Six Cases of Nevus Oligemicus

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Abstract. Hamartoma or nevus oligemicus is an uncommon lesion that is characterized by selective vasoconstriction of the deep dermal vascular plexus with respect to the superficial one and whose cause has not been clearly established. This selective vasoconstriction gives rise to fixed, acquired, and asymptomatic lesions in the form of livid, erythematous macules that are typically cold to touch compared with surrounding skin.

We report the cases of 6 young men with lesions clinically compatible with nevus oligemicus on the abdomen and flanks. Measurement of the surface temperature of the lesion revealed a decrease of up to 2.5°C with respect to healthy surrounding skin and allowed a definitive diagnosis to be made.

We describe the additional studies undertaken, the differential diagnosis, and the possible etiologic agents, and discuss the cases reported in the literature to date.

In our opinion, nevus oligemicus is an underdiagnosed lesion that is much more common than has been reported in the literature.

Key words: hamartoma, nevus, oligemicus.

CASE REPORT

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Introduction

The etymology of oligemicus is oligo, meaning little, and emicus, meaning blood; thus, the term refers to "poor vascularization." Hamartoma or nevus anemicus is defined as a circumscribed cutaneous abnormality characterized by localized vasoconstriction of the deep dermal vascular plexus (which regulates temperature) compared with more superficial dermal vessels (which handle nutrition). Clinically, the condition leads to cold, erythematous lesions. Localized hypothermia is a key finding for the diagnosis of the condition.

Nevus oligemicus was first described in 1981 and only an additional 3 cases have been reported. We report another 6 cases diagnosed in the past 5 years.

Case Description

The clinical and epidemiologic findings of the patients are summarized in Table 1. The patients were 6 men between 26 and 57 years of age; all were considerably overweight and had a sedentary profession (4 truck drivers, 1 candidate...
for a government position, and 1 bank employee). Patient 4 had undergone appendectomy and surgery for melanoma on the back 6 years earlier. All others had no relevant personal or family history. In 5 of the patients, the dermatologic examination revealed a fixed erythematous cyanotic macule with irregular borders in the middle of the abdomen (Figure 1). In the sixth, the erythematous area appeared on both flanks (Figure 2). The lesions remained stable and whitened on pressure. The skin was cold in the erythematous area, particularly in the middle of the plaque.

The laboratory workup included complete blood count, complete biochemistry, antinuclear antibodies, lupus anticoagulant, cryoglobulins, and Borrelia burgdorferi serology, and all results were normal or negative.

A skin biopsy was taken in the first 3 cases. The histologic study revealed a normal number of dermal vessels, although with dilatation of the papillary dermal capillaries and endothelial hyperplasia. In the reticular dermis, hyperplasia

Table 1. Clinical and Epidemiologic Findings

<table>
<thead>
<tr>
<th>Cases</th>
<th>Profession</th>
<th>Sex/Age, y</th>
<th>Precipitating Factors</th>
<th>Clinical Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 1</td>
<td>Truck driver</td>
<td>M/4</td>
<td>None</td>
<td>Cold erythema</td>
</tr>
<tr>
<td>Patient 2</td>
<td>Truck driver</td>
<td>M/43</td>
<td>None</td>
<td>Cold, livid erythema</td>
</tr>
<tr>
<td>Patient 3</td>
<td>Candidate for government position</td>
<td>M/26</td>
<td>None</td>
<td>Cold erythema</td>
</tr>
<tr>
<td>Patient 4</td>
<td>Truck driver</td>
<td>M/49</td>
<td>None</td>
<td>Cold erythema</td>
</tr>
<tr>
<td>Patient 5</td>
<td>Truck driver</td>
<td>M/54</td>
<td>None</td>
<td>Cold erythema</td>
</tr>
<tr>
<td>Patient 6</td>
<td>Bank employee</td>
<td>M/57</td>
<td>None</td>
<td>Cold erythema</td>
</tr>
</tbody>
</table>

Abbreviations: M, male; ND, not done.

Table 2. Clinical and Epidemiologic Findings Previously Described in the Literature

<table>
<thead>
<tr>
<th>Cases</th>
<th>Profession</th>
<th>Sex/Age, y</th>
<th>Precipitating Factors</th>
<th>Clinical Findings/Course</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fays et al(^1)</td>
<td>NK</td>
<td>M/46</td>
<td>None</td>
<td>Livid erythema</td>
</tr>
<tr>
<td>Davies et al(^2)</td>
<td>NK</td>
<td>M/16</td>
<td>Cold baths</td>
<td>Livid area</td>
</tr>
<tr>
<td>Friedel et al(^3)</td>
<td>NK</td>
<td>F/15</td>
<td>None</td>
<td>Livid, cyanotic area</td>
</tr>
<tr>
<td>Dupré et al(^4)</td>
<td>NK</td>
<td>M/45</td>
<td>None</td>
<td>Cold, cyanotic, erythematous area</td>
</tr>
</tbody>
</table>

Abbreviations: F, female; M, male; ND, not done; NK, not known.

Figure 1. Patient 2. Striae distensae and erythematous-cyanotic plaque following 15-kg weight loss.

Figure 2. Patient 6. Cold erythema on the flanks.
of the endothelial cells had caused obstruction of the capillary lumens (Figure 3).

Analysis of surface skin temperature (with an MX-TDI2388 contact thermometer, MX onda, Madrid, Spain) showed a decrease of 2°C to 2.5°C in the affected area compared with adjacent healthy skin.

Various pharmacological tests with histamine, thurfyl nicotinate, and 0.05% clobetasol propionate showed normal vasomotor responses of both the healthy skin and the hamartoma. An intradermal injection of histamine (1.0 ng histamine in 0.1 mL saline solution) plus topical application of thurfyl nicotinate caused a bump in the first case and erythema and edema in both healthy and affected skin in the second. The application of 0.05% clobetasol propionate ointment with an occlusive dressing for 2 hours caused identical intensity in the whitening of both areas.

The clinical characteristics and decreased temperature of affected skin led to a diagnosis of nevus anemicus. Only the first case received treatment with 20 mg of oral prednisone for 1 month, with a subsequent decrease to 10 mg daily the second month, with no changes observed. None of the other patients received any medications.

No improvement or worsening of the original lesions and no new lesions were observed during the follow-up period, which varied from 2 to 6 years. Patient 5 lost 15 kg, but with no changes in the plaque (Figure 1).

Discussion

Hamartoma or nevus anemicus is a common entity, understood as a functional, rather than anatomical, abnormality. The etiology is not well known, but may be due to hormone receptor abnormalities, with variations in adrenergic activity, increased sympathetic tone, and
consequently vasoconstriction and slower deep dermal vascular flow, thus cooling the lesion surface. However, the superficial vascular plexus (responsible for nutrition) is intact or thought to have suffered relative vasodilation, which led in our patients to the erythematous appearance of the lesions.\(^2,4\)

These 6 cases are consistent with the diagnosis of nevus oligemicus, first described by Davies et al\(^2\) in 1981 in a man with an area of fixed, persistent, livid erythema on the trunk, with a striking drop in local temperature compared with adjacent healthy skin.

Clinically, the lesions present as erythematous-violaceous areas that are fixed, irregular, and cyanotic or, in some cases, whitish, telangiectatic plaques.\(^1\) They are usually located in the abdominal region and thighs, but cases have also been described on the hands\(^4\) or chest and thighs.\(^5\) The main clinical characteristic is the persistence of cold skin temperatures to touch, in the absence of evident arterial ischemia in the vicinity.\(^1,2,4,5\) The main diagnostic sign is skin temperature, which is usually at least 2°C lower than the adjacent skin.\(^1,2,4,5\)

No triggers have been described except for cold baths, as reported in 1982 by Dupré and Viraben\(^4\) (Table 2). We found possible factors favoring this condition among the patients described in this article because all were obese and had a sedentary lifestyle related to their profession. Although 1 patient lost 15 kg, resolution of the lesions did not occur. Despite most lesions appearing above the belt adjustment area, we were unable to determine if pressure was the source of the abnormality because the nevi oligemicus did not disappear when a belt was not worn.

Laboratory analysis and immunologic studies are normal and histopathology shows discrete vasodilatation of the superficial dermis and obstruction of the deep vessels, as reported by Dupré and Viraben\(^4\) and observed in the 3 patients from our study in whom biopsy was performed. However, skin biopsy is not essential to diagnose the condition, which is essentially clinical and demonstrated by temperature measurement and localized hypothermia.

The differential diagnosis can be performed with inflammatory erythema and capillary hemangioma.\(^2\) In the first case, the erythema is not fixed, and there are local increases in temperature and epidermal changes seen in the histologic study. Hemangiomas are associated with characteristic histologic findings not found in nevus oligemicus.

We consider nevus oligemicus to be a common condition, perhaps underreported by patients, underdiagnosed, or even minimally reported in dermatology journals. We contribute 6 new cases to add to those reported in the literature.

**Conflicts of Interest**
The authors declare no conflicts of interest.

**References**