

Observations and considerations on a case of Mondor's syndrome associated with gigantomastia



Ann. Ital. Chir., 2011; 82: 481-485

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AIM: *The study of the Mondor's syndrome as a possible complication of gigantomastia.*

MATERIAL AND METHOD: *A 49 year old woman, with gigantomastia, came to our observation for the presence of a large superficial vein of the left breast affected by thrombophlebitis, compatible with Mondor's syndrome.*

RESULTS: *A "Thorek" breast reduction was performed; the patient was discharged on the second postoperative day and the follow up was smooth and free from any complications.*

DISCUSSION: *In the case we observed, gigantomastia had a clear role in the pathogenesis of the Mondor's disease; in fact, the excessive weight of the breasts caused stretching of the mammary dorsal vein evolving in phlebitis. In literature, among the causes related to Mondor's disease the pendulous breast is described, probably with the same dynamic we have seen in this patient, but has never reported, so obviously, the correlation between the two events. It's considered as good practice in the pre-operative evaluation, to make differential diagnosis with any cancer, that, in a certain percentage, is associated with Mondor's syndrome, and once established the causes of the pathology, it is imperative to eliminate them to prevent any relapses.*

CONCLUSION: *Among the various causes recognized as predisposing to Mondor's syndrome, there has never been described gigantomastia so far. The observation and treatment of a patient in whom the gigantomastia certainly led to the onset of Mondor's disease, has led us to consider this as one of the possible complications of gigantomastia, providing an additional cue to the treatment of this condition even at an early stage.*

KEY WORDS: Gigantomastia, Mondor's syndrome

Introduction

Mondor's syndrome is a relatively uncommon affection characterized by superficial thrombophlebitis of the major thoracic veins, usually the thoracic-epigastric veins and, more rarely, the lateral thoracic and superior epi-

gastric veins^{1,2}. It was described for the first time in 1870 by Fagge³, then reported by Fessinger and Matthieu (1922), Williams (1931), Daniels (1932), Moscheowitz (1933) and Robinson (1935), and owes its present name to 4 clinical cases reported by H. Mondor in 1939⁴. The incidence of this pathology varies between 0.5% and 0.8%^{5,6} but presumably this does not reflect the true incidence of the disease as these case studies take into account only the symptomatic patients. It affects mainly women with a ratio of 3 to 1 compared to men⁷ and most of these women are in adult-senile age, especially those in overweight and with pendulous breast⁶. Although the main tissues affected by the disease are in the mammary region, the lateral chest or the epigastric area, extrathoracic clinical variants have been reported, with involvement of atypical sites, such as the

Pervenuto in Redazione Marzo 2011. Accettato per la pubblicazione Giugno 2011.

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posterior cervical region, the axilla, the upper extremities, the antecubital cavity, the abdomen, the groin and the penis ².

Clinical manifestations vary from almost asymptomatic conditions, to those that more frequently include the appearance of a subcutaneous cordon-like swelling, linear or more rarely serpiginous, painful on palpation, with or without a feeling of tightness, itching and / or burning, and sometimes accompanied by redness, swelling and / or retraction of the overlying skin ^{1,8}. Fever and joint pain can occasionally overlap to complete the clinical set of symptoms. Diagnosis is mainly based on symptomatology and objectivity. In fact, laboratory tests are usually in normal range, including the inflammatory markers ⁹, but as described in the literature, an association of Mondor's syndrome with breast cancer, is ranging from 4% ¹⁰⁻¹³ to 12.7% ¹⁴ (this difference may in part be due to inadequate diagnostic investigations performed on patients with Mondor's syndrome), and in front of this hypothesis, a careful diagnosis, clinical and instrumental, is required, in order to prevent that a tumor, often not palpable, may go unrecognized. So, it's best to look for the typical instrumental aspects of the breast Mondor's syndrome, such as the finding of a mammographic tubular density and a ultrasound / doppler of the superficial veins with or without intraluminal thrombus in case of absent flow in U.S. Doppler examination ^{2,15}.

Generally the clinical evolution of the disease is towards the resolution in a period of 2-4 months, and commonly the prognosis is favorable, with complete "*restitutio ad integrum*" ⁵. In most cases, a symptomatic therapy, based on the use of NSAIDs or other analgesics is sufficient. Usually, surgical treatment is reserved only for cases associated with a cancer but it has been suggested in rare cases of severe local pain with poor response to therapy or in cases in which skin retraction generated distress and / or functional limitations ². The observation of a patient with gigantomastia, in our department, after the onset of the symptoms typical of Mondor's syndrome, led us to some considerations on the subject of this work. Among the many associations described and the different pathogenetic hypotheses suggested from time to time, there is no description of the Mondor's syndrome in patients with gigantomastia. This last condition, characterized by an increase in breast volume requiring a gland weight reduction of about of 1000-1500 g. ¹⁶ and due to idiopathic, hormonal or iatrogenic causes ¹⁷, has so far been associated only with diseases such as SLE, lymphomas¹⁸, or hypercalcemia ¹⁹, without the support of depth scientific studies yet.

Material and method - Results

The patient that came to our observation was a 49 year old woman (A.R.), slightly in overweight. For about three

years, the patient reported a slow and gradual increase in volume of both breasts without any hormonal changes, except for a mild hypothyroidism treated with organotherapy. About 20 days before coming to our observation, the patient had reported the presence of a thrombophlebitis of a large superficial vein, passing longitudinally between the upper quadrants of the left breast. For unclear reasons, the patient refused to be checked and after about 15 days, to the previous symptoms she noticed the formation of a large hematoma at the first bifurcation of that vein that then evolved into a large abscess. At this point, the patient was referred to our outpatient observation, where we immediately drained the abscess, extracting about 500 cc of smelly purulent-like liquid. The cultural examination showed the prevalence of "*Proteus mirabilis*". In addition to broad-spectrum antibiotic coverage required for the first few days, the treatment consisted mainly in a large surgical drainage of the abscess cavity and its first daily irrigation with H₂O₂ 50% diluted in saline and then only with saline solution, taking care to leave in the site a "Penrose" type laminar drainage, which was replaced each time. The healing of the abscessual cavity was obtained after about one month. A large clear plate with poorly defined margins and hard-fibrous consistency, of uncertain radiographic and echographic meaning, lasted nearby. The MRI contrast imaging, then carried out, allowed us to exclude the coexistence of neoplasm and confirmed it to be only a scar. We have therefore performed the "Thorek" ^{16,18,20} breast reduction in this patient, both to prevent that the weight of the breasts and the stretching of its veins could determine the recurrence of the problem and for the obvious psychological and social discomfort that the gigantomastia was determining. The patient was discharged on the second postoperative day and the follow up was smooth and free from any complications.

Discussion

Although the etiology of Mondor's syndrome still remains obscure, most of the scientific literature tends to believe that the true cause of the thrombophlebitis should be found in traumatic factors, or as the onset of a particular tension of muscles in the upper limb due to repeated stresses or infections ^{8,21}. The use of constricting clothing, breast augmentation or breast reconstructive surgery and the lesions derived performing breast diagnostic instrumental imaging can also be considered as causes of trauma ^{8,14}. Has also been considered as a possible cause. the action of stinging jellyfish²². A viral or infective origin, has been suggested in a variable percentage from 4.6% to 12% of cases ⁸. In the clinical case we observed the gigantomastia had a clear role in the pathogenesis of Mondor's disease and its complications. In fact, the excessive weight of the breast caused

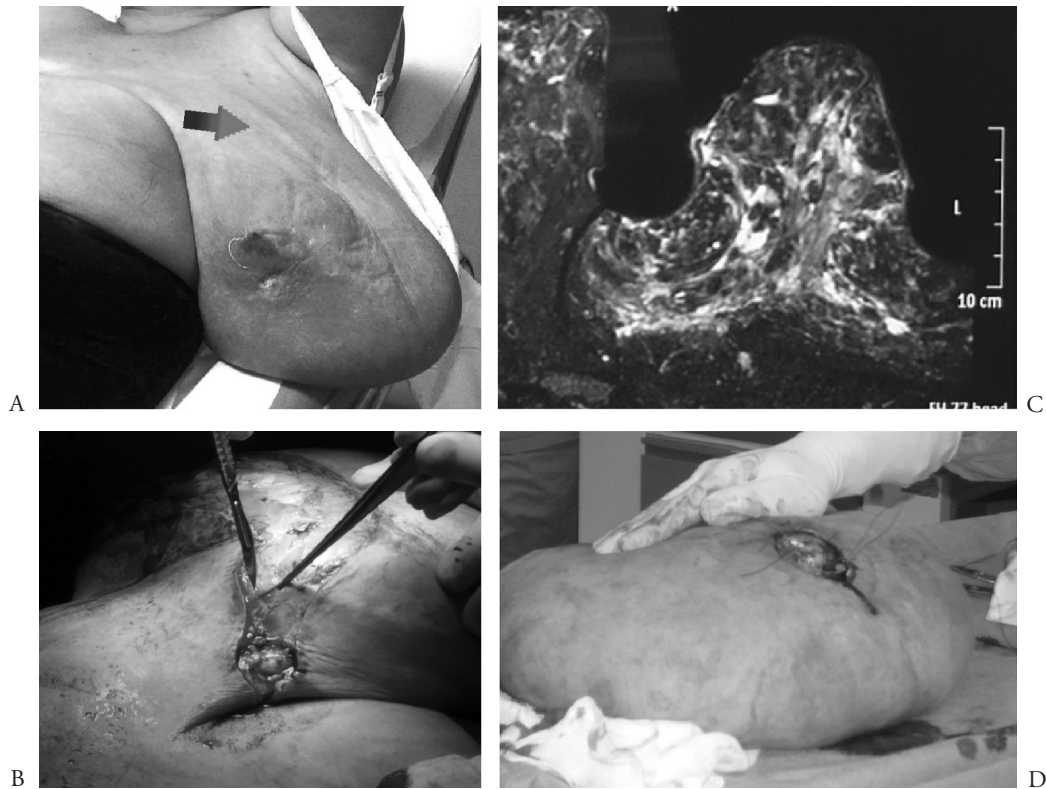


Fig. 1: A) Gigantomastia in which are evident the dorsal vein stretched and thrombosed (→) and the abscess of the IMQ; B) MRI of the breast where it is clear the outcome of the abscess cavity; C) Demolitive phase of the intervention, in which is underlined the mammary dorsal vein thrombosed; D) Size of the portion of parenchyma reduced by the left breast.

a stretching of the mammary dorsal vein evolving in phlebitis. The carelessness of the patient, led to a partial rupture of the vein, resulting in the formation of a hematoma, which turned to a large abscess cavity.

As a matter of fact, in literature, among the causes related to Mondor's disease the pendulous breast⁶ is described, probably with the same dynamic that we have seen in this patient, but the correlation between the two events was never reported so clearly. It's considered as good practice in the pre-operative evaluation, make differential diagnosis with any cancer, that, as has been said, in a certain percentage, is associated with Mondor's syndrome¹⁰⁻¹⁴. The diagnosis can be supported by traditional imaging techniques such as ultrasound and mammography and when, as in the case we reported, they leave any doubts of interpretation of certain specific aspects, the use of MRI and an echoguided biopsy, are recommended^{17,23,24}. Having established, without any doubt, the causes that determined the Mondor's syndrome, as occurred in our case, it is imperative to eliminate them, and thus prevent the possibility of recurrence of the disease.

Conclusions

Among the various known causes promoting the development of Mondor's syndrome, there has never been described gigantomastia so far.

The observation and treatment of a patient in whom the gigantomastia certainly led to the onset of Mondor's disease, led us to consider this one of the possible complications of gigantomastia, providing an additional cue to the treatment of this condition even at an early stage. Mondor's syndrome is a relatively uncommon affection characterized by superficial thrombophlebitis of the major thoracic veins, usually the thoracic-epigastric and, more rarely, lateral thoracic and superior epigastric veins. The case that came to our observation was a 49 year old woman, affected by gigantomastia, showing thrombophlebitis of a large superficial vein of the left breast, compatible with Mondor's syndrome. After an accurate diagnostic assessment, a "Thorek" breast reduction, was performed, both to prevent that the weight of the breasts and the stretching of its veins could determine the recurrence of the problem, as for the obvious psychological

and social discomfort that the gigantomastia was determining. The patient was discharged on the second post-operative day and the follow up was smooth and free from any complications.

In the case we observed, gigantomastia had a clear role in the pathogenesis of the Mondor's disease; in fact, the excessive weight of the breast caused stretching of the mammary dorsal vein evolving in phlebitis. In literature, among the causes related to Mondor's disease the pendulous breast is described, probably with the same dynamic we have seen in this patient, but such a strong correlation between the two events has never been reported so far. It's considered as good practice in the pre-operative evaluation, to make differential diagnosis with any cancer, that, in a certain percentage, is associated with Mondor's syndrome, and once established the causes of the pathology, it is imperative to eliminate them to prevent relapses. The case we reported, led us to consider the Mondor's syndrome as one of the possible complications of gigantomastia, providing an additional cue to the treatment of this condition even at an early stage.

Riassunto

La sindrome di Mondor è una affezione relativamente rara caratterizzata da tromboflebite superficiale delle vene principali del torace, di solito le toraco-epigastriche e, più raramente, le toraciche laterali e le epigastriche superiori. Il caso giunto alla nostra osservazione è relativo ad una donna di 49 anni, affetta da gigantomastia, che presentava la tromboflebite di una grande vena superficiale della mammella sinistra, compatibile con la sindrome di Mondor. La paziente, una volta formulata la diagnosi, è stata sottoposta ad una mastoplastica riduttiva secondo "Thorek", sia per evitare che il peso della mammella e lo stiramento dei suoi vasi potessero determinare il ripetersi del problema, sia per l'evidente disagio psicologico e sociale che la gigantomastia aveva determinato. La paziente è stata dimessa in seconda giornata postoperatoria e il decorso è stato privo di complicazioni.

Nel caso da noi osservato, la gigantomastia ha avuto un ruolo chiaro nella patogenesi della malattia di Mondor; infatti, l'eccessivo peso delle mammelle ha causato lo stiramento della vena mammaria dorsale con conseguente evoluzione in flebite. In letteratura, tra le cause correlate alla malattia di Mondor è descritta la mammella pendula, probabilmente con la stessa dinamica che abbiamo visto in questa paziente, ma non è mai stata segnalata, prima d'ora, una così stretta correlazione tra i due eventi. È buona norma, nel periodo pre-operatorio, porre diagnosi differenziale con eventuali patologie tumorali che, come si è detto, in una certa percentuale, si associano alla sindrome di Mondor, e una volta stabilite le cause della patologia, è indispensabile intervenire per eliminarle, ed evitare così spiacevoli recidive. Il caso da noi osservato ci ha

portato a considerare la sindrome di Mondor come una delle possibili complicanze della gigantomastia, fornendo un ulteriore spunto per il trattamento di detta condizione anche in fase preventiva.

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