

Mondor's Syndrome

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ABSTRACT

Mondor's Disease is a rare pathology, known as a superficial thrombophlebitis of the chest wall, benign and self-limiting, with a resolution that takes between 6 weeks to 6 months. It is more commonly observed in women than men. In most cases, it is idiopathic, and doesn't require specific handling, however, occasionally it can be a manifestation of other serious systemic pathologies. We report the case of a 34years old man, in emergency room with pain in a descendent path at the level of the right anterior axillary line and feeling of induration. His diagnosis was confirmed by echography, and associated systemic compromise was ruled out.

KEYWORDS: Mondor's syndrome; Mondor's disease; Superficial thrombophlebitis

INTRODUCTION

The first case of disease was reported in 1869 by Fagge, who considered it a form of scleroderma, and later by Hernri Mondor in 1939. It is an uncommon condition, probably underdiagnosed due to its benign and fairly asymptomatic course [1]. Historically it has been described as a syndrome characterized by superficial sclerosing thrombophlebitis of the veins in the anterior chest wall. The superior epigastric vein on the outer inferior quadrant of the breast is usually the most affected. There are some reports that indicate that this syndrome can affect the superficial veins of the penis [2], as well as other locations as upper limbs and inguinal region. It usually appears with unilateral compromise and rarely with bilateral.

CLINICAL CASE

34 years old man, with history of scleroderma, with pain on the right rib cage at the level of the anterior axillary line, with 20 days of evolution and increase 24 hours before his admission at emergency room. Pain extends towards the hypocondrium and flank of the

same side, is intense, is worsened by palpation of the area and doesn't have any elements that improve it. Patient denies any other associated symptoms. Physical examination shows a good general condition, vital sings: systemic arterial pressure 118/82 mmHg, breathing rate 18 rpm, heart rate 80 lpm and temperature in 36.2°C. In the anterior axillary line, on the right anterior hemithorax, a pain path extends towards the distal until the ipsilateral flank. There's no induration to touch, nor any anomalies on the skin. There's no axillar, cervical or inguinal adenomegalies.

Patient receives initial blood test and ultrasound exam of soft tissues on the thoracoabdominal wall that report: In the epifascial vein of the right hemithorax, in its T12 to T10 course, endoluminal echogenic images are shown, with no flow to any insonation angle with color Doppler, findings consistent with superficial acute thrombosis (Figure 1). Angio tomography of the chest was made to rule out other injuries or associated thoracic thrombosis, which was normal. Patient was discharged with indication of ambulatory control and symptomatic management.

Quick Response Code:



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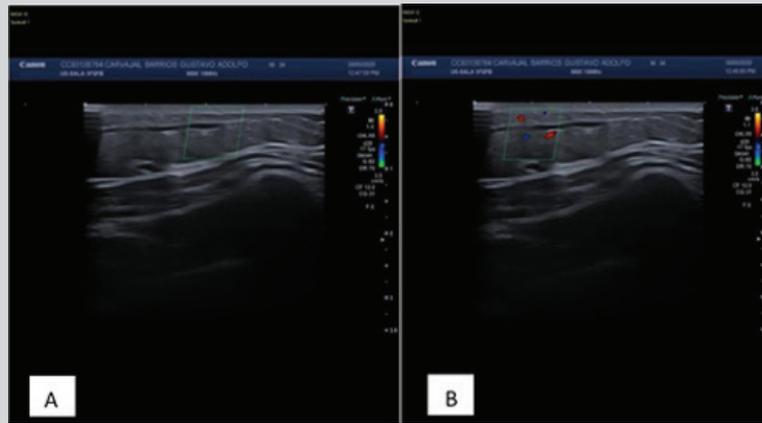


Figure 1: Dilated epifascial vein of the right hemithorax in thickness of subcutaneous cellular tissue at 2.2 mm from the skin, not compressible with endoluminal anechoic material approximately 50 mm (A), without flow with color doppler (B). Findings consistent with superficial acute thrombosis.

DISCUSSION

Epidemiology

Mondor's Syndrome is an uncommon disease. Few cases reported in the literature. Its prevalence in the primary care context is hard to establish. Commonly, appears in women between 30 to 60 years old, with average age of 35 years old [3], with relationship women to men of 3:1. No correlation has been found with regards of race or family history [4].

Physiopathology

Mondor's Syndrome physiopathology hasn't been properly described. A study made by Ichonose et al. [5] reported that the most predominant histopathological findings are thrombophlebitis, with a minority of cases resulting in lymphangitis. However, several precipitants that include the trauma, have been proposed. For the particular case of penis involvement, the causes could be sexually transmitted diseases, vigorous and prolonged sexual activity, as well as prolonged sexual abstinence, intravenous drug abuse and the use of Cunningham clamps [6,7]. Other association described are postsurgical stasis, venous stasis posterior to immobility, genetic pathologies but no case on that regards has been reported yet [2,8], and secondary to some systemic pathologies.

CLINICAL PRESENTATION

Usually, it manifests as a subcutaneous induration of sudden onset, often not painful, with edema, that looks like a rope on the vein and can be up to 5cm long, with linear depression [9]. Tends to be unilateral [10]. Typically it appears as a fibrous cord with a diameter between 3 to 5 mm, initially red [11], palpable on the subcutaneous tissue. Usually becomes more evident with the lifting of the breast or the abduction of the ipsilateral arm. It is not associated to lymphadenopathies or other injuries on the compromised area [2].

Some authors separate it on three different kinds according to the area affected: Mondor's disease on the thorax wall, Mondor's disease of the venous territories and Mondor's disease posterior to breast surgery. [4] Clinical manifestations are the same in all the areas described. Common symptoms include localized pain (that increases with the lift of the arm or during inhalation), erythema and edema of the adjacent skin [12]. Usually these follows the distribution of one of the 3 main superficial veins of the chest wall,

and in exceptional cases, of veins in the abdominal wall too. The dorsal vein of the penis is frequently compromised as well. Even though the most reported is the one on the chest wall, there are some cases involving upper limbs, inguinal [1] and cervical regions [13].

DIAGNOSIS

The clinical findings described are the main tool for the diagnosis of Mondor's syndrome. This is confirmed by ultrasound that shows the thickening of the vessel wall and obliteration of the lumen [11]. It is important to note that it is not always possible to identify the compromised vessel, specially, on the more advanced cases [1]. Biopsy is rarely indicated, except when there is compromise or affection of small arteries and lymphatics [11].

By histopathology, the thrombophlebitis is a severe vasculitis often accompanied with a thrombosis of medium and large size vessels on the dermis [5]. On earlier stages of the disease, a dense inflammatory cellular infiltrate, composed mainly of neutrophils, can be seen on a microscope. Edemas are often shown, as well as fibrin deposits on weird occasions. Another main characteristic of the microscopy is the presence of thrombus that occlude the lumen of the vessels [14].

On more advanced stages, lymphocytes, histiocytes and multinucleated giant cells may be found inside the wall of the vessel for a period of 2 to 6 weeks, with spontaneous resolution due to a recanalization process [9]. Doppler and nuclear magnetic resonance are useful for monitoring and evaluation of the recanalization. Biopsy is the most precise method when there is suspicion of malignancy and/or vasculitis [2].

Differential diagnosis include: lymphangiectasia, lymphangiomas, cellulitis, erythema nodosum and metastatic carcinoma of the skin [11]. It is important to differentiate Mondor's syndrome from other pathologies like neoplasms of vascular origin. For patients that do not respond to the initial treatment, a biopsy sample is indicated [15].

TREATMENT

Mondor's syndrome is self-limited, it resolves spontaneously in weeks or months without risk of embolism. Surgical resection is indicated if there is an associated bacterial infection without response to adequate antibiotic management [7]. Remember that

the etiology may be vasculitis, in this case the management is aimed at a possible autoimmune etiology.

COMPLICATIONS

There are no inherent complications of this disease known yet. The main risk is the oversight of serious pathologies with similar symptoms and clinical findings. There is a case reported by Kamangar et al. [16], in which the cutaneous metastases of a thyroid carcinoma were mistaken as Mondor's syndrome. Some other examples can be its association with metastatic cancer to the lung [17] or the distinction needed between this entity and the malignancies of vascular origin [7].

CONCLUSION

Mondor's disease is uncommon pathology, characterized by the presence of a hardened cord usually located on the thorax. In most cases it is related to thrombophlebitis, of self-limited evolution, that might not need a specific treatment but could be associated or be the initial manifestation of other serious pathologies like malignancies. Therefore, it is important adequate study. If there is any suspicion of another pathology, a biopsy should be performed.

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