Acquired Bilateral Telangiectatic Macules
A Distinct Clinical Entity

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Within the last several years, we observed 13 adult patients with multiple telangiectatic pigmented macules confined mostly to the upper arms. Until now, these patients might have been diagnosed with conditions such as telangiectasia macularis eruptiva perstans (TMEP) or acquired brachial cutaneous dyschromatosis (ABCD). Herein, we report the findings of further analysis of these clinical and histologic findings.

Report of Cases

All patients provided their written informed consent, and the study was approved by the Ajou University Hospital institutional review board. We examined 13 patients with multiple telangiectatic pigmented macules on the upper arms who presented between January 2003 and December 2012. Chart review was performed for the clinical evaluation of the enrolled patients. Clinical features associated with age, sex, location of skin lesions, symptom duration at the time of presentation, and underlying diseases were investigated. Three-millimeter punch biopsies of both the lesional and the normal perilesional skin were performed in 7 patients and of only the lesional skin in the remaining 6 patients. Hematoxylin-eosin (H&E) staining was used to study the general histopathologic changes in the skin lesions. Epidermal thickness was measured under H&E staining as well. Melanin pigments were visualized with Fontana-Masson staining. Immunohistochemical staining was performed using monoclonal antibodies to human gp100 (NKI/beteb; Monosan; 1:10), tyrosinase (TYR; Thermo Scientific; 1:50), microphthalmia transcription factor (MITF; Leica Biosystems; 1:10), factor VIII–related antigen (Thermo Scientific; 1:100), podoplanin (D2-40; Cell Marque; 1:100), and c-kit (CD117; Cell Marque; 1:50).

The amount of melanin pigment was evaluated as the ratio of pigmented area to the measured epidermal area (PA/EA) under Fontana-Masson staining. Expression of NKI/beteb and TYR were calculated as the ratio of the stained area to measured EA (SA/EA). The number of MITF-positive melanocytes (Mc) per 1-mm length of rete ridge (Mc/1R) was counted. The number of factor VIII-related antigen-positive vessels per 1 mm² area within a 0.1-mm distance from the dermal-epidermal junction was counted. Perivascular mast cell number per vessel unit (>55 μm) was compared under c-kit staining.

IMPACT
We evaluated 13 distinct patients with multiple telangiectatic pigmented macules confined mostly to the upper arms to determine if the clinical and histopathological features of these cases might represent a specific clinical entity.

OBSERVATIONS
We retrospectively investigated the clinical, histopathologic, and immunohistochemical features of 13 patients with multiple telangiectatic pigmented macules on the upper arms who presented between January 2003 and December 2012. Epidermal pigmentation, melanogenic activity, melanocyte number, vascularity, epidermal thickness, and perivascular mast cell number of the specimens were evaluated. Clinically, the condition favored middle-aged men. On histopathologic examination, the lesional skin showed capillary proliferation and telangiectasia in the upper dermis. Histochemical and immunohistochemical analysis revealed basal hyperpigmentation and increased melanogenic activity in the lesional skin (P < .05). No significant difference in epidermal thickness or mast cell number was observed between the normal perilesional skin and the lesional skin.

CONCLUSIONS AND RELEVANCE
The clinical and histopathologic features of these lesions were relatively consistent in all patients. In addition, the features are quite distinct from other diseases. Based on clinical and histologic features, we suggest the name acquired bilateral telangiectatic macules for this new entity.
For statistical analysis, an image analysis program (Image-Pro PLUS software, version 4.5; Media Cybernetics) was used. A Kruskal-Wallis test was performed using SPSS Statistics Desktop 20.0.0 software (IBM). A $P < .05$ was considered statistically significant.

Mean patient age was 42.9 years (age range, 37-52 years; 11 men, 2 women), and all patients had Fitzpatrick skin types III and IV. The skin lesion duration ranged from 1 to 8 years. None of the patients had a family history of similar skin lesions or specific medical history such as a chronic drug intake. All patients had asymptomatic, irregular, dark red to brown telangiectatic macules developed insidiously and distributed over both upper arms (Figure 1). Darier sign was negative in all cases. The skin sites of disease involvement included both upper arms (13 of 13 patients), both lower arms (7 of 13 patients), both thighs (1 of 13 patients), trunk (1 of 13 patients), and neck (1 of 13 patients) (Table 1).

Twelve patients were observed in follow-up without specific treatment, and none of the patients showed improvement. Mean duration of follow-up was 3.8 years (range, 1.3-10.3 years). One patient was treated with intense pulsed light (IPL; Lumenis One, LUMENIS) at 3- to 4-week intervals. Treatment fluence ranged from 21 to 23 J/cm². Energy was delivered in double pulse trains of 3.0 milliseconds with pulse delays of 120 milliseconds. Cutoff filters of 590 nm were used. After 4 treatment sessions, the skin lesions showed moderate improvement.

Histopathologic examination revealed mild capillary proliferation and telangiectasia in the lesional skin of all the patients. No D2-40 immunoreactivity was observed in the lesional or normal perilesional skin. The lesional skin specimens showed marked hyperpigmentation in the basal layer of the epidermis on H&E staining. Capillary proliferation and perivascular inflammatory cell infiltration in the dermis was observed (Figure 2). A significant difference was observed in mean (SD) epidermal pigmentation in the normal perilesional and lesional skin (PA/EA, 0.097 [0.057] vs 0.249 [0.062]) ($P = .02$). The mean (SD) NKI/beteb levels (SA/EA, 0.003 [0.002] vs 0.017 [0.009]) ($P = .07$) and number of melanocytes (Mc/1R, 1.861 [0.808] vs 3.882 [1.633]) ($P = .12$) did not differ significantly between the perilesional and lesional skin. However, TYR levels (SA/EA, 0.005 [0.003] vs 0.032 [0.015]) ($P = .046$), which are used as markers of melanogenic activity, were significantly higher in the lesional skin compared with the normal perilesional skin (5.626 [2.707] mm² vs 10.648 [4.943] mm²) ($P = .046$). The mean epidermal thicknesses of the normal and lesional skin samples were 31.10 μm and 33.62 μm, respectively, not significantly different ($P = .62$).

### Table 1. Demographic Data for All 13 Study Patients

<table>
<thead>
<tr>
<th>Patient No./Sex/ Disease Duration, y</th>
<th>Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/M/1-1/2</td>
<td>Both upper arms</td>
</tr>
<tr>
<td>2/F/3</td>
<td>Both upper arms</td>
</tr>
<tr>
<td>3/M/2</td>
<td>Both upper arms</td>
</tr>
<tr>
<td>4/M/4</td>
<td>Both upper arms, both anterolateral thighs</td>
</tr>
<tr>
<td>5/M/5</td>
<td>Both upper and lower arms</td>
</tr>
<tr>
<td>6/M/5</td>
<td>Both upper arms</td>
</tr>
<tr>
<td>7/M/8</td>
<td>Trunk, both upper and lower arms</td>
</tr>
<tr>
<td>8/M/2</td>
<td>Both upper and lower arms</td>
</tr>
<tr>
<td>9/M/3</td>
<td>Neck, both upper and lower arms</td>
</tr>
<tr>
<td>10/F/3</td>
<td>Both upper and lower arms</td>
</tr>
<tr>
<td>11/M/1</td>
<td>Both upper and lower arms, both legs</td>
</tr>
<tr>
<td>12/M/NA</td>
<td>Both upper arms</td>
</tr>
<tr>
<td>13/M/NA</td>
<td>Both upper and lower arms</td>
</tr>
</tbody>
</table>

Abbreviation: NA, not assessed.
perivascular mast cell number of the normal and lesional skin samples using c-kit staining. The mean numbers of mast cells in the normal and lesional skin samples were 2.18 and 2.50, respectively, and no significant difference was noted ($P = .50$) (Table 2).

### Discussion

The clinical features of these lesions were relatively consistent in all patients. The lesions occurred insidiously and mostly...
in men during middle age (age 32-50 years), and the patients had irregular and dark red to brown telangiectatic macules on both upper arms. None of the patients had a history of excessive sun exposure.

Some disorders should be discussed in the differential diagnosis. Clinically, TMEP is characterized by telangiectatic tan to brown macules on the trunk and extremities. Histologically, TMEP shows infiltrates of mast cells, mainly in the upper dermis usually clustered around dilated capillaries and venules. Unlike TMEP, our cases showed no significant difference in perivascular mast cell number was observed between the lesional and perilesional skin in our patients.

The newly described ABCD is a disorder characterized clinically by asymptomatic gray-brown patches on the arms. However, in contrast to our cases, it is accompanied by epidermal atrophy and slight actinic elastosis and occurs on the dorsal aspects of the forearms in postmenopausal women.

Generalized essential telangiectasia is characterized by the dilatation of veins and capillaries without preceding or coexisting skin lesions. However, it involves a large segment of the body and develops most frequently in women in their 40s and 50s, usually appearing first on the lower legs and then spreading to the upper legs, abdomen, and arms.

Poikiloderma is a morphologic term combining atrophy, telangiectasia, and pigmentary changes over an area of the skin. However, unlike patients with poikiloderma, our patients showed no significant difference in epidermal thickness.

Unilateral nevoid telangiectasia syndrome involves large punctate and stellate telangiectases on the skin with characteristic unilateral occurrence. These presentations can frequently be assigned to certain dermatomes.

Solar lentigines commonly occur as multiple lesions in sun-exposed areas such as the face and extensor surfaces of the forearms. Histologically, the rete ridges are subtly elongated. In contrast, our patients had skin lesions mainly on their upper arms, and rete ridge elongations were not observed on histologic examination.

In the present study, epidermal hyperpigmentation and increased vessel density were observed in the lesional skin. In recent studies, more vascularization was observed in hyperpigmented lesions than in the normal perilesional skin in melasma. These results support the hypothesis that vascular components may be closely correlated with cutaneous pigmentation. Although the number of melanocytes in our cases was not increased in the lesional skin, melanogenic activity was increased. The exact vascular factor associated with melanogenesis has not yet been determined. However, recent studies have proposed that vascular endothelial growth factor (VEGF) might play a key role in melanogenesis. Kim et al suggest that VEGF might not only affect cutaneous angiogenesis but also exert paracrine effects on melanocytes. It is known that VEGF activates the arachidonic acid pathway, which might affect melanogenesis. An additional in vivo study is needed to support these data.

Because most of the patients described herein showed no significant change of the skin lesions during follow-up, this condition is thought to be chronic and persistent. However, moderate improvement was observed in a patient treated with IPL, which might be effective in treating both hyperpigmentation and erythema in this condition because of its broad spectrum of light.

Conclusions

We describe herein an acquired telangiectatic disorder with an adult male predominance that involves pigmentation mainly on the upper arms. Based on its clinical and histologic features, we suggest the name acquired bilateral telangiectatic macules for this entity.

REFERENCES