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Case report

Abdominal Mondor's disease presenting as acute abdominal pain. A case report and literature review

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Abstract. We describe here the case of 41 yrs old male patient, who was admitted to the emergency department complaining for abdominal pain lasting for two days. The patient self-reported a history of idiopathic deep vein thrombosis five yrs before the visit. A subcutaneous cordlike induration, tender and painful, was clearly palpable in the left lower abdominal quadrant. Routine blood tests did not reveal any substantial abnormality, except increased D-dimer concentration. Ultrasound evaluation of the abdominal wall revealed diffuse thrombosis of the left superficial inferior epigastric vein, involving several small tributaries branches, extended until 1.5 cm from the confluence with the common femoral vein, which was finally classified as an atypical case of Abdominal Mondor's disease. Complete thrombophilia screening was negative. The patient was discharged with warfarin therapy 48 hours from admission. At 30 days follow-up, the patient self-reported a nearly complete recovery. (www.actabiomedica.it)

Key words: Mondor's disease, thrombophlebitis, thromboembolism

Introduction

Superficial venous thrombosis (SVT) is a painful and frequent disorder (1), which is generally considered to have a good prognosis (2, 3). Some emerging evidence indicate that SVT, especially when localized in the lower limbs, may be however associated with venous thromboembolism (VTE). The percentage of patients with SVT who are also diagnosed with deep venous thrombosis (DVT) varies between 6-53%, whereas symptomatic pulmonary embolism (PE) may be present in up to 10% of cases (3). The heterogeneous epidemiological data about the frequency of this association are mainly attributable to the low number of studies published on this topic as well as to the limited number of patients included, so that the actual risk of both DVT and PE in SVT patients remains largely unknown (4, 5). In a recent large observational study, Décousus et al reported that VTE may accompany symptomatic SVT in nearly 25% of patients, most of whom with DVT and only 3.9% with symptomatic PE. It was also observed that 8.3% of patients with isolated SVT at admission developed at least 1 symptomatic thromboembolic event at 3 months (6). It seems thus plausible to raise the issue as to whether SVT may really be a benign condition (as it is widely thought), or rather a marker of a more clinically significant thromboembolic risk. Accordingly, the optimal therapeutic strategies for treatment of apparently isolated SVT (i.e., with no DVT or PE at presentation) are still unclear (7-9). The only accepted recommendations for management of SVT patients are those available in the American College of Chest Physicians (ACCP) Guidelines, suggesting the use of

prophylactic dose of fondaparinux or low molecular weight heparin (LMWH) for 45 days over no anticoagulation in patients with SVT of the lower limb of at least 5 cm in length (Grade 2B), and the administration of 2.5 mg daily fondaparinux over a prophylactic dose of LMWH in patients with SVT who are treated with anticoagulation (Grade 2C) (8). It is also noteworthy that the vast majority of scientific literature is focused on upper or lower-limbs SVT, whereas only anedoctal reports exist on other locations. These mainly entail the so-called Mondor's disease (MD), i.e. an SVT involving either the chest wall or penile veins. We describe here a case of acute abdominal pain in a 41 yrs old male, who was finally diagnosed with a superficial inferior epigastric vein thrombosis, clinically classifiable as MD.

Case report

M.F., a 41 yrs old male, presented to the emergency department (ED) of the Academic Hospital of Parma (Italy) complaining for abdominal pain, which was lasting for two days. The pain was referred to the left lower abdominal quadrant, and was not associated with either vomit or fever. The patient self-reported a history of "idiopathic" DVT (i.e., no trauma, malignancy, acute medical illnesses, surgery, immobilization, long distance air flight) five yrs before the visit, but was managed in a different hospital, so that no official clinical documentation was available. He also reported to have been treated with oral anticoagulant therapy (i.e., warfarin), which he personally withdrawn after a couple of months. The day before the visit the patient was evaluated for the same abdominal pain in another hospital, and was rapidly discharged with a diagnosis of "non specific abdominal pain". A subcutaneous cordlike induration, tender and painful, was clearly palpable in the left lower abdominal quadrant at presentation in our ED. No other relevant physical findings were present. The patient underwent routine blood tests, and the only suggestive abnormality was an increased D-dimer concentration (822 ng/mL; reference range <245 ng/mL). He was then subjected to ultrasound evaluation of the abdominal wall, with Doppler sonography (Philips iU22, with high frequency 12.5 MHz linear-array probe) of the superficial abdominal veins. The exam revealed a diffuse thrombosis of the left superficial inferior epigastric vein, involving several small tributaries branches, extended until 1.5 cm from the confluence with the common femoral vein (Fig. 1), which was finally classified as an atypical case of abdominal MD. No other thromboses of the lower limbs veins were detected. Ultrasonography of the abdomen did no show any additional pathological finding. Considering the history of "idiopathic" DVT, the patient was given enoxaparin 8000 I.U. b.i.d. and admitted to the Internal Medicine Unit. A complete screening for thrombophilia was then performed, which did not reveal and abnormality in the parameters tested, including homocysteine, antithrombin, coagulation proteins C and S, factor V Leiden and prothrombin 20210 gene polymorphisms, factor VIII, Lupus Anticoagulant (LAC), anti-cardiolipin and anti-beta-2-glycoprotein antibodies. After insistent request, the patient was finally discharged with warfarin therapy after 48 hours. At telephone follow-up at 30 days, the patient self-reported a nearly complete recovery, with minimal subcutaneous tenderness in the left lower abdominal quadrant, only in case of pressure (e.g., due to the belt). Once again, the patient self-interrupted warfarin treatment two weeks after hospital discharge.

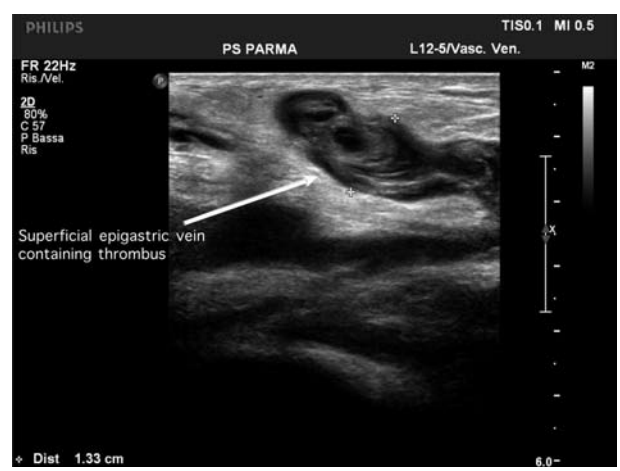

Figure 1. Ultrasound evaluation of the abdominal wall, showing diffuse thrombosis of the left superficial inferior epigastric vein, involving several small tributaries branches, extended until 1.5 cm from the confluence with the common femoral vein

Discussion

MD is a relatively rare clinical entity, characterized by a sclerosing thrombophlebitis of the subcutaneous veins of the anterior chest wall, mainly the lateral thoracic, the thoracoepigastric or the superior epigastric veins. It was first described by the French surgeon Henri Mondor in 1939, in a series of four patients presenting with SVT of thoracoepigastric vein and its tributaries (10). The sudden appearance is typically as a subcutaneous, red and tender cord, which subsequently evolves as a painless, tough, fibrous band accompanied by tension and skin retraction. Similar findings may rarely occur in the abdomen, arm, and axilla (11). This condition has been recently classified in three forms, each exhibiting a specific aetiology and management: type 1 MD, involving the chest wall; type 2 MD, involving other venous districts, mainly the dorsal vein of the penis; type 3 MD, occurring after breast surgery. The last form, although most often linked with breast cancer surgery, may however occur as a result of excision of axillary nodes after gel silicone breast implant rupture (12). Only single cases of type 2 MD with non-penile locations (i.e., brachial, femoral, and calf veins) has been reported (13). Overall, less than 500 MD cases have been published in the literature. The typical patient profile is that of a 30 to 60 years old woman (the female-to-male ratio varies between 3:1 and 20:1) (14). From histological perspective, four phases of the disease have been described, developing from thrombus formation, thrombus organization, recanalization and residual thick-walled fibrosis (15). The vast majority of MD cases are self limited in 3 weeks to 6 months, so that there is no universal consensus about the optimal therapeutic management. Several therapeutic approaches have been suggested, including no treatment, administration of non steroidal anti inflammatory drugs (NSAIDs), LMWH, antibiotics (especially in cases of penile MD after infection), and even surgery (13, 14, 16). When a predisposing or triggering condition can not be identified, the major benefits for the patients come from reassurance during follow-up (outpatient) visits.

As regards this specific case report, a single case of abdominal pain due to thrombosis of the inferior epigastric vein has been reported more than 50 years ago, which has been classified as atypical MD (17). A single case of MD presenting as tenderness and progressing swelling in the left hypochondriac region has also been recently reported (18), along with a very unusual case of MD mimicking a Spigelian hernia (19). As such, the case described in this report has some peculiarities. The first is indeed the clinical presentation as acute abdominal pain, which is very unusual in MD. An additional interesting aspect is that the patient was presumably affected by a thrombophilic condition, which could not be identified with a comprehensive thrombophilia testing or physical examination. This is probably the major strength of this case report, since comprehensive thrombophilia screening was not available in the previously described cases.

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