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A Hard Ball for a Tennis Player: A Rare Case of Large Calcifying Sertoli Cell Testicular Tumor

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ABSTRACT

A 46 year old tennis player was addressed to our clinic after incidental finding of right testicular calcification on plain x-ray of the spine. Urologic consultation revealed a hard non-tender testicular mass which required inguinal orchiectomy. Final histology revealed large cell calcifying Sertoli cell tumor: we herein present the case and review current physiopathology of such rare testicular disease.

Introduction

Sertoli cell tumors are a rare entity and represent around 1% of all testicular tumors in men. Various histologic subtypes are described, in particular sclerosing Sertoli cell tumor, large cell calcifying Sertoli cell tumor (LCCSCT) and Sertoli cell tumor not otherwise specified. This tumor is usually non-malignant and is frequently associated to genetic syndromes Peutz-Jaghers and Carney complex. To date, less than 100 cases have been reported. We herein describe a particular case which was firstly diagnosed on plain X-ray of the lombosacral column, thanks to the intense calcifications of the lesion.

Case presentation

A 46 year old male, active tennis player, presented lumbar osteomuscular pain while playing tennis. His family doctor had prescribed plain X-rays of the vertebral column. The films were inconclusive on spinal pathology but detected abnormal right testicular calcifications (Fig. 1), and therefore the patient was referred to our urology clinic. On clinical examination the patient presented a hardened, non-tender right testicle, suspicious for neoplasia. No clinical lymphadenopathy was palpable. Patient was in excellent health condition, presented no significant co-morbidity and did not assume any medication. A testicular ultrasound was performed which confirmed the presence of a hypervascularized mass, of the size 44 mm × 30 mm, with multiple calcifications. Although this is not currently recommended in initial staging for

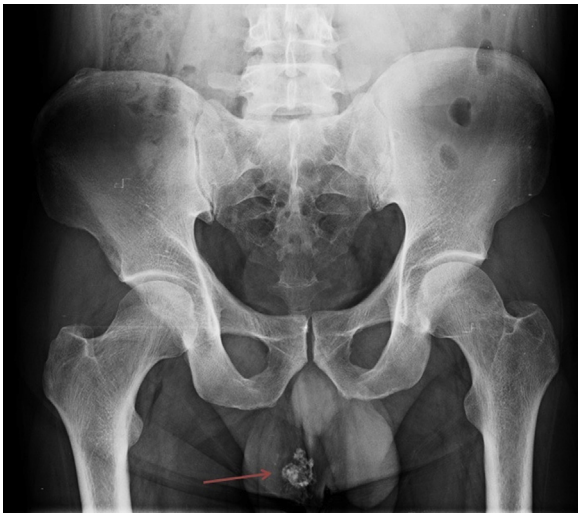

Figure 1. Plain X-ray finding incidental right testicular macrocalcification, which lead to diagnosis. Arrow pointing at calcifications.

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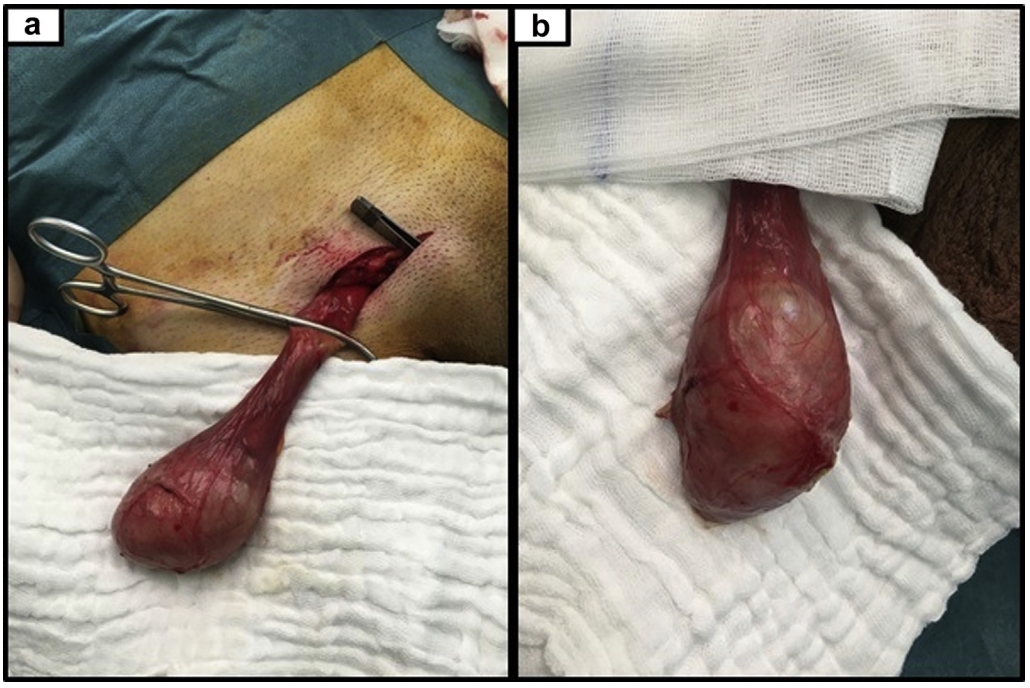

Figure 2. a. Intraoperative view of the dissected right testis. b. Close up.

testicular mass, a PET-FDG scan was performed prior to surgery which found a highly hypermetabolic lesion confined to the right testis, with no evident distant metastasis.

The patient was then scheduled to undergo radical inguinal orchiectomy (Fig. 2). Surgery and post-operative recovery were uneventful. Anatomopathologic evaluation of the specimen (Fig. 3) reported the presence of a calcified, well delimited mass measuring $40 \times 38 \times 30$ mm, with no infiltration of the vaginal tunic. A large intratumoral calcification was found, and on microscopic examination, osteoid metaplasia was observed. Immunostaining with placentar phosphatase alcaline was negative, confirming the absence of intratubular germ cell neoplasia in the rest of the testicle. Final pathologic diagnosis was therefore large calcifying cell Sertoli cell testicular neoplasia, with complete excision (Fig. 4). A genetic analysis was performed via an Illumina Truseq Amplicon Cancer Panel kit, finding no mutation of the STK11 gene (Peutz-Jaghers syndrome). Similarly, no mutation was found in other genes including FGFR3, ATM, or p53.

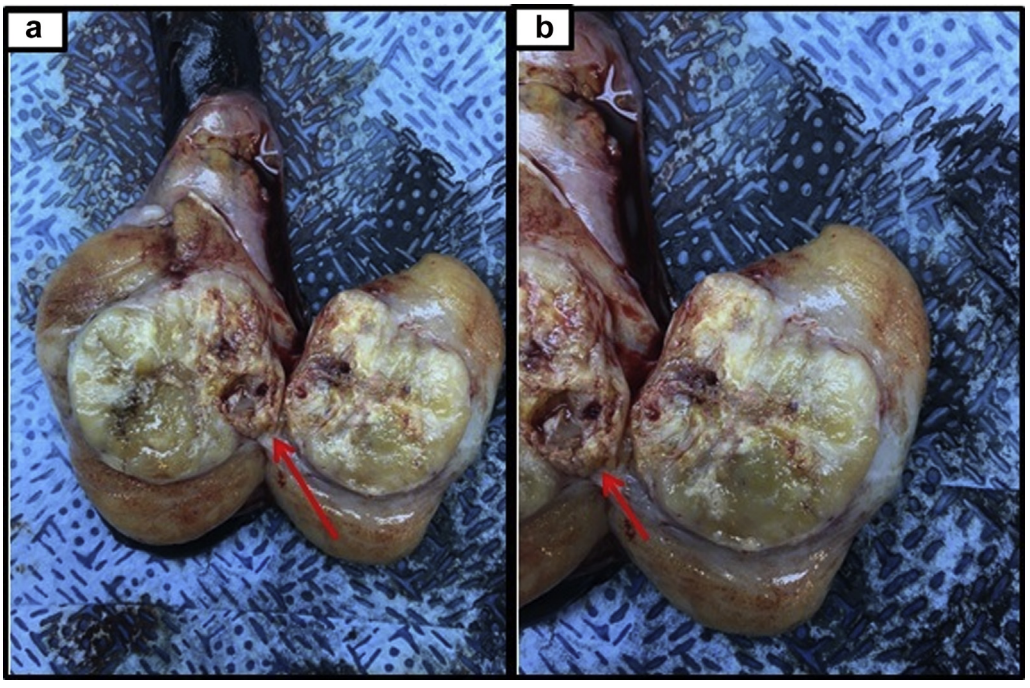

Figure 3. a. Macroscopic intratumoral histology. Arrow pointing at calcifications. b. Close up.

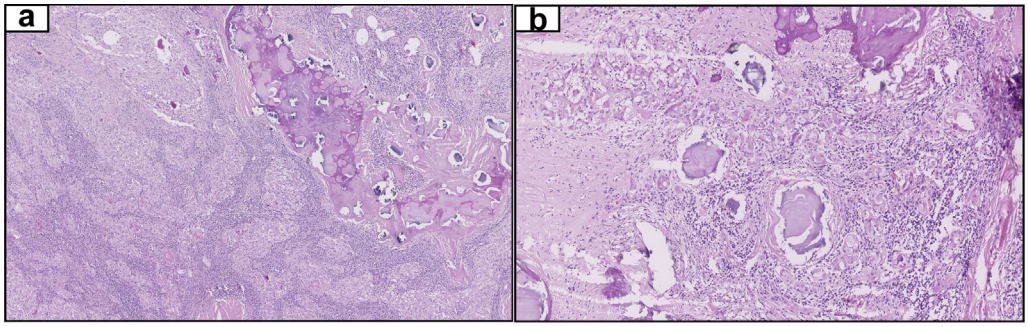

Figure 4. a. H&E slide of the histology (×50). b. Close up (×100).

Discussion

LCCSCTs are a rare entity which was first described in 1980 by Proppe and Scully, who reported on 10 cases.[1] These cancers represent a frequently benign pathology, more common in young boys. LCCSCT usually present a distinctive histology: an intratubular growth pattern, with hyalinization of the tubular basement membrane, with cells arranged in circumscribed nodules. Large round Sertoli cells provided with abundant cytoplasm are typical, as well as massive calcifications which are fundamental for the diagnosis of LCCSCT.[1] This tumor can be multifocal and bilateral in about 25% of cases and is frequently associated to extragonadal endocrine symptoms. Some of these cancers possess hormonal synthesis capacity, as has been demonstrated by Perez-Atayde on immunehistochemical marking. Malignant cases with consequent metastases are rare, accounting for around 5% of cases: these are usually detected in older patients and present with large lesions (>4 cm), lymphovascular invasion, necrosis and a high mitotic index, which was not the case for our patient. Importantly, these lesions are frequently associated to genetic syndromes of Peutz-Jaghers and Carney complex in about half the cases.

Peutz-Jeghers syndrome is an autosomal-dominant syndrome, frequently characterized by a mutation in *LKB1/STK11* gene.[2] Clinical characteristics include hamartomatous polyps of the gastrointestinal system and skin hyperpigmented melanin depositions. Patients with Peutz-Jeghers syndrome have a 15-fold increased risk of developing intestinal cancer compared to general population. It was first described in 1921 by Jan Peutz who described the syndrome in a Dutch family.[2] Carney syndrome is also inherited in an autosomal dominant fashion, with a common inactivating mutation of the PRKARIA gene.[3] It presents with spotty cutaneous pigmentation, endocrinopathy and the following tumors: cardiac myxomas, pituitary adenomas, Sertoli cell tumors, thyroid cancer and schwannomas.[3] Interestingly, in our case, genetic work up revealed no aberrant genetic mutation.

Our patient was referred to urology due to testicular macrocalcifications. Intratesticular calcifications are a frequent finding in men, with a prevalence of over 5%. Their etiopathology is unclear, though they may be a consequence of spermaticitic cell degeneration and failure of phagocytosis by Sertoli cells, which in turn determines an immune inflammatory reaction.[4] Another hypothesis is the increased permeability of the basal membrane with formation of interstitial calcified depositions.[5] Currently the association between testicular microcalcifications and cancer remains controversial, although there is a link between the two pathologies as conformed by multiple studies. Current guidelines advise to perform standard physical examination with no extra exams in men with testicular calcifications and no other risk factors. In patients with a history of testicular cancer and contralateral calcifications instead, no clear guideline is available. Experts generally advise tight surveillance, ultrasound and eventually testicular biopsy in case of doubt. In the current case report, the patient clearly presented a testicular mass suspect for neoplasia, and as such underwent inguinal orchiectomy. Nonetheless, cases of organ-sparing surgery have been documented with good oncologic outcomes in LCCSCTs, and should be considered for younger patients with bilateral disease, given the frequent benign behavior of this tumor.

Conclusions

We herein report a case of LCCSCT with massive intratumoral calcifications, detected hazardously on a plain X-ray, describing its pathophysiology and histologic characteristics. We hope that the present report will help in future diagnostic and treatment decisions.

Conflict of interest statement

The authors declare that there is no conflict of interest regarding the publication of this paper.

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