

Two Ultrasonographic Patterns in Maculopapular Cutaneous Mastocytosis: A Preliminary Report

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ABSTRACT Maculopapular cutaneous mastocytosis (MPCM) is a variant of cutaneous mastocytosis resulting from abnormal accumulation of mast cells in dermal tissues. Ultrasonography is a contemporary, safe, non-invasive, and real-time diagnostic method. High-frequency ultrasonography (HFUS) utilizes probes emitting frequencies of 20 MHz and higher, visualizing and measuring areas of healthy and lesional skin in various dermatologic conditions. We examined 4 patients with MPCM using a *taberna pro medicum*TM (Germany) device. We present 2 characteristic ultrasonographic patterns of MPCM lesions. Three subjects presented with an abnormal, widened, and hypoechogenic area representing the dermis. One subject demonstrated an anechogenic fusiform structure below the epidermis that represented a localized pattern of infiltrate. This patient responded better to psoralen and ultraviolet A radiation (PUVA) therapy than the other subjects. Our research implies that HFUS may be used as an additional method in evaluating skin lesions in MPCM.

KEY WORDS: high frequency ultrasonography, cutaneous mastocytosis, diagnosis

INTRODUCTION

Mastocytosis refers to a heterogeneous disease that is divided into systemic and cutaneous forms (1). Cutaneous mastocytoses comprise maculopapular cutaneous mastocytosis (MPCM; formerly known as urticaria pigmentosa), diffuse cutaneous mastocytosis (DCM), and solitary mastocytoma of the skin. MPCM occurs mainly in childhood and manifests with tan or brown macules and papules most commonly localized on the trunk. Additionally, Darier's sign is mentioned as a pathognomonic feature of this clinical entity, as the formation of an itchy urticarial wheal after rubbing skin lesions indicates the presence of mast cell infiltration in the dermis. The latter may be visualized in histologic evaluation of skin biopsy, especially utilizing immunohistochemical staining for tryptase and CD117 (c-Kit) particle.

Ultrasonography is a contemporary diagnostic method used in many fields of medicine, including

dermatology. Skin thickness measuring was first introduced in the late 1970s (2). Different frequency spectrums have been evaluated over the years. Frequencies of 7.5-15 MHz are frequently used for the evaluation of lymph nodes and subcutaneous lesions. Probes emitting and receiving waves of 20 MHz and higher were introduced into clinical practice in the 1990s. Higher frequencies provide better visualization of upper skin layers, better resolution, and are capable of showing smaller structures (3). The disadvantage is decreased depth of ultrasonographic penetration. High frequency ultrasonography (HFUS) has proven useful in visualizing several parameters of healthy and lesional areas of the skin without performing a biopsy. These parameters include the intensity of the infiltrate in chronic inflammatory disorders or the depth of skin tumors. HFUS of the skin is a quick, non-invasive, real-time, and safe diagnostic

method. Despite these advantages, the utilization of HFUS in dermatology remains limited, partly due to special skills required from a physician, subjective assessment of several phenomena, and devices which are relatively costly. Here we present our experience with HFUS in cutaneous mastocytosis, which has not been previously reported to the best of our knowledge.

PATIENTS AND METHODS

We examined 4 patients admitted to our Department of Dermatology, each presenting with typical MPCM lesions (2 women, 2 men, aged 23, 24, 28, and 32 years, respectively) (Figure 1). Diagnoses were uniformly confirmed by skin biopsy. The sonographic phenomena were evaluated utilizing a 20 MHz *taberna pro medicum*TM (Germany) device. The data were collected and saved using DUBmicro[®] tpm and DUB 6100 v1.0 software. The parameters of axial and lateral resolution were approximately 80 μ m and 200 μ m, respectively. The length and the depth of investigation were 12.8 mm and 8.0 mm, respectively. Measurements and echogenicity of the structures were assessed in both A-mode and B-mode. The densitometry value was defined as the mean height of the reflection amplitude in a standardized scale of 255 amplitude levels. Bright colors in B-mode depicted hyperechogenic structures, while dark colors corresponded with hypoechogenic structures. Lesional skin areas were assessed, as well as areas of clinically unaltered skin, to provide a reference point for each individual. The study was approved by the local ethics committee.



Figure 1. Maculopapular cutaneous mastocytosis with typical reddish papular lesions on the trunk.

RESULTS

Healthy skin areas presented with a hyperechogenic entrance echo, with a normoechogenic area below (representing the dermis) and a hypoechogenic or anechogenic zone associated with subcutaneous tissues (Figure 2). The border between the dermis and subcutaneous tissues was linear. HFUS also revealed linear hyperechogenic structures which represented muscle fascia.

In lesional skin areas, our research demonstrated 2 distinctive ultrasonographic patterns. Three patients out of four demonstrated a normoechogenic entrance echo and a widened, hypoechogenic area representing the dermis (3400-3800 μ m vs. 2800 μ m in clinically unchanged skin, 2700-2900 μ m vs. 2200 μ m and 2800-3000 μ m vs. 2400 μ m, respectively). Decreased echogenicity of dermis was a manifestation of a diffuse cell infiltrate (Figure 3). The fourth patient presented with a normoechogenic entrance echo and an anechogenic, fusiform structure below the epidermis, depicting a characteristic localized pattern of the infiltrate (Figure 4). Notably, this patient responded significantly better to PUVA-therapy (12 irradiations) than the other subjects (data not shown).

DISCUSSION

The diagnosis of skin diseases is based chiefly on clinical examination. Physician determines the type of primary lesions and their measures. The third dimension of the lesions is assessed indirectly, for example by palpation. Occasionally biopsy is required, determining the depth and histologic features of the lesions. This procedure is necessary in case of skin tu-

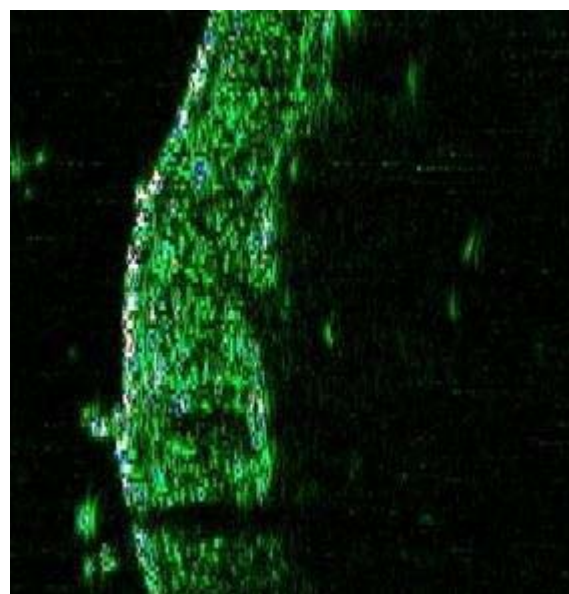


Figure 2. Ultrasonographic image of healthy skin.

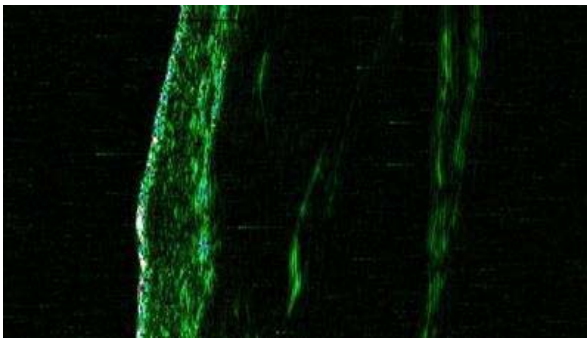


Figure 3. HFUS examination of MPCM lesions reveals a normoechogenic entrance echo. The dermis is markedly widened (3400-3800 μm vs. 2800 μm in clinically unchanged skin) and hypoechogenic.

mors. Ultrasonographic imaging of the skin is an additional modality that aids diagnosis and monitoring of skin diseases. Numerous investigators remarked on its effectiveness in assessing various dermatologic conditions. Hoffmann *et al.* (4) reported that an echopoor area underneath the entry echo was present in active psoriatic plaques, depicting acanthosis and inflammatory infiltrate. Successful therapy resulted in reduction of the echopoor area and an increase in density. Corresponding observations were performed in patients suffering from atopic dermatitis (5). Ultrasonographic evaluation of patients with scleroderma also seemed useful as it was better than clinical examination alone in determining the stage of the disease (6). Recently Maj *et al.* (7) reported that HFUS provides valuable information concerning size and depth of malignant melanoma and may be regarded as a complementary method in preoperative evaluation of the tumor. Other reports concerning malignant skin tumors were also published (3,8).

To our knowledge, our research is the first published report regarding the usefulness of HFUS in evaluating MPCM. The intensity of the infiltrate in dermis varied between patients, and we assume that HFUS might provide the physician with an objective method of assessing the severity of the disease. We demonstrated two different patterns of infiltrate in MPCM lesions, although the explanation for these observations is unknown. The association between the characteristic ultrasonographic pattern of the infiltrate and positive response to PUVA treatment may be coincidental. However, another study of ours performed in patients with cutaneous T-cell lymphomas suggested that infiltrate manifesting as a nodular hypoechogenic area under the epidermis is also associated with positive response to treatment, including PUVA-therapy (results unpublished).

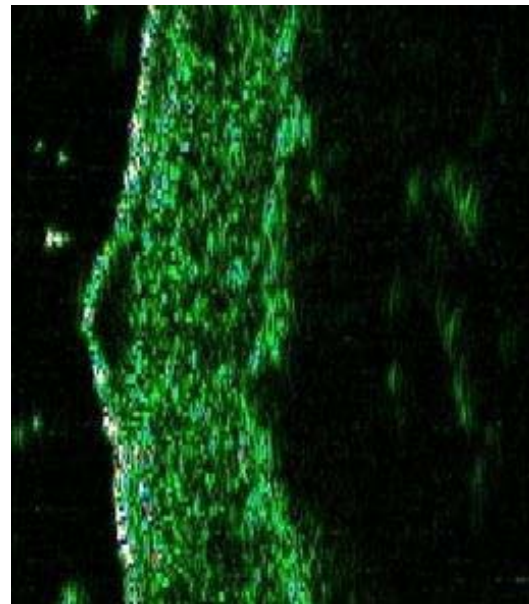


Figure 4. An example of a localized pattern of infiltrate.

CONCLUSION

In conclusion, HFUS provides the physician with a useful method of assessing MPCM lesions, possibly complementing clinical staging and histological grading. Additionally, the pattern of the infiltrate may indicate the therapeutic outcome in this dermatosis. Further studies regarding this subject seem advisable.

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