Mondor’s disease in this case report: is there any correlation with antiphospholipid syndrome?

Maladie de Mondor dans cette observation clinique : existe-t-il une corrélation avec le syndrome des antiphospholipides ?

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Résumé

La maladie de Mondor (MD) ou thrombophlébite des veines sous-cutanées de la région thoracique est une affection rare, d'étiologie inconnue dans la plupart des cas. Nous présentons ici un cas singulier associant maladie de Mondor et syndrome des antiphospholipides chez une femme de 63 ans chez qui on a noté une bonne évolution sous traitement antiagrégant plaquettaire, sans récidive clinique. L'association avec le syndrome des antiphospholipides est extrêmement rare et a été décrite dans quelques cas.

Mots-clés : maladie de Mondor, syndrome des antiphospholipides, sein

Summary

Mondor’s disease (MD) or thrombophlebitis of the subcutaneous veins of the chest region is a rare condition, with unknown etiology in most cases. Herein, we report a singular case associating Mondor’s disease and antiphospholipid syndrome in a 63-year-old woman, with a good outcome and no clinical recurrence after antiplatelet therapy. The association between Mondor’s disease and antiphospholipid syndrome is extremely rare and has been described in few cases.

Keywords: mondor’s disease, antiphospholipid syndrome, breast

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Introduction

Mondor’s disease (MD) occurs with palpable subcutaneous cord-like indurations beneath the skin. Usually, MD is a benign, self-limited disease that resolves spontaneously in four to eight weeks. MD was first described by Faage in 1869 as a form of scleroderma, but it was not until 1939 that a French surgeon, Henri Mondor characterized the condition that now bears his name (1-4).

The term antiphospholipid syndrome (APS) was coined in the 1980s to describe a condition of autoantibody-induced thrombophilia. This autoimmune prothrombotic condition is characterized by venous thromboembolism, arterial thrombosis, and pregnancy morbidity in the setting of laboratory evidence of elevated levels of antiphospholipid antibodies (aPLs) (5)

We report a case where a superficial thrombophlebitis of the internal quadrant of a left breast wall was associated with persistent B2GP1 antibodies. This raises the question: is there any correlation between Mondor’s disease and antiphospholipid syndrome?
Case report

We report a case of a 63-year-old woman with an obstetric history of preterm deliveries in unidentified context and a family history of concealed lupus erythematosus in 3 daughters. She presented with a chief complaint of pain in her left breast. She had no previous trauma or surgery and denied using any drugs. On physical examination, she was afebrile with stable vital signs and in no apparent distress. Her breast examination was significant for a superficial thrombophlebitis of the internal quadrant of the left breast. There was also an ecchymosis in this area (Figure 1). No axillary lymphadenopathies of pathological significance, or solid or cystic nodules were observed in the breast. A mammographic investigation carried out with a view to eliminating a mammary neoplasia, returned without abnormality. The diagnosis of Mondor’s disease was made, confirmed by an ultrasonographic examination of the breast in which an elongated hypoechoic image became evident, compatible with a thrombosed vein (Figure 2). The Thrombophilia Testing were normal. Elevated titres of IgG anti B2GP1, IgM antiB2GP1 antibodies were found and confirmed three months later. Neither IgM aCL antibodies, IgG aCl antibodies nor lupus anticoagulant were found. Antinuclear antibodies were detectable at 1/80 titer. Owing to the presence of aCL, the treatment with antiplatelet agent (Aspirin™ at the dose of 100 mg per day) was introduced. After one year, she had no recurrence of superficial thrombophlebitis and did not develop any other thromboembolic event.

Figure 1. Ecchymosis on the left breast

Figure 2. Sonogram reveals markedly dilated tubular structure with echogenic intraluminal thrombus (arrow)

Discussion

Mondor’s disease is a rare entity and has an estimated incidence of 0.84 % to 0.96 % of breast clinic patients (1,3); The reported typical patient's profile is a 30 to 60-year-old woman (2); and reported sex-ratio is usually three females for one male (2). Diagnosis of Mondor’s disease is clinical, based on history and physical examination (3). The differential diagnoses of cord-like lesions in the breast include infection, Erythema nodosum, inflammatory breast cancer and indurated carcinoma (1). However, ultrasound and Doppler ultrasonography may be necessary not only to confirm MD, but also to exclude other differential diagnoses such as presence of an underlying compressing mass. This ultrasonographic exploration shows direct or indirect signs of thrombus: presence of an internal echogenicity in the superficial vein in Gray scale, absence of venous flow signal on color Doppler and incompressible vein on compression by ultrasound probe. It should be pointed out that all available tools (colorflow, eflow, Bflow, pulsed Doppler…) are important and enhance the exploration (1-3).

The histopathological findings are limited to subcutaneous veins that show thrombosis and organization (4). There was damage to tunical intima of the vein and presence of an abundant blood clot in the lumen. Vessels were prominent with plump endothelial cells, an inflammatory infiltrate, and connective tissue proliferation in the wall. Necrosis, ulceration and thrombus can be found (4). Immunohistochemical staining can
distinguish between blood and lymphatic vessels. Blood vessels are positive for CD31 and CD34 monoclonal antibodies and for von Willebrand factor, and negative for LYVE1 and D240. The majority of cases have no clear etiology or precipitating event and the exact cause remains unclear. Various authors, however, have associated the disease with local trauma and breast surgery, including core biopsies, local inflammation, muscular strain, wearing of tight clothing, breast injections by drug abusers, Infectious agents have also been considered. The occurrence of some cases is “secondary” to another underlying disease, including malignancy. Thus, a small proportion of cases (2.4% to 12%) are associated with breast cancer. (1-3) Association with a hypercoagulative state, vasculitis and thrombophilia is very rare. The Antiphospholipid syndrome (APS) is the most common cause of acquired thrombophilia and accounts for 15 to 20% of all episodes of deep vein thrombosis, one third of new cases of cerebrovascular accident (CVA) occurring in patients aged less than 50 years, and 10 to 15% of recurrent fetal loss. The classification criteria for APS were developed in 1999 in Sapporo and subsequently revised in 2006 at an international congress held in Sydney. At present, these criteria include the requirement of at least one clinical criterion (thrombotic event or gestational morbidity) and at least one laboratory criterion (confirmed positive aPL at two or more separate time points with a 12-week minimum interval). Thus, superficial thrombophlebitis is not retained as clinical criteria for antiphospholipid syndrome. Some cases have noted an association between MD and antiphospholipid antibodies. The first description of a thrombotic event in hereditary protein C deficiency and anticardiolipin antibodies was published in 1999. Four years later, the French doctors of the Besançon Hospital published two original observations of Mondor’s disease from separated cases of two young patients. On the basis of the association with antiphospholipid antibodies secondary to autoimmune disease (autoimmune hepatitis, Systemic lupus erythematosus) in the majority of cases, specific treatment beyond symptom control is not required. Anti-inflammatory drugs may be effective to decrease the time to resolution. When treatment is not effective or there is recurrence of disease, surgical resection of the superficial vein should be necessary. However, the positivity of anticardiolipin antibodies in this case suggests that antiagregant therapy is necessary. Most lesions resolve over a period of 2 to 8 weeks.

Conclusion

Observations from the present study suggest that superficial thrombophlebitis must be related to complications of APS and should thus lead to the exploration of antiphospholipid antibodies, in particular in an autoimmune disease context, the longer follow-up of the positive cases should help to define the correct diagnosis and therapeutic attitude.

Declaration of Conflicting Interests

The Authors declare that there is no conflict of interest.

Author’s contribution

All the authors have contributed equally.

Références

6. Wester JPJ, Wester JPJ, Kuenen BC, Meuwissen OJA. Th, de Maat CEM. Mondor’s disease as...