SHORT REPORT

Extraspinal Ependymoma: An Unusual Cause of Deep Vein Thrombosis

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Introduction. Extrinsic compression of the iliac veins by neoplasms is a rare cause of deep vein thrombosis (DVT). We report a case of DVT heralding the presence of an unusual tumour, an extraspinal ependymoma.

Report. A 39 year old female presented with a left leg DVT, initially presumed to be idiopathic. However, subsequent investigations indicated by persisting symptoms revealed a rare pelvic tumour obstructing the left iliac vein.

Discussion. We review the current literature surrounding ilio-femoral thrombosis caused by extrinsic compression and venous stasis. To our knowledge, DVT caused by an extra-spinal ependymoma has not been reported in the past.

Keywords: Deep vein thrombosis; Iliac obstruction; Ependymoma.

Introduction

Extrinsic compression and resulting obstruction of the iliac veins is a recognised cause of DVT. Rarely, the underlying cause of obstruction proves to be a neoplastic lesion, with the DVT unmasking the tumour. We report a case of DVT caused by a rare pelvic tumour, adding to the differential diagnoses of secondary DVT.

Report

A previously healthy 39 year old woman presented with a painful grossly swollen left lower limb. She denied any trauma and was fully mobile prior to the onset of her symptoms. Examination revealed no cellulitis or neurovascular deficit. A venous duplex scan was carried out which showed left sided occlusive femoro-popliteal thrombosis but the iliac veins could not be visualised as bowel gas obscured a full examination. A presumptive diagnosis of idiopathic DVT was made and she was treated with low molecular weight heparin given subcutaneously and oral warfarin therapy. Over the following months, she experienced intermittent episodes of restrictive shortness of breath with persistent limb swelling. Results of cardio-respiratory and haematological investigations including a thrombophilia screen were normal. However, an ultrasound scan of her abdomen showed a 7 cm mass in her pelvis and a subsequent CT scan revealed a partially cystic non-enhancing mass measuring 6.8 cm situated in the pelvis just lateral to the aortic bifurcation, displacing the left iliac artery and compressing the left iliac vein (Fig. 1). Further clarification of the location of the lesion was gained by magnetic resonance imaging which showed a well defined left sided retroperitoneal lesion at the level of L5/S1, separate from the ovaries and adnexae (Fig. 2).

As the lesion was inaccessible to percutaneous biopsy, she underwent a laparotomy during which a large semi-cystic mass adherent to the left internal iliac vein and presacral plexus (with no spinal communication) was excised leaving no macroscopic remnants. Histology showed a very cellular tumour with uniform oval cells arranged in a pseudo-rosette formation around blood vessels. On immunohistochemistry, the tumour cells were positive for glial
fibrillary acidic protein (GFAP) and negative for cytokeratin. These features were consistent with a diagnosis of the cellular variant of extra-spinal ependymoma.

The patient made an uneventful recovery and was discharged back to the community. Adjuvant therapy was not considered due to the benign appearance of the tumour and lack of evidence supporting further treatment for such neoplasms. A follow up CT scan of her abdomen one year after surgery showed no evidence of recurrence. The patient remains well with no further paroxysmal episodes of shortness of breath.

Discussion

Studies have demonstrated that there is an anatomical predilection for acute ilio-femoral thrombi to form on the left side.\(^1\) This is generally due to compression by the right common iliac artery crossing the underlying left iliac vein, as first described by May and Thurner and also, in a significant proportion caused by the presence of venous spurs. Extrinsic compression of the iliac veins causing DVT has also been attributed to leiomyoma uteri, endometriosis, a distended urinary bladder, iliac artery aneurysm, heterotopic ossification within the hip flexor, debris from hip arthroplasty implant failure and from an extra-adrenal pheochromocytoma within the organ of Zuckerkandl.\(^2\) In addition, adventitial cystic disease of the external iliac vein has been reported to present with a DVT. However, there has been no previous report of an extraspinal ependymoma causing ilio-femoral thrombosis.

Ependymomas are glial tumours arising from ependymal cells that line the central canal or its remnants, accounting for nearly 60% of spinal intramedullary tumours in adults.\(^5\) Rarely, they can be found outside the confines of the CNS with most of such primary extraspinal ependymomas occurring within the sacro-coccygeal region.

In our patient, the presence of the tumour was heralded by the DVT possibly caused by extrinsic compression of the left iliac vein. Although the patient was presumptively diagnosed with idiopathic DVT, the diagnosis of the underlying neoplasm came to light after an ultrasound scan of her abdomen was performed to rule out any obstructive pelvic pathology. As reported by various studies, the incidence of finding a subsequent neoplasm after diagnosing an unexplained DVT is in the order of 2–25%. In order to detect an occult tumour in patients with idiopathic DVT, screening tests may be warranted, especially if there are no identifiable risk factors that may have contributed to the DVT.

As the biological behaviour of extraspinal ependymomas is unclear, their prognosis is unpredictable. In light of these facts, long term clinical and radiological follow up may be the only prudent option until more evidence with regards to their management emerges.

References


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