Short Communication

Macular Amyloidosis in Kashmiri Females

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Background: Macular amyloidosis, the commonest form of primary localized cutaneous amyloidosis is a pruritic eruption consisting of small, dusky brown or greyish pigmented macules distributed symmetrically over the upper back and arms. Aim: To study the clinical and histological features of macular amyloidosis. Material and Methods: Fifty Kashmiri females in the reproductive age group with a clinical diagnosis of macular amyloidosis were evaluated after performing skin biopsy and staining with Congo red. Results: Mean age of females was 27.8±2.34 years and mean duration of lesions was 32.3±4.2 months. History of itching, friction, hair style changes, use of cosmetics and family history was positive in most of the patients. Amyloid deposits were observed in 42% of biopsies. Conclusion: Macular amyloidosis is the common cause of truncal pigmentation in adult females. Role of sex hormones needs to be evaluated.

Keywords: Truncal pigmentation, amyloidosis, friction.

INTRODUCTION

Macular amyloidosis (MA) or frictional amyloidosis is a pruritic eruption consisting of small, dusky-brown or greyish pigmented macules distributed symmetrically over the upper back and arms. About 50% of patients have a reticulated or rippled pattern of pigmentation. This condition is relatively unknown in western countries with a higher prevalence in Asia and Middle East (Taheri 2007; Rados et al., 2008). MA is the commonest form of primary localized cutaneous amyloidosis (PLCA) and various incriminating factors include genetics, sunlight, friction, female gender, atopy, environmental factors and EBV (Rados et al., 2008). MA usually presents as small 2-3mm greyish brown or brown pruritic (82%) or nonpruritic (18%) macules, which gradually join to form symmetric patches with a characteristic rippled pattern that most frequently involves the interscapular area and less frequently the upper arms, chest and thighs (Eswaramoorthy et al., 1999).

MATERIAL AND METHODS

Fifty Kashmiri females in the reproductive age group with a clinical diagnosis of macular amyloidosis (confirmed by at least two observers) were enrolled in the study at SKIMS medical college hospital for a period of two years, after obtaining clearance from institutional ethical committee. A detailed history was elicited in relation to the etiological factors followed by cutaneous examination. A detailed clinical history of each patient was taken regarding duration and precipitating factors like friction, sunlight, hair styling. History of other possible causes of pigmentation such as drugs, cosmetic use, atopy, previous inflammatory conditions and systemic illness was taken. A thorough cutaneous examination of the patient was done for the sites of pigmentation. Oral mucosa and nails were examined for pigmentation. General physical and systemic examination was also done. Routine laboratory investigations like haemogram, liver, renal and thyroid function tests were carried out. Skin biopsy was performed on all the patients having clinical signs of the disease. Specimens were then stained with congo red with polarization.

RESULTS

All the females were in the reproductive age group of 17-
Table 1. Histopathological findings of macular amyloidosis patients

<table>
<thead>
<tr>
<th>Histopathological findings</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyperkeratosis &amp; acanthosis</td>
<td>47</td>
</tr>
<tr>
<td>Basal cell pigmentation</td>
<td>40</td>
</tr>
<tr>
<td>Pigment incontinence</td>
<td>42</td>
</tr>
<tr>
<td>Condensed collagen in papillary dermis</td>
<td>45</td>
</tr>
<tr>
<td>Eosinophilic deposits in papillary dermis</td>
<td>14</td>
</tr>
<tr>
<td>Positive congo red staining</td>
<td>21</td>
</tr>
</tbody>
</table>

Figure 1. Rippled macular greyish pigmentation on upper back.

Figure 2. Rippled macular pigmentation on neck and supraclavicular area.

54 years with the mean age of 27.8±2.34 years. Out of 50 patients, 46% were in the age group of 21-30 years, followed by 22% in 11-20, 18% in 31-40, 10% in 41-50 and 4% in 51-60 year age group. Patients with Fitzpatrick skin type IV had a more delayed onset than in those with skin types III and II. The duration of lesions varied from 3 months to 8 years with a mean duration of 32.3 ±4.2 months. Itching was present in 28 patients with varying severity, 10 patients had mild, 13 moderate and 5 had severe itching. History of friction was present in 5 patients for years and objects mainly used were bath sponge, brush and plant stick. Only 6 patients had history of cosmetic use and 3 had the habit of changing hair styles frequently. Eight patients had similar history in the family and 2 patients had history of atopy. Menstrual history was abnormal in 9 patients with menorrhagia, irregular cycles and oligomenorrhea being the common.

Three patients attributed their lesions to pregnancy. Sun protected sites were commonly involved with interscapular area in 30%, neck in 22%, forearms in 18%, infraclavicular in 14%, arms in 10% and shins in 6%. Lichen amyloidosis was seen on the shins in 3 patients. None of our patients had any specific systemic disease except for diabetes mellitus in 2 patients and hypothyroidism in 3 patients. Histopathological findings are tabulated in table 1. Congo red staining was positive in 21 patients.

DISCUSSION

Macular amyloidosis is a relatively common cutaneous disease in Asia and the Middle east (Rados et al., 2008; Siraqusa et al., 2001). Age distribution is mainly between
20 and 50 years of age (Rasi et al., 2004). The sex distribution is reported to be 9:1 in females and males (Rasi et al., 2004). The diagnosis of macular amyloidosis is based on long term persistence of the disorder, localized pruritus and constant scratching urge, greyish-brown pigmentation over the scapula and detection of amyloid in histologic slides (Shanon and Sagher 1970). Rippled pigmentation is not necessarily specific or diagnostic of amyloidosis and is associated with atopic dermatitis (Onuma et al., 1994). Siraqusa M et al. described friction amyloidosis to be caused by the use of cotton towels, horse-hair gloves or artificial and rough sponges to clean the skin (Siraqusa et al., 2001). There is history of prolonged rubbing over a period of 2-5 years with various objects such as bath sponges, brushes, towels, plant sticks and leaves (Rasi et al., 2004; Shanon and Sagher 1970).

It is thought that the amyloid may be partly derived from the products of apoptotic necrosis (Chang et al., 1999). Many cases of long-standing notalgia paraesthetica result in the formation of amyloid, possibly secondary to chronic friction (Venkataram et al., 2001; Goulden et al., 2004). Histochernically, H and E stain can give a primary clue for the diagnosis of amyloidosis and crystal violet stain is a very simple and sensitive method to detect the existence of amyloid (Looi 1991). No beta 2-microglobulin and advanced glycation end product immunoreactivity was found in the amyloid deposits of macular amyloidosis unlike nodular amyloidosis (Fujimoto et al., 2002). The amount of amyloid present in macular amyloidosis is often very small and difficult to detect (Wang et al., 2001). Biphasic amyloidosis ie, macular amyloidosis with lichen amyloidosis is seen in 25% of primary cutaneous amyloidosis (Bedi and Datta 1979). Histologically MA differs from PCA by the smaller size of amyloid deposits in the papillary dermis. There is no difference in their tinctorial and immunohistochemical characteristics, deposits are permanganate-resistant and negative for AA protein, Ig light chains and keratin (Al-Ratrout and Satti 1997; Breathnach 1988; Manabe et al., 1987).

MA and LA generally follow a chronic course with intractable pruritus. There have been isolated reports of the beneficial effect of dermabrasion, topical dimethylsulfoxide, etretinate, cyclophosphamide, cyclosporine and recently 532-nm and 1064-nm Q-switched Nd:YAG laser therapy for reduction of pigmentation in macular amyloidosis (Marschalko et al., 1988; Behr et al., 2001; Ostovari et al., 2008).

To conclude, macular amyloidosis is a common cause of truncal pigmentation in females in Kashmir with no definite etiology and being common in the reproductive age group, role of sex hormones needs to be evaluated taking a larger sample of patients.
REFERENCES


