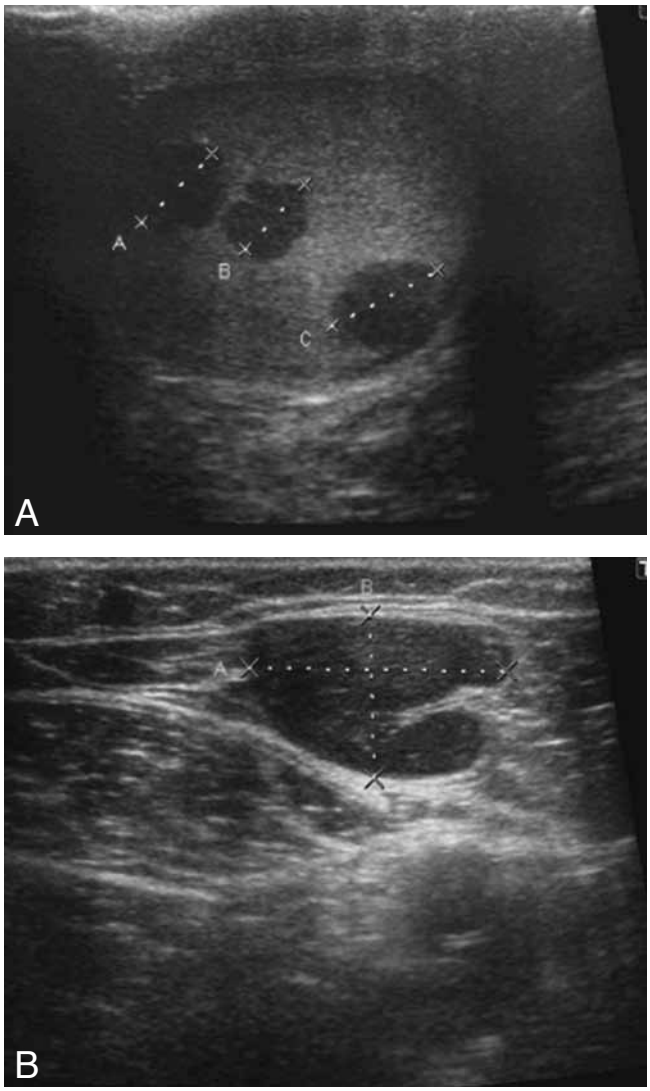

Fig. 1. — Ultrasound of the right testis (A), right inguinal lymph node (B). Image A demonstrates a normal volume of the testis and multiple sharply demarcated hypoechoic intratesticular nodules. Image B demonstrates the pathological enlarged right inguinal lymph node from which a biopsy specimen was obtained.

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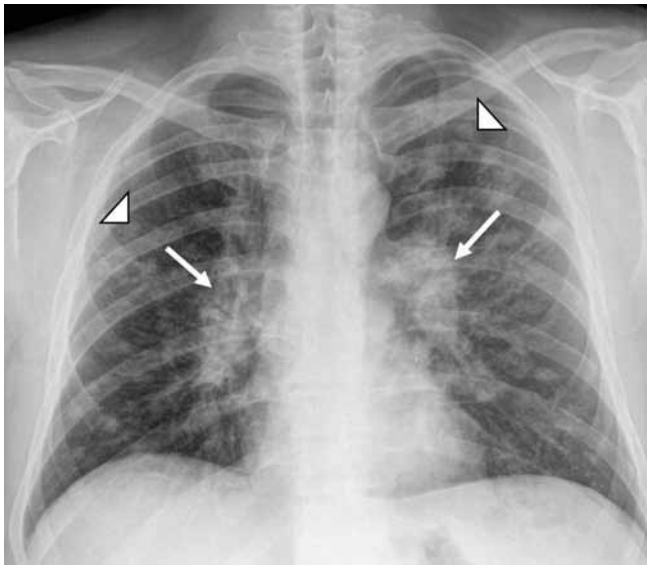

Fig. 2. — Postero-anterior radiograph of the thorax displays bilateral hilar adenopathies (arrows) with multiple nodular opacities bilaterally (arrowheads) with dominant predilection for the upper lobes.

abdominal lymph nodes and the lungs. This however was contradictory with the lab results. The differential diagnosis further included lymphoma and systemic disease such as a granulomatous disease. Based on the routine blood examination, chronic infectious process such as tuberculosis was unlikely. Finally, histological examination of a biopsy specimen from an inguinal lymph node revealed the diagnosis of sarcoidosis. Retro-anamnesis revealed itchy lesions at the lower limbs several weeks ago consistent with erythema nodosum. Apparently, treatment with topical corticosteroid therapy relieved the complaints and the precise etiology of the skin lesions remained unclear after this clinical contact. Further pulmonary work up was consistent with the diagnosis of sarcoidosis: spirometry showed a slightly decreased total lung capacity suggesting a restrictive lung pattern with normal diffusion capacity. No

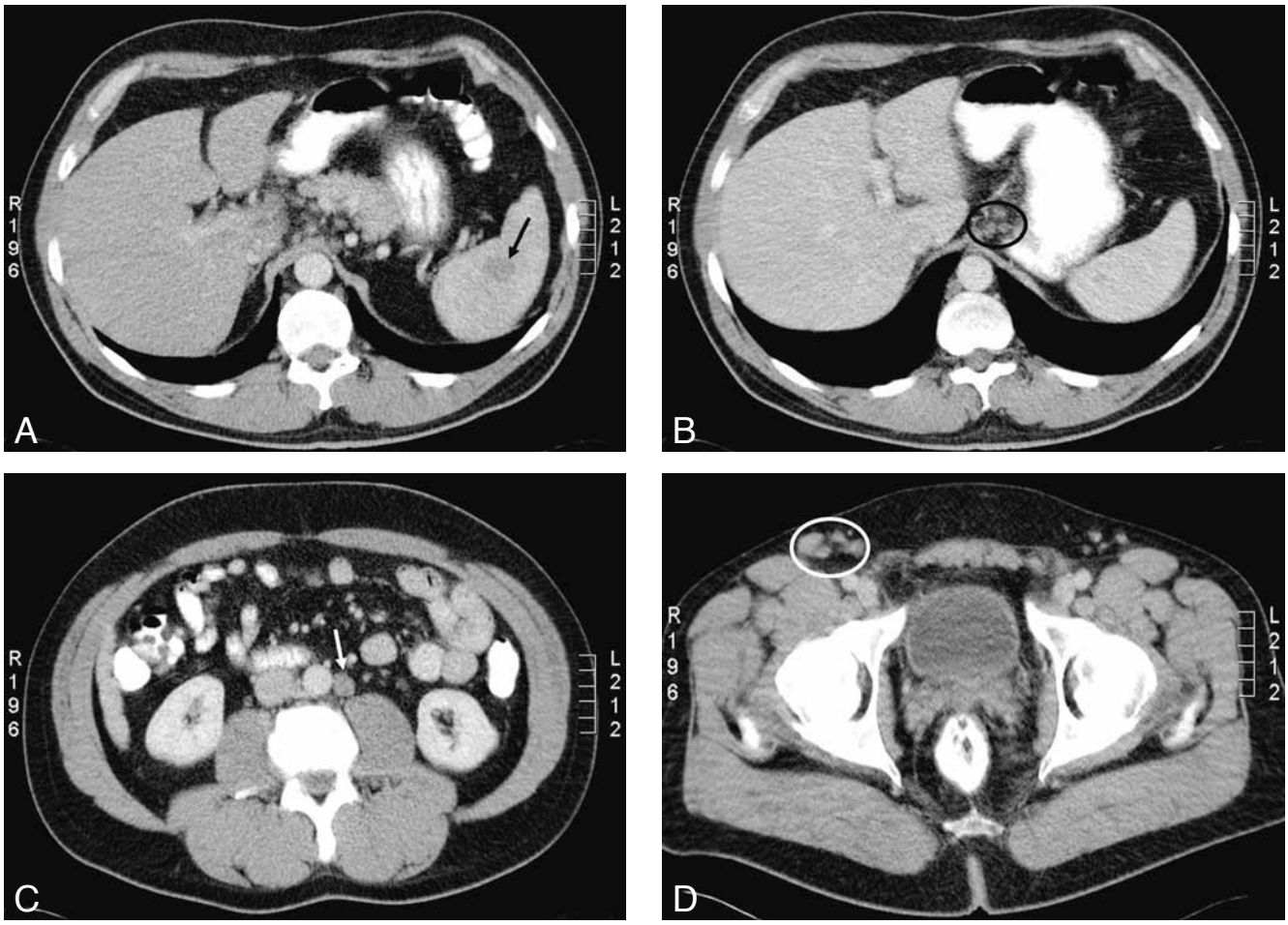

Fig. 3. — Axial (A, B and C) contrast-enhanced CT of the abdomen and pelvis (D). Figure A demonstrates clearly a well demarcated hypodense intrasplenic lesion 3 cm in size. Image B demonstrates multiple enlarged lymph nodes at the retroperitoneum and gastrohepatic ligament (circle) and image C an enlarged paraaortic lymph node (arrow). Figure D demonstrates the pathological lymph node aggregate (circle) in the right inguinal region from which the biopsy was obtained.

other vital organs were involved and the patient is in follow up every 4 months.

Discussion

Sarcoidosis is a systemic granulomatous disease of unknown multifactorial etiology in which the combination of an unbalanced immune system, genetic predilection and environmental impact are key factors (1). The disease primarily affects young African-American female adults. The estimated prevalence is 1 to 6 cases per 100000 people worldwide with a peak incidence that occurs between the third and fifth decade. The histological hallmark is the occurrence of non-caseating epitheloid granulomas and multinucleated giant cells (2). The final diagnosis is based on the combination of clinical, radiological and histological features and after exclusion of other pathologies with similar histology characteristics (3). In cases where a biopsy cannot be obtained, diagnosis is based on the clinical and radiographic features in addition to certain demographic information (4). In sarcoidosis, pulmonary and lymph node involvement are the two most frequently affected organ sites. Extrathoracic involvement occurs in 15-45% of patients, involving many organs including the spleen, liver, skin, eyes, muscles, central nervous system and salivary glands (5). Male genitourinary involvement is an unusual manifestation of this granulomatous disease reported in approximately 0,2% of all sarcoidosis patients (6). The incidence of genital sarcoidosis at necropsy series is less than 5% (7).

Imaging features of pulmonary sarcoidosis on conventional radiographs and CT include bilateral hilar adenopathies and involvement of lung parenchyma (8). Further discussion of the pulmonary imaging features is beyond the scope of this case report.

Abdominal sarcoidosis may mimic other systemic diseases such as lymphoma, tuberculosis or metastases with hepatosplenomegaly being the most frequent imaging finding. Nonspecific focal hepatosplenic lesions (8-10), as demonstrated in our patient, are less frequent (Fig. 3).

For evaluating the scrotal content, ultrasonography is the key imaging modality. Multiple hypoechoic and hypovascular lesions that synchronously affect the epididymis and testis are typical for testicular sarcoidosis. These lesions are rather small (ranging from a few millimeters to 3 cm), nodular and sharply demarcated. When bilateral hypoechoic testicular solid nodules are found, the differential diagnosis is wide. Although testicular tumors represent only 1% of all malignant neoplasms in men, it is the most common malignancy in men aged 15-34 years (11) and this is the same age group in which sarcoidosis of the testis is most frequent (12). However, in our patient the presence of a primary testicular tumor is unlikely since the prevalence of bilateral testicular neoplasms is very low (1-3%). Bilateral multifocal testicular tumors mimicking testicular sarcoidosis include seminoma, Leydig cell nodules (Leydigiomas), adrenal rests, chronic granulomatous disease processes, lymphoma, leukemia and metastasis (11). Seminoma may be bilateral in approximately 3% of the cases. Multiple and bilateral Leydig cell nodules are incidental findings in patients referred for subfertility. Adrenal rests are part of a syndrome and present as tail-shaped lesions located near the rete testis. Lymphoma and leukemia usually have a sonographic appearance of bulky masses involving major parts of the testicular parenchyma, which is clearly not the case in our patient. Also, metastasis to the testis has been reported and needs to be considered (melanoma, gastric cancer and prostate carcinoma).

In addition, the absence of specific tumor markers as lactate dehydrogenase, serum alpha-fetoprotein and human chorionic gonadotropin are helpful in differentiating sarcoidosis from primary testicular tumor. The distribution of lymph nodes is also helpful for further narrowing the differential diagnosis. Inguinal lymph node metastases would indicate scrotal involvement of a primary testicular tumor, which was completely ruled out based on the imaging studies. The sentinel nodes for testicular tumors are the retroperitoneal areas near the renal hilum (11).

Conclusion

Genitourinary sarcoidosis is extremely rare and ultrasound is the primary imaging modality to evaluate the scrotal content. The combination of a detailed patient history, laboratory results and documentation of multisystem involvement on chest X-rays and CT of the chest and abdomen allow narrowing of the differential list. Histological confirmation is essential for a final diagnosis.

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