CASE REPORT

Retroperitoneal seminoma, a rare cause of testicular pain

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SUMMARY

Extragenital retroperitoneal seminomas are a rare cause of testicular pain in males. We report the case of a 46-year-old man, who presented with right-sided testicular pain, clinically mimicking testicular torsion. The patient underwent a scrotal exploration, which revealed normal testes. He went on to have a contrast-enhanced CT abdomen and pelvis, which revealed a 55 mm para-aortic mass, displacing the duodenum and inferior vena cava, with an adjacent 22 mm lymph node. Histological analysis revealed that it was a seminoma.

BACKGROUND

There are only a few reported cases of primary retroperitoneal seminoma presenting with testicular pain. Seminomas typically arise from the testis. Those that arise elsewhere are known as extragenital germ cell tumours (EGCT) and account for only 1%–2% of germ cell tumours.¹ They are thought to occur from the transformation of primordial germ cells, which have been mislaid during the migration through the midline dorsal mesentery in embryogenesis.² They are typically found anywhere within the midline, most commonly in the mediastinum and retroperitoneum.¹

Clinical diagnosis of an EGCT can be challenging and is most commonly prompted by signs and symptoms of compression secondary to a growing mass. The most common symptoms include abdominal and back pain. Diagnosis can then be confirmed

with fine needle aspiration cytology or histology.³ It is important to be aware of atypical presentations as in the case described in order to avoid incorrect and delayed diagnosis. There should be a lower threshold for investigating patients with persistent testicular pain in the presence of a normal testicular examination, due to referred pain from retroperitoneal tumours to the testis.

CASE PRESENTATION

A 46-year-old man was admitted to the local accident and emergency department, complaining of a sudden onset of acute right-sided testicular pain. He did not have any urinary symptoms (dysuria, frequency or haematuria), fever, weight loss, altered bowel habit, nausea or vomiting. There was no significant medical history or family history. His vital signs were entirely normal. On examination, he had a soft and non-tender abdomen. No masses were palpable. There was no evidence of an inguinal or femoral hernia and testicular examination was unremarkable, besides severe pain on palpation of the right testicle.

INVESTIGATIONS

Initial investigations revealed an increase in C-reactive protein (76 mg/L) only. There were no significant abnormalities within the full blood count, urea and electrolytes or liver function tests. A urine dipstick was found to be negative for blood, protein or white cells, making renal colic and epididymo-orchitis less likely as a cause. Plain X-rays of the chest and abdomen and an ultrasound of the testes were carried out, which showed no significant abnormality.

Given the timing of onset and severity of right testicular pain, with no other causative pathology identified, the patient underwent an immediate scrotal exploration. This showed normal testes. Due to the suspicion of testicular torsion, ultrasound imaging of the abdomen was not undertaken prior to exploration. However, if there had been significant diagnostic doubt, an ultrasound abdomen and pelvis, including the renal tract could have been carried out to rule out possible intra-abdominal pathology or renal colic.

Subsequent CT imaging of the abdomen and pelvis with contrast ([figure 1](#)) revealed a 55 mm mass in the aortocaval region, with consequent displacement of the adjacent duodenum and inferior vena cava (IVC), suggestive of a retroperitoneal tumour. Adjacent to the mass, there was also a

Figure 1 CT abdomen and pelvis with contrast (coronal plane). The 55 mm retroperitoneal tumour is visualised in the aortocaval region, with consequent displacement of the adjacent duodenum and inferior vena cava.

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Figure 2 CT-guided biopsy. The biopsy needle is visualised entering the retroperitoneal tumour via a percutaneous approach.

22 mm lymph node and small subcentimetre lymph nodes within the adjoining mesentery.

The patient underwent a CT-guided core biopsy of the para-aortic mass (figure 2). This showed fibrovascular connective tissue containing scattered CD3+ T lymphocytes together with cores of necrotic cells expressing optical coherence tomography (OCT) 3/4 surface proteins. The appearances were suggestive of a malignant tumour; however, the necrosis precluded a firm diagnosis and further biopsies were recommended for analysis.

DIFFERENTIAL DIAGNOSIS

The main differential diagnosis at initial presentation was testicular torsion. This is an uncommon diagnosis in someone of this male's age but in the presence of acute onset of severe testicular pain and the absence of features to suggest epididymo-orchitis and renal colic, this was the diagnosis of exclusion.⁴ Epididymo-orchitis can also present with testicular pain, which can be severe, but it is typically associated with swelling secondary to inflammation of the ipsilateral epididymis and/or testes. In addition, the patient may be feverish, with leucocytes and/or nitrites in their urine and raised inflammatory markers. Renal colic typically presents with loin to groin pain, with associated microscopic haematuria. Clinically, there was no evidence to suggest epididymo-orchitis or renal colic.

Having confirmed a retroperitoneal mass, our initial differentials included lymphoma, sarcoma, metastatic lymph nodes¹ or an extragonadal tumour (seminomatous/non-seminomatous). On histological analysis, the tumour was found to be composed of discohesive polygonal cells, with associated lymphohistiocytic inflammatory infiltrate. These stained positive with OCT 3/4 surface protein and were focally placental-alkaline phosphatase positive, for which the differentials could include seminoma, embryonic choriocarcinoma or yolk sac tumour. However, the immunohistochemical stains for CD30, cytokeratin monoclonal antibody (MNF116), alpha feta-protein (AFP) and beta-human chorionic gonadotropin (b-HCG) were negative, therefore ruling out a non-seminomatous tumour.

Serum tumour markers for EGCTs including AFP and b-HCG were not raised. The lactate dehydrogenase level was 159 units/L. A final diagnosis of seminoma was confirmed based on

the morphological appearance, immunohistochemical staining and the absence of raised serum tumour markers.

TREATMENT

As our main initial differential diagnosis was of testicular torsion, we undertook a scrotal exploration under general anaesthesia. Although this revealed normal testis, the patient was symptomatically better after the procedure.

His case along with his CT images and biopsy findings were discussed in the local multidisciplinary meeting. The consensus was for an elective laparotomy to excise the mass. This was a joint case between the urologists and vascular surgeons, given the close nature of the mass to the IVC.

Intraoperatively, he was found to have a dense 3–4 cm mass, adherent to the IVC and aorta at the origin of the superior mesenteric artery. The mass was mobilised and completely excised. The right testicular vessel was ligated and the resultant specimen sent for histology.

OUTCOME AND FOLLOW-UP

The histology from the excised mass showed necrotic tissue, originating as a seminoma. The histology from the lymph node also confirmed this. Due to the absence of any testicular disease, the impression was that the mass had originated as a retroperitoneal seminoma, rather than metastasising secondary to a primary tumour in the testes.

The patient made a good recovery postoperatively, with complete resolution of his testicular pain following excision of the retroperitoneal mass and decompression of the gonadal vasculature. At his 4-week follow-up appointment, he remained clinically well and reported no further symptoms. He did not require bilateral prophylactic orchidectomies as his testes were normal during previous scrotal exploration and on a repeat scrotal ultrasound scan.

The oncologists reviewed the patient, who expressed a wish to forego further chemotherapy. Follow-up CT imaging 6 months postsurgery showed no evidence of recurrence. In the absence of raised tumour markers with complete surgical resection of the tumour, the patient elected for high-risk surveillance over chemotherapy. This would entail 3-monthly blood tests and a further CT abdomen and pelvis with contrast, after a 6-month interval.

DISCUSSION

EGCTs localising in the retroperitoneum are rare. They account for only 4.4% of all malignant primary retroperitoneal tumours.⁵ As in this case, 50% are found to be seminomas on histology. The process by which they appear is through failed migration of the germinal epithelium in the midline during embryogenesis, from the urogenital ridge to the scrotum.¹

The median age for presentation of seminomatous EGCTs is 41 years, similar to that of primary gonadal tumours.³ Typical clinical symptoms of seminomatous EGCTs from most common to rare, include abdominal pain, back pain, weight loss, fever, vena caval thrombosis, scrotal oedema and dysphagia.³ Diagnosis can be further supported with serum tumour markers; 50% of retroperitoneal seminomas are associated with raised serum AFP and 74% are associated with raised serum b-HCG.³

There is only one other documented case study of a retroperitoneal seminoma presenting primarily with testicular pain.¹ This is thought to occur from compression of the gonadal neurovasculature, resulting in cremasteric spasm.⁵ As in this case, retroperitoneal masses are often identified only after normal testicular investigations with persistent symptoms. These include blood tests, tumour

markers, urinalysis, ultrasound and even exploration. Frequently, it is only as an incidental finding on a CT abdomen and pelvis that a retroperitoneal neoplasm is identified.

Management of a retroperitoneal mass includes complete surgical excision⁶ and histological analysis to confirm diagnosis. In addition, adjuvant chemotherapy consisting of cisplatin, etoposide and bleomycin is recommended for all patients, particularly in the presence of positive tumour markers. A minimum of three cycles are recommended for those patients considered to have a 'good' prognosis as opposed to four for those with an 'intermediate/poor' prognosis. There is no role for radiotherapy in current clinical practice, except for salvage therapy in those patients with refractory disease.³

During the multidisciplinary stage, it is important to establish whether the tumour is a primary retroperitoneal neoplasm, or secondary to a metastatic primary elsewhere, for example, a primary testicular seminoma. Features on ultrasound of the testes to suggest primary malignancy would include echogenic foci, microlithiasis and heterogeneous tissue.⁷ In this case, the testes showed no abnormality on examination or on ultrasound imaging; therefore, the retroperitoneal tumour was considered to be a primary neoplasm. It has also previously been advocated that a midline retroperitoneal tumour is suggestive of a primary retroperitoneal tumour, whereas a lateral tumour would suggest a metastatic retroperitoneal tumour secondary to a primary in the ipsilateral testis.⁸

Retroperitoneal seminomas commonly metastasise to the retroperitoneal lymph nodes (26%), mediastinal lymph nodes

(12%) and cervical lymph nodes (6%), whereas non-seminomatous tumours have a higher frequency of visceral metastases, particularly to the lungs (49%).³ As they are often only detected due to local compression of adjacent structures, retroperitoneal tumours can be quite advanced at the time of presentation. However, the 10-year survival of a primary EGCT is approximately the same as a primary gonadal tumour (88%). The presence of positive tumour markers and/or metastases at the time of diagnosis can reduce survival.³

It is important to be aware that the risk of developing metachronous testicular cancer 10 years after diagnosis is 1.4% in seminomatous EGCTs.³ Therefore, it is imperative that these patients undertake routine testicular examination.

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Learning points

- ▶ Patients presenting with persistent testicular pain with a normal ultrasound, urinalysis and blood tests should be considered for further imaging of the retroperitoneum either by means of an ultrasound or CT scan.
- ▶ All male patients presenting with a primary retroperitoneal mass should undergo regular self-examination of the testicles to allow early detection of malignancy.
- ▶ Treatment of a retroperitoneal seminoma should include complete surgical excision with adjuvant chemotherapy to prevent tumour recurrence.

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