CASE REPORT

Case Report: Solitary mastocytoma treated successfully with topical tacrolimus [version 1; referees: 2 approved, 1 approved with reservations]

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Abstract
Solitary mastocytoma, a rare dermatological entity accounts for 10-15% of cutaneous mastocytosis. We report a rare case of solitary mastocytoma presenting at birth, treated successfully with topical tacrolimus. Along with reassurance and strict avoidance of triggering factors, no recurrence was reported within the one year follow-up period.
Introduction
Solitary mastocytoma, a rare dermatological entity, represents the second most common type of cutaneous mastocytoma. Solitary mastocytomas constitute 10–20% of all childhood cutaneous mastocytosis. They usually present within 2 years of age, mostly within first 3 months.

We report a case of solitary mastocytoma presenting a birth that was treated successfully with topical tacrolimus with no recurrences noted during a one year follow-up period.

Case report
An eighteen month old girl presented with a solitary, itchy dark coloured, minimally elevated lesion over her left elbow that had been evident since birth. The lesion used to itch and swell on scratching, bathing and toweling of the area. The child was otherwise healthy and no other systemic manifestations were noted. Clinical examination revealed a solitary, 3.5 × 6.5 cm, non-tender, minimally elevated plaque with central shiny skin and peripheral marginal hyperpigmentation over left elbow. On scratching the lesion with the blunt end of a pin, the central shiny skin became edematous and itchy (positive Darier’s sign) (Figure 1). Hematological and biochemical investigations were within normal limits. A 5 mm biopsy of the skin tissue obtained from the center of the lesion revealed a dense monomorphic inflammatory infiltrate consisting of round to oval cells with clear cytoplasm and centrally located nuclei in the upper and mid dermis (Figure 2a, 2b). Special staining with toluidine blue revealed metachromatic staining of the monomorphic mast cells, confirming the diagnosis of mastocytoma (Figure 3).

The child was treated with topical tacrolimus 0.03% ointment which was applied on the lesion site twice daily. The child was also prescribed an oral antihistamine (levocetirizine syrup, 1.25 mg once a day). By the end of third month, complete subsidence of the lesion was noticed with residual hyperpigmentation, negative Darier’s sign, and no signs of atrophy. This treatment was continued for another four months which led to resolution of the lesion with residual hyperpigmentation, negative Darier’s sign, and no signs of atrophy. Treatment was continued with only a once a day treatment.

Figure 1. A solitary, 3.5 × 6.5 cm, non-tender, minimally elevated plaque with edematous central shiny skin made more apparent on scratching the lesion with the blunt end of a pin (positive Darier’s sign) with peripheral marginal hyperpigmentation over the left elbow.

Figure 2. a, Dense monomorphic inflammatory infiltrate in upper and mid dermis; b, Dense monomorphic inflammatory infiltrate consisting of round to oval cells with clear cytoplasm noted at 40x magnification.

Figure 3. Metachromatic staining of the monomorphic mast cells with Toluidine blue staining.
application of topical tacrolimus for a month after clinical resolu-

tion to prevent further recurrence (Figure 4). Reassurance and strict

avoidance of triggering factors such as pressure, friction (rubbing or
toweling of the lesion), extreme temperature changes, intake of mast
cell degranulating agents like aspirin, NSAIDS, morphine, codeine
(especially in the form of cough preparations) has led to no recur-
rence of the child’s symptoms during a 1 year follow-up period.

Figure 4. Complete subsidence of the lesion with residual
marginal pigmentation noted at the end of three months of
therapy. The central atrophic scar due to biopsy can be seen in the
centre of the lesion.

Discussion
Solitary mastocytoma, the second most common type of cutane-
ous mastocytosis, accounts for 10–15% of cutaneous mastocytosis1.
Nearly half of solitary mastocytomas present within the first 3 months
of life and the remaining half during the first year2. Solitary masto-
cytoma presenting in adults has also been noted3. The most com-
mon locations of mastocytomas are on the trunk, neck, and arms.

Most solitary mastocytomas are about 1–5 cm in diameter and are
seen as skin areas that are colored yellow to brown and present as
minimally elevated plaques with a smooth shiny surface having a
soft to rubbery consistency. The lesion turns edematous and itchy
on manipulation [rubbing or trauma to the lesion]. Mild tender-
ness and the formation of vesicles or bulla can also occur4. These
features can sometimes be so mild that they may not come to the
attention of parents.

Diagnosis is by biopsy that reveals a dense monomorphic inflam-
matory infiltrate consisting of round to oval mast cells containing
a clear cytoplasm and centrally located nuclei in the dermis. Con-
firmation of diagnosis is usually by special staining with toluidine
blue that reveals the metachromatic staining of the monomorphic
mast cells5.

The course of solitary mastocytomas is benign and the disease is
self-limited. Systemic involvement is uncommon and complete
spontaneous resolution is expected in months to years6, time.
Reassurance along with avoidance of triggering factors such as

pressure, friction (rubbing or toweling of the lesion), physical exer-
tion, extreme temperature changes, emotional stress, intake of mast
cell degranulating agents like aspirin, NSAIDS, morphine, codeine
(particularly in cough preparations), alcohol and radio contrast dyes
are of utmost importance8.

In symptomatic patients, oral H1 and H2 antihistamines are com-
monly used. Topical steroids with or without occlusion, intral-
esional steroids, oral sodium cromoglycate, oral ketotifen and
surgical excision are other treatment options6,7. Though topical
steroids have shown good results, their topical and systemic side
effects are a matter of concern, especially when treating infants.

Tacrolimus and pimecrolimus are topical immunomodulators, the
first in a new class of topical calcineurin inhibitors. These drugs act
as immunosuppressants by binding to a cytosolic ligand in the cyto-
plasm of T cells called FK506-binding protein (FKBP) and inhibit
the cytoplasmic enzyme calcineurin, thus inhibiting the activation
and maturation of T cells and blocking transcriptional activation of
several cytokine genes – interleukin (IL)-2 [mainly], IL-4, IL-10,
interferon-γ, tumor necrosis factor-α, and granulocyte–macrophage
colony-stimulating factor8.

Other immunomodulatory effects of tacrolimus include the inhibi-
tion of mast cell adhesion and the inhibition of the release of media-
tors from mast cells and basophils9, which might explain its efficacy
in the improvement of the lesion and alleviation of the symptoms in
cutaneous mastocytosis.

These immunomodulators offer advantages over corticosteroids
in terms of a more selective action, no associated systemic side-
effects, and the absence of associated skin atrophy, depigmentation
and telangiectasia.

This case report demonstrates that topical calcineurin inhibitors can
be considered as a safe and efficacious modality of treatment in
cutaneous mastocytoma.

Consent
Written informed consent for publication of the clinical details and
clinical images was obtained from the father of the patient.

Author contributions
Dr. Sukesh M.S. and Dr. Ameet Dandale were involved in clinical
diagnosis, work-up, treatment and writing up of this case report.
Dr Smita Ghate contributed to the histopathologic diagnosis, Dr Rachita Dhurat contributed to the conception and design and
final approval of the paper; Dr Ankur Sarkate contributed to the
assimilation of all data and the histopathological pictures.

Competing interests
No competing interests were disclosed.

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References

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The authors described an 18-mo old girl with a solitary mastocytoma, which was successfully treated with tacrolimus. This might suggest that the mastocytoma requires therapy. This is not the case. Mastocytomas are self-limited and usually don’t need therapy. The most important management is the avoidance of known trigger factors. It is not clear whether the reduction is due to the self-limiting nature of the tumor or to the therapy.

Pathogenetically and from the mechanism of action of tacrolimus, which prevents mast cell degranulation, an improvement of mastocytoma can be expected. However it should be made clear that the indication for treatment of a solitary mastocytoma should be made very cautiously.

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.

Competing Interests: No competing interests were disclosed.

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I thank the authors for this interesting article.

I believe it is worthy of indexation because there is surprisingly little in the literature about this alternative treatment. I might make a few small suggestions for consideration:

1. Is solitary mastocytoma really "rare"? I see an awful lot of them for such a designation even taking into consideration referral bias. Uncommon might be a better descriptor.

2. Could the line "Diagnosis is by biopsy..." be modified? I do not think these absolutely must be biopsied and in fact almost never do. If there is a + Dariers sign and a strong clinical suspicion this presentation is specific enough that I do not think biopsy is mandatory. As a pediatric dermatologist I do all I can to avoid biopsying when not absolutely necessary and I worry readers might take this
line to imply diagnosis mandatory for diagnosis of mastocytoma. It is not.

3. Can we still call tacrolimus and pimecrolimus "new" given they have been available almost 15 years now?

4. I think in fairness the authors must mention the boxed warning about this class of medications somewhere. If the authors cite the concerns for topical and systemic side effects of topical steroids as they do I think they must balance this by mentioning the biggest barrier to using these agents - the black box.

5. I might have included two references the authors omitted:
     This describes two cases similar to the authors' treated successfully with pimecrolimus
     Cutaneous mastocytosis - albeit not mastocytoma - treated with a number of things including topical tacrolimus.

I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

**Competing Interests:** No competing interests were disclosed.
I have read this submission. I believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.

Competing Interests: No competing interests were disclosed.