Primary retroperitoneal seminoma: an unusual cause of testicular pain

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Introduction

Germ cell tumours in men usually arise from the testes, with only 1–2% originating from other locations.1 These so-called primary extra-gonadal germ cell tumours (EGCTs) are rare, accounting for 0.15–0.2% of all malignancies.2 They are characterized by their midline location, and have been reported to occur anywhere from the pineal gland to the coccyx, with the commonest locations being the mediastinum and the retroperitoneum.3 Lack of awareness of this condition, together with its rarity, often results in diagnostic delay. The commonest presenting symptoms of retroperitoneal germ cell tumours (GCTs) are abdominal and back pain, but can include weight loss, night sweats, venous thrombosis and scrotal oedema among others.3 However, there have been no reported cases of testicular pain as a presenting complaint. Here we report such a case, and highlight the difficulties in diagnosis.

Case report

A 47-year-old man initially present to the emergency department with a history of sudden-onset, intermittent left testicular pain which had been present for a few weeks. He denied any systemic symptoms and had no family history of note. General physical examination was unremarkable, and examination of his testes revealed no abnormality. Routine laboratory tests for full blood count, urea and electrolytes, and C-reactive protein were within normal limits. A testicular ultrasound scan also failed to show any abnormality in either testis and a renal tract ultrasound scan was reported as normal. He was treated with a course of antibiotics for suspected epididymo-orchitis, but this failed to relieve his symptoms.

It was subsequently thought that he may have a distal ureteric calculus causing referred pain to the testicle, and so he underwent a CT scan of his renal tract, almost six months after the initial presentation. This revealed a 10 × 12 cm retroperitoneal mass (Figures 1 and 2). CT-guided biopsy was non-diagnostic, and so he underwent laparotomy and biopsy, the histology of which showed classical seminoma. Tumour markers of LDH, β-HCG and alpha feta protein were within normal limits.

He underwent four cycles of cisplatin-based chemotherapy with complete response.

Discussion

Testicular pain can be caused by a number of conditions, but there are currently no reported cases of EGCTs presenting solely with pain in the testicles. Common investigations for patients presenting with testicular pain include urine microscopy, serum inflammatory markers, tumour markers if indicated by clinical findings, and a testicular ultrasound scan. This case highlights that even if all investigations are normal, persistent symptoms should be further investigated with a CT scan of the abdomen and pelvis to exclude a rarer cause, and reduce the time delay to diagnosis. An unenhanced CT is now frequently the first-line investigation for patients presenting to the emergency department with features of acute ureteric colic, and has the added advantage of being able to identify non-urologic causes of pain. It has been suggested that this
form of imaging can identify other significant pathologies in up to 12% of cases.4

Retroperitoneal masses can be caused by a range of benign and malignant conditions. Commonly, these include metastatic lymphadenopathy, lymphoma and liposarcoma. However, retroperitoneal germ cell tumours are rare lesions, accounting for only 4.4% of all malignant primary retroperitoneal tumours.5 Half of these are seminomatous in nature. During embryogenesis, germinal epithelium migrates in the midline from the urogenital ridge to the scrotum. It has been suggested that failure of this migration may ultimately lead to the formation of primary germ cell tumours.6 Alternatively, it has been postulated that these tumours may arise in germ cells that are normally distributed in midline locations throughout the body to perform various functions.7

Determining whether a tumour is a primary retroperitoneal malignancy or a metastatic tumour from an occult or regressed testicular primary is often difficult, but it is important to establish as recurrence in the testes have been reported up to 18 years after removal of the original retroperitoneal malignancy.8 Previous reporters have attempted to set criteria to aid in distinguishing these two possibilities,9 and radiologically it has been suggested that midline tumours are suggestive of a primary retroperitoneal tumour whereas lateral tumours suggest a primary in the ipsilateral testis. Ultrasonic features to suggest a regressed testicular tumour as the cause (burned-out phenomenon) include echogenic foci, microlithiasis, scar formation and inhomogeneity.10 However, all these features are non-specific and not always present. Therefore, if any testicular abnormality is found on ultrasound or clinical examination in combination with a retroperitoneal mass then the patient should undergo ipsilateral inguinal orchidectomy for careful histological analysis. If all testicular investigations are normal then the retroperitoneal tumour can be considered a primary. However, in this situation, we recommend that the patient be advised to perform regular testicular self-examination to identify any testicular recurrence early.

A study characterizing the clinical features of retroperitoneal germ cell tumours showed that they may present with a variety of symptoms.3 The most common presenting features are abdominal and back pain, but in a smaller proportion of patients symptoms can include weight loss, fevers/night sweats, venous thrombosis, a palpable mass, a hydrocoele, and rarely haematuria. The non-specific nature of these symptoms often leads to a delay in diagnosis. In our case, the patient presented with testicular pain but no clinical, biochemical or radiological testicular abnormality was found. A CT scan was only performed to exclude the possibility of a ureteric calculus causing his testicular pain, and led to the finding of a retroperitoneal mass. This highlights the need to be aware of the possibility of EGCTs in male patients presenting with ongoing non-specific symptoms that could be related to the genitourinary system, where initial investigations have failed to identify any abnormality.

**Conclusion**

Retroperitoneal seminomas are rare tumours, and it is difficult to know whether they are true
primary tumours or metastases from an occult or regressed testicular primary. In many cases, patients may present with a variety of symptoms and imaging findings are often non-specific. Consequently, the diagnosis is commonly overlooked initially. We recommend that male patients with persistent non-specific genitourinary symptoms and normal testicular investigations undergo a CT scan to exclude the possibility of a retroperitoneal mass. Furthermore, if no testicular abnormality is found in patients confirmed to have an EGCT, regular testicular self-examination is advised to identify the development of tumour recurrence early.

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