A case report of May–Thurner Syndrome associated with a large intra abdominal cyst

Fozia Saeed, Yazan S. Khaled, Venugopal K. Shankar

ABSTRACT

Introduction: May–Thurner Syndrome (MTS) usually presents with signs and symptoms of deep vein thrombosis or chronic venous insufficiency. We report the first case of MTS in a previously healthy adolescent female associated with a large intra-abdominal cyst arising from the right ovary.

Case Report: The patient presented with signs and symptoms suggestive of deep vein thrombosis. Imaging confirmed extensive deep vein thromboses extending into the common iliac vein. Further imaging successfully diagnosed MTS and also identified a cyst arising from the right ovary. Catheter-directed thrombolysis followed by removal of the cyst and the right ovary was performed. Following re-thromboses few days later, the patient underwent further management with surgical thrombectomy with left common iliac vein stenting and IVC filter placement.

Conclusion: The present case identifies an unusual presentation of a previously undiagnosed MTS in an adolescent female. In this case, anticoagulation alone was found to be ineffective and required the assistance of endovascular intervention.
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Keywords: Cyst, Endovascular, May–Thurner Syndrome, Thrombolysis, Vascular

INTRODUCTION

May–Thurner Syndrome (MTS) is an anatomical variant of the venous system in which the left common iliac vein (CIV) is compressed by the overlying right common iliac artery (CIA) leading to (via an unknown mechanism) vessel wall injury and predisposition to thrombosis [1–2].

The MTS can present as acute deep vein thrombosis (DVT) or chronically with varying degrees of venous insufficiency or chronic venous outflow obstruction [2]. It is a rare condition. The diagnosis is usually established following cross sectional, venography or ultrasonic investigation [2]. Treatment options include local thrombolysis or surgical thrombectomy for acute DVT and angioplasty and endovascular stenting for the underlying venous abnormality.

The purpose of this case report is to describe an unusual presentation of MTS in association with a large intra-abdominal ovarian cyst.

CASE REPORT

A 16-year-old Caucasian female, presented to the emergency department (ED) with a three day history of
left leg swelling and pain. She did not complain of any chest pain or shortness of breath and was systemically well. There was no previous history of DVT or of any recent travel, surgery or trauma. However, she had recently commenced use of a combined oral contraceptive pill (COCP) three months previously. There was no significant family history.

On examination, the whole left leg was markedly swollen with mild tenderness in the calf. It was warm and well perfused with all pulses palpable. There were no signs of chronic venous insufficiency. The right leg was normal.

Initial investigations demonstrated a raised white blood cell count at 11.25x10⁹/L and C-reactive protein (CRP) of 117 mg/L. Renal function and coagulation parameters were normal.

Ultrasound venous duplex of the lower limbs and the iliac segments revealed occlusive thrombus in the left common iliac vein (CIV), external iliac vein (EIV), femoral vein (FV), popliteal vein (PV) and short saphenous veins. A large (22x9x18 cm) intra-abdominal cyst was also found, compressing the inferior vena cava (IVC). Subsequent MRI scan of pelvis and high resolution MRV confirmed these findings - and that the cyst arose from the right ovary (Figure 1).

Blood screening tests for coagulopathy, thrombophilia and for gynecological malignancies were negative.

Following discussion at the departmental multidisciplinary team (MDT) meeting, the patient underwent three days of catheter directed thrombolysis through the popliteal vein with the agent, Alteplase (Actilyse, Boehringer Ingelheim Ltd, Berkshire, UK). Two fountain catheters (5 French and 4 French, Merit Medical Systems Inc. Utah, USA) were placed, one in the upper superficial femoral vein (SFV) and the other in the CIV. Catheter repositioning was performed after check venography on three occasions, with eventual successful clearance of acute thrombus from the iliac and femoropopliteal venous segments after 72 hours. The IVC remained compressed with sluggish venous return from the left leg. A decision was made not to attempt immediate insertion of an IVC filter and stent into the left CIV as the pelvic cyst obscured anatomical venous landmarks making it difficult to place them accurately.

The patient was aggressively anti-coagulated with unfractionated heparin (target activated partial thromboplastin (APTT) ratio range of 2.5–3.5). An open right salpingo-oophorectomy and ovarian cystectomy was performed 48 hours after cessation of thrombolysis. On opening the cyst, there were extensive nodules internally and pathological examination revealed the lesion to be a benign serous cystadenofibroma. The patient was restarted on IV heparin immediately postoperatively.

Repeat MR venography (on postoperative day 1) showed extensive re-thrombosis of the CIV, EIV, FV, and SFV despite the anticoagulation. The IVC was patent and no longer compressed (Figure 2). The recent laparotomy precluded the use of further thrombolysis. Left femoropopliteal and iliac venous surgical thrombectomy and left iliac venous stenting (Iliac vein stent, Sinus stent, OptiMed, Germany) was performed. The approach involved exposure of the left iliac bifurcation through an inguinal incision. An IVC filter was also inserted.

Outcome and follow up

The patient had an uneventful recovery postoperatively and was discharged home on POD-4 following stent insertion. The IVC filter was removed six weeks following insertion and the patient was then commenced onto Warfarin (INR target of 2-3). On follow-up after 4 months, the patient remains asymptomatic and fully independent.

Figure 1: MRI scan of pelvis with contrast: There is a large thin walled abdomino-pelvic cyst that has a craniocaudal extent of 22 cm and transverse measurements of 18.5x7 cm.

DISCUSSION

The MTS is related to an anatomical abnormality in which the right common iliac artery compresses the left CIV anterior to the sacral promontory and the fifth lumbar vertebra. It is hypothesized that arterial pulsation not only compresses the vein but also induces endothelial injury within its wall, resulting in the formation of venous spurs and thrombi [3]. MTS may present acutely or more commonly, may manifest as chronic venous insufficiency.
with varying degrees of trophic changes, varicose veins, edema and ulceration of the affected limb [3].

The exact incidence of this syndrome is unknown, however, compression (or at least flattening) of the left CIV in asymptomatic individuals from the general population is a common incidental finding on cross sectional imaging [3]. A recent literature review reported an incidence of 2–5% diagnosis of MTS in patients presenting with chronic venous insufficiency [2]. The syndrome is more common in females with a peak incidence in second to fourth decade of life [2].

In our case, the patient presented with symptoms and signs of DVT in the left lower limb but on investigating further, it was revealed that a large right ovarian cyst was also present, compressing surrounding structures including the IVC. This finding has not been reported in previous case reports and literature reviews discussing MTS. One can propose that the thromboses were as a result of the ovarian cyst alone with an incidental finding of MTS. Following removal of the ovarian cyst, the CT venogram performed confirmed the presence of MTS (Figure 3). The uncertainty remains and the presence of MTS as well as the cyst may have together precipitated the thrombotic events experienced by the patient.

The relevance of the ovarian cyst in this case also highlights the importance of individualized tailored management of patients. For example, the removal of the ovarian cyst resulted in re-thromboses of the lower limb. This may be attributed to the withholding of heparin in order to perform the surgical procedure of cyst removal.

The patient did not receive intra-operative heparin infusion as the risk at the time outweighed the benefit. It was thought that the risks of pulmonary embolism were low as the above knee and iliac vein thrombus had cleared entirely preoperatively.

The diagnosis of MTS is clinically challenging due to the rarity of this condition and the lack of guidelines or standard criteria for diagnosis. In addition, most patients remain asymptomatic with MTS and may remain undiagnosed [4]. We believe that our patient became symptomatic as a result of the mass effect of the enlarging pelvic cyst that was compressing the IVC in addition to the May–Thurner anatomical abnormality as well as beginning the use of the COCP, all of which have predisposed to extensive venous thromboses which manifested as leg pain and swelling.

To date, there is no international consensus on the best management of this condition. In the past, treatment for MTS involved anticoagulation alone. Using anticoagulation management alone may prevent the propagation of the thrombus but it does not eradicate the thrombus and neither does it relieve the extrinsic compression. This may therefore lead to further complications such as post-thrombotic syndrome and recurrence of thrombosis [2]. Case reports mostly suggest that t-PA infusions following initial clot lysis should continue with regular repeat imaging to guide further management [5–7]. Regular post-thrombolysis angiogram checks were performed in our patient and with re-positioning of catheters when required, successful lysis was performed prior to the patient undergoing a right salpingo-oophorectomy. Prior to endovascular management, open surgical procedures were performed commonly. However, due to the advancement in surgical technology, less invasive techniques such as endovascular repair became widely accepted [1]. Following thrombolysis and cyst removal, we deployed an endovascular stent into the left CIV. Case reports and small series studies have reported
encouraging results in the short-term with the use of stents [8]. One study suggested the use of a larger self-expanding stent that was placed at the area of narrowing with extension into the IVC, in order to minimize the risk of migration of the stent [9]. Patency rates of iliac vein stents are reasonably good ranging between 95% and 100% [9]. It is also recommended to insert an IVC filter prior to proceeding with any lower limb interventions as this reduces the likelihood of pulmonary events [6].

In our case, the initial management options available included (a) performing thrombolysis first followed by ovarian surgery, (b) performing ovarian surgery and surgical thrombectomy concurrently, or (c) inserting an IVC filter and performing ovarian surgery followed by thrombectomy or aggressive anticoagulation.

In consideration of the patient’s age combined with the long term effects associated with extensive iliofemoral DVT, it was decided to use thrombolysis rather than anticoagulation alone. Similarly, the decision to not insert an IVC filter prior to ovarian surgery was thought to be appropriate as the ovarian cyst was compressing the IVC and distorting the anatomy, which rendered the deployment of the device challenging and risky. The endovascular treatment allows for reversal of the obstructive component in MTS preventing further sequelae such as chronic venous insufficiency, which in our patient, was not yet apparent. Following endovascular treatment, it is advised to commence long term anticoagulation for a minimum of six months as per national guidelines [10].

CONCLUSION

In this case report, a previously undiagnosed May-Thurner Syndrome (MTS) was identified due to a large intra-abdominal cyst complicating the usual presentation with lower limb deep vein thrombosis. The case portrays the need for aggressive management of patients with MTS and the selection of treatment options should be customized according to the underlying pathology. The best outcome may be achieved through a combination of local thrombolysis and angioplasty with or without stenting. It is important to ensure optimal care and management for patients with MTS and this can be achieved through managing these patients in tertiary centers that are better equipped and prepared for dealing with complex vascular conditions like MTS.

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Author Contributions

Fozia Saeed – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Yazan S. Khaled – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Venugopal K. Shankar – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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