Erythromelanosis Follicularis Faciei: A Case Report and Review of the Literature

Khalid Al Hawsawi, Ohood Aljuhani, Ghassan Niaz, Haneen Fallatah, Abrar Alhawsawi

Key Words
Erythromelanosis follicularis faciei · Erythromelanosis follicularis faciei et colli · Keratosis pilaris

Abstract
Erythromelanosis follicularis faciei is a rare sporadic condition of unknown etiology characterized by reddish-brownish patches and follicular papules that appear commonly on the face and rarely on the neck. Herein, we report a 16-year-old male who had asymptomatic facial skin lesions since early childhood. His family history revealed a similar case in his younger brother. His parents are not consanguineous. Skin examination revealed diffuse nonscaly brownish patches with erythematous background and multiple skin-colored, hypopigmented follicular papules on both cheeks. A summary of previous reports of erythromelanosis follicularis faciei in the literature is presented in this report.

Introduction
Erythromelanosis follicularis faciei (EFF) is a rare sporadic condition of unknown etiology characterized by erythematous hyperpigmented patches and follicular papules on the face. It was first described in Japanese patients in 1960 by Kitamura and collaborators. When the neck is affected, the condition is called erythromelanosis follicularis faciei et colli (EFFC) [1]. The pathogenesis is unknown. However, a combination of vasodilation and hyperpigmentation has been found in the affected areas. Some authors consider EFF as part of the spectrum of keratosis pilaris atrophicans disorders [2]. EFF is characterized clinically by the
presence of red-brown patches on the lateral aspects of the cheeks, and rarely lateral aspects of the neck. Numerous pinhead-sized follicular papules are present within the involved areas that may sometimes appear relatively hypopigmented. Bilateral distribution is the main characteristic, but unilateral cases have been described [1–15]. EFF is usually asymptomatic. However, a burning sensation has been described in a few patients [16–21]. Keratosis pilaris elsewhere in the body is a common association with EFF. Histopathologically, there are hyperkeratosis, slight follicular hyperkeratosis (follicular plugging), increased basal layer pigmentation, dilatation of superficial dermal blood vessels, and periadnexal lymphocytic infiltrate [9].

**Case Report**

A 16-year-old male presented with asymptomatic facial skin lesions which he had since early childhood. There was no history of predisposing factors. He had been using topical treatment but without any help. Family history revealed a similar case in his younger brother. His parents are not consanguineous. Skin examination revealed diffuse nonscaly reddish-brownish patches and multiple skin-colored, hypopigmented follicular papules on both cheeks (fig. 1). On the basis of the above classical clinical findings, the diagnosis of EFF was made. The patient was reassured and put under periodic follow-up.

**Discussion**

Erythromelanosis follicularis faciei (EFF) is a pigmentary disease associated with erythema and follicular papules on the face. It affects all races. However, it shows a preponderance in the people of Asian ancestry [10–20]. The cause is unknown, but the hereditary component (autosomal recessive) seems to play a role in the pathogenesis [12, 19, 22, 24]. Table 1 summarizes the previous reports of erythromelanosis follicularis faciei (EFF) in the literature.

EFF primarily affects adolescents. However, it has been reported in children as young as 2 years old and in adults as old as 46 years old. Similarly, the onset of the disease shows a wide range, starting from birth to as old as 43 years old. The male:female ratio is 2:1 [1–24]. Differential diagnoses include keratosis pilaris rubra, poikiloderma of Civatte, Riehl’s melanosis, and pigmented peribuccal erythrosis of Brocq. In skin type I patients, there may be only erythema, leading to a significant overlap with keratosis pilaris rubra, and it remains to be answered whether EFFC and keratosis pilaris rubra are two spectrums of the same condition [2, 9–18]. Poikiloderma of Civatte is observed in middle-aged women as reticulated dyschromia with atrophy and erythema affecting preferably photoexposed areas and sparing the submental region [5–15]. Treatment is not well defined. Various modalities have been described. Topical agents have been used, including, ammonium lactate, retinoids, hydroquinone, vitamin C, salicylic acid peels (20–30%), glycolic acid peels, tacacone ointment, and metronidazole gel. The evidence for their use is anecdotal. Limited courses of isotretinoin (0.1–1 mg/kg/day) have been tried in severe cases. A combination of laser treatment (pulsed dye laser) for erythema and Q-switched Nd:YAG laser for hyperpigmentation have been tried but they require multiple sessions [1–24].
Statement of Ethics

Consent has been obtained from the parents of the patient for the purpose of using patient’s photographs for print or online publication.

Disclosure Statement

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References

## Table 1. Summary of previous reports of EFF in the literature

<table>
<thead>
<tr>
<th>First author</th>
<th>Cases, n</th>
<th>Patient age, years</th>
<th>Gender</th>
<th>Patient age at disease onset</th>
<th>EPP cases in the family, n</th>
<th>Site of lesion</th>
<th>Keratosis pilaris</th>
<th>Other associated conditions</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shahshahani [1]</td>
<td>60</td>
<td>range: 4–39 years; average: 22</td>
<td>M = 43 F = 17</td>
<td>at birth, 3 cases 1st decade, 16 cases 2nd decade, 33 cases 3rd decade, 1 case</td>
<td>8</td>
<td>cheeks and neck</td>
<td>trunk, arms, thigh</td>
<td>milia in 4 cases</td>
<td>not available</td>
</tr>
<tr>
<td>Volks [2]</td>
<td>3</td>
<td>15 years</td>
<td>F</td>
<td>no</td>
<td>cheeks</td>
<td>trunk, limbs</td>
<td>no</td>
<td>no</td>
<td></td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>F</td>
<td>1 year</td>
<td>no</td>
<td>cheeks</td>
<td>limbs</td>
<td>no</td>
<td></td>
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<tr>
<td></td>
<td>4</td>
<td>F</td>
<td>birth (mother)</td>
<td>no</td>
<td>cheeks, forehead</td>
<td>arms, limbs</td>
<td>no</td>
<td></td>
<td></td>
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<tr>
<td>Silva [3]</td>
<td>1</td>
<td>11</td>
<td>M</td>
<td>years</td>
<td>no</td>
<td>face, shoulder, arms</td>
<td>face, neck</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>Augustine [4]</td>
<td>3</td>
<td>19</td>
<td>F</td>
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<td>(sister and brother)</td>
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<td>upper back, shoulders, arms</td>
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<tr>
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<td>F</td>
<td>6 years</td>
<td>no</td>
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<td>upper back, shoulders, arms</td>
<td>no</td>
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</tr>
<tr>
<td></td>
<td>13</td>
<td>M</td>
<td>8 years</td>
<td>no</td>
<td>cheeks</td>
<td>shoulders</td>
<td>no</td>
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<tr>
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<td>2</td>
<td>17</td>
<td>F</td>
<td>12 years</td>
<td>no</td>
<td>face, neck, arms</td>
<td>arms, upper back, thigh</td>
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<td>topical salicylic acid 2% and retinoic acid 0.01%</td>
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<tr>
<td></td>
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<td>F</td>
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<td>no</td>
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<td>arms, upper back, thigh</td>
<td>no</td>
<td></td>
<td></td>
</tr>
<tr>
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<td>1</td>
<td>17</td>
<td>M</td>
<td>11 years</td>
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<td>cheeks, lower lip and auricles</td>
<td>shoulder areas</td>
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<td>24</td>
<td>F</td>
<td>23 years</td>
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<td>not available</td>
<td>no</td>
<td>topical retinoic acid</td>
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<tr>
<td></td>
<td>14</td>
<td>F</td>
<td>10 years</td>
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<td>no</td>
<td></td>
<td></td>
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<td>2</td>
<td>26</td>
<td>M</td>
<td>not available</td>
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<td>not available</td>
<td>no</td>
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<td></td>
<td>22</td>
<td>M</td>
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<td>no</td>
<td>cheeks</td>
<td>not available</td>
<td>no</td>
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<tr>
<td>Kim [9]</td>
<td>10</td>
<td>range: 12–46 years; average: 22</td>
<td>M = 8 F = 2</td>
<td>range: 8–43 years; average: 16.5 years</td>
<td>no</td>
<td>cheeks and neck</td>
<td>1 patient with keratosis pilaris on arms</td>
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<td>18</td>
<td>M</td>
<td>childhood</td>
<td>1 (brother)</td>
<td>maxilla, cheeks, neck</td>
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<td>Tuzun [12]</td>
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<td>17</td>
<td>M</td>
<td>10 years</td>
<td>2 (sister and father)</td>
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<td>arms and trunk</td>
<td>diabetes mellitus and congenital leukokeratosis</td>
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<tr>
<td>Lee [13]</td>
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<td>18</td>
<td>M</td>
<td>13 years</td>
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<td>cheeks, neck</td>
<td>no</td>
<td>no</td>
<td>no</td>
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<td>35</td>
<td>F</td>
<td>25 years</td>
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<td>cheeks</td>
<td>arms</td>
<td>no</td>
<td>topical ammonium lactate 12% or metronidazole gel</td>
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<td></td>
<td>43</td>
<td>F</td>
<td>24 /2 years</td>
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<td></td>
<td></td>
<td></td>
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<td></td>
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<tr>
<td>McGillis [15]</td>
<td>1</td>
<td>13</td>
<td>F</td>
<td>8 years</td>
<td>no</td>
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<td>upper arms</td>
<td>no</td>
<td>topical tretinoin 0.05%, ammonium lactate and hydroquinone</td>
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<tr>
<td>Watt [16]</td>
<td>1</td>
<td>15</td>
<td>M</td>
<td>several months</td>
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<td>upper arms</td>
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<td>no</td>
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<tr>
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<td>F = 7 M = 5</td>
<td>not available</td>
<td>not available</td>
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<td>not available</td>
<td>not available</td>
<td>dual wavelength laser (pulsed dye laser + Q-switched Nd:YAG laser)</td>
</tr>
<tr>
<td>Kim [18]</td>
<td>11</td>
<td>14.4 ± 7.7</td>
<td>M = 6 F = 5</td>
<td>not available</td>
<td>not available</td>
<td>preauricular and maxillary regions</td>
<td>not available</td>
<td>not available</td>
<td>topical tacalcitol ointment</td>
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<tr>
<td>Lalit [19]</td>
<td>1</td>
<td>21</td>
<td>M</td>
<td>19 years</td>
<td>no</td>
<td>cheeks forehead chin</td>
<td>arms, shoulders, back</td>
<td>no</td>
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</tr>
<tr>
<td>Li [20]</td>
<td>1</td>
<td>20</td>
<td>M</td>
<td>12 years</td>
<td>no</td>
<td>preauricular area, neck</td>
<td>shoulders, lateral arms, thighs</td>
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<td>dual-wavelength laser system (pulsed dye laser + Q-switched Nd:YAG laser)</td>
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<td>Sodaily [21]</td>
<td>3</td>
<td>15, 18, 28</td>
<td>M</td>
<td>childhood</td>
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<td>cheeks</td>
<td>arms and legs</td>
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<td>17</td>
<td>M</td>
<td>9 years</td>
<td>1 (sister)</td>
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<td>upper arms</td>
<td>no</td>
<td>not available</td>
</tr>
<tr>
<td></td>
<td>17</td>
<td>M</td>
<td>childhood</td>
<td>2 (mother and grandmother)</td>
<td>cheeks</td>
<td>face, neck</td>
<td>no</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sardana [23]</td>
<td>5</td>
<td>19</td>
<td>M</td>
<td>13 years</td>
<td>no</td>
<td>temples, cheeks, neck</td>
<td>arms, shoulders, back</td>
<td>not available</td>
<td>oral isotretinoin</td>
</tr>
<tr>
<td></td>
<td>13</td>
<td>M</td>
<td>not available</td>
<td>no</td>
<td>not available</td>
<td>topical retinoic acid cream</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>18</td>
<td>M</td>
<td>12 years</td>
<td>no</td>
<td>not available</td>
<td>tretinoin and hydroquinone cream</td>
<td></td>
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</tr>
<tr>
<td></td>
<td>11</td>
<td>M</td>
<td>5 years</td>
<td>no</td>
<td>not available</td>
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<td></td>
</tr>
<tr>
<td></td>
<td>13</td>
<td>F</td>
<td>not available</td>
<td>no</td>
<td>not available</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yanez [24]</td>
<td>2</td>
<td>15</td>
<td>M</td>
<td>10 years</td>
<td>1 (sister)</td>
<td>cheeks, forehead and neck</td>
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<td>topical retinoic acid cream 0.05%</td>
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<td>18</td>
<td>F</td>
<td>childhood</td>
<td>1 (brother)</td>
<td>cheeks</td>
<td>upper limbs</td>
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<td></td>
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<tr>
<td>Al Hawsawi, this study</td>
<td>1</td>
<td>16</td>
<td>M</td>
<td>childhood</td>
<td>1 (brother)</td>
<td>cheeks</td>
<td>no</td>
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<td>no</td>
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Fig. 1. Diffuse nonscaly reddish-brownish patches with multiple skin-colored, hypopigmented follicular papules on both cheeks.