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Classification of the Clinical Presentation, Severity, and Response to Treatment in Idiopathic Granulomatousis Mastitis

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Idiopathic granulomatous mastitis (IGM), also called idiopathic lobular mastitis, is a rare, benign inflammatory condition of the breast with currently unknown etiology. The cornerstone of the design of a clinical trial relies on the type and the severity of the clinical findings and the response to the treatment strategy applied. Without a standard classification, we may not expect to accomplish uniform investigations in different centres. According to the considerable experience of the authors and considering our previous publications, we suggest a classification in four grades according to the clinical manifestation (inflammatory, cutaneous, soft tissue, and systemic findings). Severity can predict the treatment response, so we suggest the therapeutic modalities be chosen based on this factor.

Idiopathic granulomatous mastitis (IGM), also called idiopathic lobular mastitis, is a rare, benign inflammatory condition of the breast with currently unknown etiology.[1] The course of the disease is sometimes chronic and devastating for the patients.[2] It mostly affects women of childbearing age with a history of pregnancy or lactation within the last five years of the onset of the disease.[3-5] This is a rare condition in developed countries; however, it is more prevalent in developing counties. Recently there has been an increase in the number of studies in this regard. The reports mostly come from middle-east countries, including Iran and Turkey.[4]

Although an increase in the number of studies is evident, and it is a well-known clinical condition since 1972[6], there is not yet a general consensus regarding the management of this disease.[7, 8] In the literature, there is a disparity in clinical findings, diagnosis, management, and follow-up of the disease in different studies.

The cornerstone of the design of a clinical trial relies on the type and the severity of the clinical findings and the response to the treatment strategy applied. Without a standard classification, we may not expect to accomplish uniform investigations in different centres.

The clinical presentation of the disease contains a wide range of signs including inflammatory symptoms resembling infectious mastitis or inflammatory carcinoma depending on the severity and extent of the inflammation. On the other hand, some patients would refer to medical centres with breast mass and tissue thickening. This group of symptoms are mostly related to the collection with or without inflammation that may mimic breast cancer. Finally, in some situations, the breast complaints are a part of a systemic disease. The clinical manifestation of the disease is summarized in Table 1.

Three hundred seventy-four patients were analysed in our previous work.[9] The severity of the disease in that study was categorized into three levels. Considering our previous publications[9-14], we made some modifications to the classification system we used. We classify the severity of the disease in Table 2, based on the clinical manifestations and radiologic findings. We have conducted another investigation on the radio-pathologic findings of the disease in 224 patients and the severity of the disease is classified based on the description presented in this paper (Table 2). The clinical findings of the disease, along with radiologic and pathologic assessment of this large cohort of patients, will be presented soon.

Also, as an outcome measure, the researchers need a consistent classification of the response rate. This classification is crucial to evaluating the efficiency of each treatment modality, so we categorize the response to treatment based on the rate of improvement in the signs and symptoms as well as

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Table 1. Classification of the signs and symptoms

Inflammatory	Pain, Redness, Erythema, peu d'orange, skin thickening, axillary lymphadenopathy
Soft tissue*	Deep collections, tissue thickening, mass, skin dimpling, nipple retraction
Cutaneous Destructive	Thin red skin, superficial collection, Ulcer, fistula
Extramammary	Systemic lymphadenopathy, Arthralgia, Arthritis, Erythema nodosum, etc

* This group needs to be studied by clinic-radiologic investigations. They can be considered mild when the collection is single with minimal inflammatory reaction around it, moderate when the collections are multiple in a single quadrant and severe when the collections can be seen in more than one quadrant.

Table 2. Grading of the severity of the disease

Grade	Inflammatory	Soft tissue	Cutaneous destructive	Extramammary	Suggested Treatment
I	Mild	Mild	No	No	NSAIDs*
II	Mild to Moderate	Moderate to Severe	No	No	Percutaneous drainage$^{\Theta}$ ANB$^{\varrho}$ NSAIDs*
III	Sever	Moderate to Severe	Yes	No	Open drainage ANB$^{\varrho}$ NSAIDs * +_CS$^{\varepsilon}$ +_ISM$^{\eta}$
IV	Any	Any	Any	Yes	Based on the protocols for systemic disease

* Non-Steroidal Anti Inflammatory Drugs for three months
$^{\Theta}$ Should be done with large born needles, preferably with the guide of ultrasonography
$^{\varrho}$ Widespectrum oral antibiotics with coverage on gram-positive microorganisms for 10-14 days
$^{\varepsilon}$ Corticosteroids can be used based on clinical impression with 50 mg prednisolone in a tapering dose, discontinue in 10 weeks. Can be continued in low dose (5-10mg/day) for a maximum of 3 months
$^{\eta}$ Immunosuppressive medications can be used if only the disease in persistent in Grade III for 6 months

Table 3. Classification of "Response to Treatment" in breast-limited IGM

	Initial Control of symptoms	No Recurrences*	Severity of the recurrences*
Excellent Control	Complete$^{\Theta}$	0	-
Good Control	Complete$^{\Theta}$	1-2	Grade I-II
Fair Control	Partial$^{\varrho}$	3-4	Grade I-II
Poor Control	Fair $^{\varepsilon}$	Persistent disease	Grade III

* During a 12-months follow up
$^{\Theta}$ Resolution of >90 percent of the signs and symptoms
$^{\varrho}$ Marked reduction in the signs and symptoms
$^{\varepsilon}$ Not a big resolution in signs and symptoms

the frequency of the recurrences (Table 3). It is clear that the clinical classifications will be authenticated if they are concordant with the imaging (ultra-sonographic) findings.

Conflict of Interest

None.

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