With regard to treatment, different chemotherapy options—from CHOP to leukemia regimens—have been tested, but bone marrow transplantation is considered to be the best treatment. In a study of 57 patients, Dale et al. observed better survival in patients who had received transplants compared to other therapies (31.3 months vs 12.8 months). Preventive intrathecal chemotherapy has also been recommended.

Advanced age and the combination of negative TdT and positive BDCA-2 are negative prognostic factors. Of our 3 cases, only the patient who received a bone marrow transplant achieved remission lasting longer than 6 months. Thus, even though ours is a very small series, it supports the evidence for bone marrow transplantation as the best therapeutic option. Although TdT was positive in all our patients, this marker had no apparent bearing on survival. As has been reported in the literature, bone marrow involvement at the time of diagnosing the patients did not influence prognosis.

Our 3 cases reflect the typical presentation of plasmacytoid dendritic cell tumor, and suggest the importance of early treatment in order to improve what is normally a poor prognosis.

References


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Generalized Multinucleated Cell Angiohistiocytoma

Angiohistiocitoma de células multinucleadas generalizado

To the Editor:

Multinucleate cell angiohistiocytoma (MCA) is a benign fibrohistiocytic and vascular proliferation. This rare disorder was first described by Smith and Wilson Jones1 in 1985, and since then about 80 cases have been reported in the literature. The disorder occurs most frequently in women. Lesions are in the form of clusters of erythematous, violaceous, or brownish-colored papules located in a single anatomical area, usually the lower extremities, or the dorsal aspect of the hands, wrists, or fingers; however, it has also been described on the trunk, forehead, cheeks, oral mucosa, lips, and orbit. Generalized cases are exceptional. Although lesions are usually asymptomatic, some are pruritic.

Table 1 Immunohistochemical phenotypic profile of the patients.

<table>
<thead>
<tr>
<th>Positive markers</th>
<th>Negative markers</th>
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<tbody>
<tr>
<td>CD4</td>
<td>CD3</td>
</tr>
<tr>
<td>CD56</td>
<td>CD5</td>
</tr>
<tr>
<td>CD123</td>
<td>CD7</td>
</tr>
<tr>
<td>CD43</td>
<td>CD15</td>
</tr>
<tr>
<td>TdT</td>
<td>CD20</td>
</tr>
<tr>
<td>TCL-1 (Case 3)</td>
<td>Lysozyme</td>
</tr>
<tr>
<td>CD43</td>
<td>Myeloperoxidase</td>
</tr>
<tr>
<td>CD45RA</td>
<td></td>
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</tbody>
</table>

Abbreviations: TdT, terminal deoxynucleotidyl transferase; TCL-1, type 1 T-cell lymphoma.
A 56-year-old man, with no past history of relevance, presented with a slightly pruritic rash that had slowly developed over the previous 6 years. Examination revealed numerous sharply circumscribed erythematous, violaceous papules, some slightly brown in color, firm in consistency, and measuring 4 mm to 10 mm. The lesions were distributed on the trunk and the extremities, but particularly affected the flanks and lower part of the back (Figure 1). The patient reported no previous injuries, bites, or stings. Skin biopsy demonstrated the presence of a proliferation of capillaries in the dermis, with increased numbers of fibrohistiocytic cells and bizarre multinucleated cells with angulated cytoplasm (Figure 2). Immunohistochemistry was negative for CD34, S-100, CD68, cytokeratin, and epithelial membrane antigen, but showed increased numbers of capillaries positive for CD31.

The clinical manifestations and biopsy findings indicated a diagnosis of generalized MCA. The benign nature of the condition was explained to the patient who decided against treatment.

MCA is a dermal dendrocytic proliferation, similar to dermatofibroma, and, as with that condition, it is still not known if it is a genuine neoplasm or a reactive condition. Its benign course, its tendency to present as multiple lesions in the form of a rash in areas exposed to trauma and arthropod bites or stings, the absence of clonality, and cases of spontaneous regression would all support a reactive process.

From a histopathological point of view, MCA is characterized by a proliferation of capillaries and venules in the dermis, accompanied by an infiltrate of lymphohistiocytic cells and multinucleated cells with angulated or scalloped cytoplasm. The presence of mast cells in the dermal infiltrate has been described, but they were absent in the biopsy from our patient. Immunohistochemistry shows endothelial cells positive for the CD31 and CD34 antigens, and mononuclear interstitial cells expressing vimentin and factor Xlla, but not S-100 or CD1a. The characteristic multinucleated cells express vimentin, but positivity for CD68 is variable, and, in our case, was negative.

The pathogenesis of MCA is unknown. As for the histogenetic origin of the characteristic giant multinucleated cells, Wilson Jones et al suggested that they are derived from fibroblasts or macrophages, which, in conditions of prolonged chronic stimulation, lose their ability to divide after mitosis and so become inactive. Puig et al considered that the dermal angiogenesis and multinucleated cell formation processes were secondary to the release of cytokines by mononucleated fibrohistiocytic cells or mast cells present in the lesions.

The differential diagnosis of MCA (Table) primarily includes dermatofibroma. Requena et al consider MCA to be a variant of dermatofibroma with a prominent vascular component and bizarre multinucleated cells. The differential diagnosis should also include Kaposi sarcoma, which is distinguished from MCA both by microscopy findings and by negative immunohistochemistry findings for human herpesvirus type 8.

MCA presenting with a generalized distribution of lesions is rare, with only 3 cases reported in the literature. In 1996, Chang et al reported the case of a 24-year-old man with brownish-red papules affecting the trunk and extremities. In 1994, Del Rio et al reported the case of a 42-year-old man with skin lesions clinically and histologically compatible with generalized MCA, which developed in the context of POEMS syndrome. More recently, a case has been reported of a 40-year-old woman with lesions affecting the upper chest area, abdomen, back, and limbs. Lesion morphology and distribution in all the reported cases were consistent with those in our patient. Of the 4 patients (including our patient) diagnosed with generalized MCA, 3 were male; this contrasts with localized MCA, which predominates in women. Furthermore, although 1 patient developed generalized MCA in the context of POEMS syndrome, the other 3 cases of generalized MCA were not associated with other conditions; the finding was thus coincidental, although, admittedly, a larger series would be necessary to confirm this tentative conclusion.

<table>
<thead>
<tr>
<th>Clinical</th>
<th>Histological</th>
</tr>
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<tbody>
<tr>
<td>Dermatofibroma</td>
<td>Atrophic dermatofibroma</td>
</tr>
<tr>
<td>Angiofibroma</td>
<td>Angiofibroma</td>
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<tr>
<td>Kaposi sarcoma</td>
<td>Microvenular hemangioma</td>
</tr>
<tr>
<td>Pseudo-Kaposi sarcoma</td>
<td></td>
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<tr>
<td>Granuloma annulare</td>
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<tr>
<td>Lichen planus</td>
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<td>Pyogenic granuloma</td>
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<tr>
<td>Giant-cell fibroblastoma</td>
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</table>

Figure 1 Papular rash affecting the trunk, back, and buttocks.
The generalized variant of MCA is extremely rare, with only 3 cases recorded in the literature, to which we add a new case that is not associated with any other disease.

Conflict of Interest

The authors declare that they have no conflict of interest.

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References


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Simultaneous Presentation of Localized Basaloid Follicular Hamartoma and Epithelioid Blue Nevus in a 44-Year-Old Patient

Hamartoma folicular basalioide localizado y nevus azul epitelioide de presentación simultánea en una paciente de 44 años

To the Editor:

Basaloid follicular hamartoma is an uncommon benign neoplasm whose histological appearance is very similar to that of infundibulocystic basal cell carcinoma and trichoepithelioma. It may be familial or sporadic, generalized or localized, and is occasionally associated with autoimmune disease and cystic fibrosis. Some patients have a mutation in the PTCH gene that is similar to that observed in Gorlin syndrome and in up to 50% of isolated basal cell carcinomas.

Epithelioid blue nevus is a benign neoplasm whose histological diagnosis is complex due to its similarity with melanocytoma. It is closely associated with Carney complex.

We present the case of a 44-year-old woman with no relevant past history who consulted for a lesion on her back. The woman was unable to say how long she