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A case of adult-onset multiple angiokeratomas with zosteriform distribution

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Abstract
Angiokeratomas are benign vascular ectasias in the papillary dermis associated with epidermal changes in the form of hyperkeratosis and/or acanthosis. Clinically, angiokeratomas appear as solitary or multiple dark red to purple-black macules and/or papules, mostly with a verrucous surface. Five subtypes of angiokeratoma have been proposed — angiokeratoma corporis diffusum, angiokeratoma of Mibelli, angiokeratoma of Fordyce, angiokeratoma circumscriptum, and “solitary and multiple” angiokeratomas. We report an unusual case of multiple angiokeratomas in a zosteriform distribution with onset at age 74.

Keywords: angiokeratoma, angiokeratoma circumscriptum, zosteriform, unilateral, mosaicism, elderly, adult, vascular lesions

Introduction
The prevalence of angiokeratomas has been reported as approximately 0.16\% [1]. Overall, there is a slight male predominance and they are rarely seen in darker skin types. The “solitary and multiple” subtype may be located on any part of the body and typically appears in the second to fourth decades [2]. Well over 80\% of cases that fall into this subtype are solitary. When multiple, they are more common on the lower legs, but generally do not show any recognizable clustering or patterning. Herein, we report an unusual case of adult-onset multiple angiokeratomas with zosteriform distribution in an elderly male [3].

Case Synopsis
A 76-year-old man presented with a two-year history of lesions, which spontaneously developed on his right buttocks and posterior thigh. Physical exam revealed multiple 2-3mm purple papules on the right buttock (Figure 1) and right posterior thigh in addition to two typical, solitary angiokeratomas on the scrotum. The patient denied pain and itching of the lesions, but did report occasional bleeding. There was no family history of similar lesions. The patient’s past medical history was significant for hypertension, monoclonal gammopathy of unknown significance, coronary artery disease/stent, and mitral valve repair.

A punch biopsy extending to the level of the deep reticular dermis was taken from the right buttock. Histopathological examination demonstrated thin-walled dilated vascular spaces in the papillary dermis surrounded by epithelial collarettes. There was variable hyperkeratosis and acanthosis (Figure 2). There was no endothelial cell atypia and no involvement of the reticular dermis. Based on the clinical and histopathological features, a diagnosis of multiple angiokeratomas with zosteriform distribution was made.

Case Discussion
In 1889, the term ‘angiokeratoma’ was first coined by Vittorio Mibelli who used the term to refer to vascular lesions on the dorsum of the hands and feet in a 14-year-old girl [1]. At present, angiokeratomas are subdivided into five subtypes: (1) angiokeratoma corporis diffusum, typically associated with Fabry
disease; (2) angiokeratoma of Mibelli, occurring on acral regions and often associated with chillblains; (3) angiokeratoma of Fordyce, located on the scrotum and vulva; (4) angiokeratoma circumscription, nevoid and with an onset in early life; (5) solitary and multiple angiokeratoma. The latter is the most frequent subtype, with 83% of patients presenting with solitary angiokeratoma; the least frequent subtype is angiokeratoma circumscription [4]. The exact mechanism for the development of angiokeratomas is unknown but proposed causal factors include pregnancy, congenital causes, chillblains, trauma, and tissue asphyxia [2]. Though the aforementioned subtypes differ clinically from each other by location, morphology, and epidemiology, they can be histologically similar (Table 1).

Our patient does not fit in the category of angiokeratoma corporis diffusum owing to the late onset, localized distribution, and lack of any signs/symptoms of Fabry disease. Angiokeratoma of Mibelli is acral in distribution and is generally more plaque-like or hyperkeratotic. The patient does have two scrotal lesions in the S2 dermatome, raising the possibility of concurrent angiokeratoma of Fordyce. However, the predominant findings are extragenital and largely in the S3 and S4 dermatomes, which necessitates a separate diagnosis.

The individual lesion morphology in our case fits best with the “solitary and multiple” subtype of angiokeratoma, but the zosteriform distribution is distinctive. Similar cases have been reported in the past, but never at such a late age of onset. Eizaguirre et al. described two sisters with multiple angiokeratomas in zosteriform distribution, with an age of onset at age 10 and 12. We believe that our current case is similar to theirs, although at a much later onset.
only scattered reports of late-onset angio keras toma
circumscriptum. Del Pozo presented two late onset
cases [17]. The first is a female who presented with
an asymptomatic, hyperkeratotic, violaceous plaque
on her left buttock at the end of the first decade of
life. The second patient presented with a similar
lesion on her right buttock in the second decade of
life. Kwon et al. reported a case of angio keras toma
circumscriptum developing on the chest of a man—
in the third decade [18]. Interestingly, the lesion
developed following injury to the area. In our current
case, the individual lesion morphology is not plaque-
like, but is that of typical solitary or multiple
angio keras tomas, albeit in an unusual distribution.
For these reasons, along with the very late onset, we
believe the current case is most similar to the
previously described ‘multiple angio keras tomas with
zosteriform distribution’ [4].

The etiology of this presentation is uncertain. The
zosteriform distribution is suggestive of mosaicism.
Bechara et al. postulated that perhaps the late onset
could be explained by a secondary environmental
factor superimposed on preexisting mosaicism [19].
Alternatively, the ‘late onset’ could be analogous to
the hypertrophy noted in late lesions of nevus
flammeus [17]. However, the lack of any visible pre-
existing lesion would argue against this.

Treatment of solitary or multiple angio keras tomas is
often not needed owing to its clinically innocuous
nature. For cosmetic purposes, small lesions may be
managed by curettage and electrocautery, cy
rosurgery, and diathermy, whereas larger lesions
may respond to laser ablation (carbon dioxide or
argon laser), [17]. Given that our patient was
asymptomatic, no treatment was undertaken.

**Conclusion**

In summary, we present an unusual case of acquired
angio keras tomas in an elderly gentleman that does
not clearly fit within the standard classification
scheme. Based on existing literature, we think it is
best classified as ‘angio keras tomas with zosteriform
distribution.’
### Table 1. Angiokeratomas.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Clinical presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Angiokeratoma corporis diffusum (Fabry syndrome)</td>
<td>Before puberty</td>
<td>Predominantly in males but also occurs in females</td>
<td>Dermis, heart, kidneys, autonomic nervous system</td>
<td>Clusters of small, red papules</td>
</tr>
<tr>
<td>Angiokeratoma of Mibelli</td>
<td>Childhood to adolescence</td>
<td>Both sexes, but predominantly in young girls</td>
<td>Bony prominences of hands and feet</td>
<td>Hyperkeratotic vascular lesion(s)</td>
</tr>
<tr>
<td>Angiokeratoma of Fordyce</td>
<td>Adulthood or second to third decades</td>
<td>Male</td>
<td>Genitals</td>
<td>Dark red papules of 2 to 5 mm in diameter with a discrete keratotic surface</td>
</tr>
<tr>
<td>Angiokeratoma circumscripturn</td>
<td>At birth</td>
<td>Both sexes</td>
<td>Trunk and/or legs</td>
<td>Dark red to blue-black nodules or plaques presenting unilaterally</td>
</tr>
<tr>
<td>Solitary and multiple angiokeratomas</td>
<td>Second to fourth decades</td>
<td>Both sexes</td>
<td>Any part of body</td>
<td>Single or multiple papular lesion(s)</td>
</tr>
</tbody>
</table>

### Table 2. Grouped or agminated vascular lesions.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Pathology</th>
<th>Clinical presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Verrucous hemangioma [6]</td>
<td>At birth</td>
<td>Both sexes</td>
<td>Distal parts of lower limbs</td>
<td>Dilated capillaries and large endothelial-lined, blood-filled spaces extending well into the reticular dermis and subcutaneous tissue with an overlying hyperkeratotic epidermis</td>
<td>Dark blue papules or nodules</td>
</tr>
<tr>
<td>Tufted angioma [7]</td>
<td>First year of life</td>
<td>Both sexes</td>
<td>Predominantly, on the neck and trunk. Occasionally, on the extremities.</td>
<td>Lobules or tufts of endothelial cells in the dermis</td>
<td>Red to purple coalescent papules or plaques</td>
</tr>
<tr>
<td>Kaposiform hemangioendothelioma [8]</td>
<td>Children</td>
<td>Both sexes</td>
<td>No apparent site preference</td>
<td>Tightly packed spindle cells and small oval capillaries in cannonball or glomerular nests. The tumor grows as irregular slit-like vascular spaces that dissect between normal dermal and subcutaneous tissues.</td>
<td>Multinodular soft tissue masses, purpuric macules, plaques, and multiple telangiectatic papules</td>
</tr>
<tr>
<td>Spindle cell hemangioendothelioma [9]</td>
<td>Young adolescents</td>
<td>Both sexes</td>
<td>Typically, the distal extremity</td>
<td>cavernous blood vessels intermixed with solid areas composed predominantly of spindle cells.</td>
<td>Solitary or multiple nodules that have a smooth surface, are skin colored or blue, and firm in consistency</td>
</tr>
<tr>
<td>Multinucleate cell angiohistiocytoma [10]</td>
<td>Unknown</td>
<td>Females</td>
<td>Face and dorsal aspects of the hands</td>
<td>Vascular and histiocytic proliferations with dermal fibrosis</td>
<td>Erythematous to violaceous papules</td>
</tr>
</tbody>
</table>
Table 2, continued. Grouped or agminated/vascular lesions.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Age</th>
<th>Sex</th>
<th>Predilection</th>
<th>Associated Changes</th>
<th>Clinical Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eccrine Angiomatic Hamartoma [11]</td>
<td>Children &gt;</td>
<td>Both sexes,</td>
<td>Extremities&gt; head</td>
<td>Dilated eccrine glands associated with dilated capillaries</td>
<td>Variably sized patches plaques or nodules, red, blue, brown, or flesh-colored</td>
</tr>
<tr>
<td>Blaschko-linear</td>
<td>adults</td>
<td>male</td>
<td>neck and Trunk</td>
<td></td>
<td></td>
</tr>
<tr>
<td>“Congenital Mixed Hemato-lymphangiokeratoma</td>
<td>1 case</td>
<td>Female</td>
<td>Leg</td>
<td>Combined features of lymphangioma circumscriptum, angioma serpiginosa, and verrucous hemangioma</td>
<td>Combined red and yellow papules along lines of Blaschko</td>
</tr>
<tr>
<td>Serpiginosum” Naeviforme [12]</td>
<td>reported,</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>congenital</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Agminated Eruptive Pyogenic Granuloma [13]</td>
<td>Young</td>
<td>Both sexes</td>
<td>No specific</td>
<td>Lobular vascular</td>
<td>Grouped red papules</td>
</tr>
<tr>
<td></td>
<td>children</td>
<td></td>
<td>predilection</td>
<td>proliferation</td>
<td></td>
</tr>
<tr>
<td>Multiple agminated superficial arteriovenous</td>
<td>Adult</td>
<td>No known</td>
<td>Scalp</td>
<td>Aggregates of thin-walled and thick-walled vessels, lined with a single layer of</td>
<td>Clustered red papules</td>
</tr>
<tr>
<td>haemangioma[14]</td>
<td></td>
<td>predilection</td>
<td></td>
<td>plump endothelial cells.</td>
<td></td>
</tr>
<tr>
<td>Unilateral Agminated Angiofibromas [15]</td>
<td>Adult</td>
<td>No known</td>
<td>Face</td>
<td>Dermal proliferation of fibroblasts and capillaries</td>
<td>Unilateral discrete small red papules</td>
</tr>
<tr>
<td></td>
<td></td>
<td>predilection</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Multiple angiokeratomas in zosteriform</td>
<td>Adult</td>
<td>Both sexes</td>
<td>Buttocks and thighs</td>
<td>Thin-walled dilated vascular spaces in the papillary dermis surrounded by epithelial</td>
<td>Grouped small red papules in a unilateral and dermatomal distribution</td>
</tr>
<tr>
<td>distribution [4]</td>
<td></td>
<td></td>
<td></td>
<td>collarettes with variable hyperkeratosis and acanthosis</td>
<td></td>
</tr>
</tbody>
</table>

References

