To the Editor

In 1999, Carlson et al described 2 cases of a pigmented matrical neoplasm composed of matrical cells and dendritic melanocytes. The authors named this neoplasm, which was clearly distinct from pilomatricoma, melanocytic matricoma. This tumor mimics a normal anatomic process that takes place in the healthy bulb of an early anagen hair follicle.

We present a new case of melanocytic matricoma seen recently in our department. Only 10 such cases have been reported to date. The patient, a 66-year-old man with a history of hypertension, was referred to our department for evaluation of an asymptomatic lesion that had appeared on the bridge of his nose 1 year earlier. According to the patient, the lesion had appeared on normal skin and grown slowly. There was no family history of similar lesions. The patient had undergone cryosurgery in the past to treat facial actinic keratosis.

Physical examination revealed a blackish tumor with a diameter of 2 mm and clearly defined borders on the bridge of the nose. There was no evidence of any other skin lesions. Histopathology revealed a well circumscribed pigmented tumor in the middle and deep dermis (Figure 1). The tumor was composed of a biphasic cell population formed by melanocytes (several of which were heavily pigmented) with some mitotic activity, and epithelial cells of varying size and eosinophilic cytoplasm with abrupt transition to anucleated shadow cells (Figure 2). Also visible were small areas of calcification (Figure 3).

Immunohistochemical analysis revealed that epithelial components were positive for cytokeratin AE1/AE3 and melanocytic components for human melanoma black-45 (Figure 4).

Our findings are similar to those described in all the case reports of melanocytic matricoma published to date (Table). Clinically, melanocytic matricoma lesions are a blackish color and measure less than 1 cm in diameter; they occur in elderly patients (60-80 years), mostly men, with sun-damaged skin. There has been 1 report of melanocytic matricoma on the tail of a dog.

Histopathologic findings include pigmented nodular proliferation in the dermis composed of matrical cells, supramatrical cells, and shadow cells admixed with heavily pigmented dendritic melanocytes. Calcification and granulomatous reactions are uncommon. The small size of the lesions, their well circumscribed borders, and the lack of recurrence all suggest a benign neoplasm rather than a matricoma, despite the presence of variable cytologic atypia and frequent mitoses (characteristic of matrical cell tumors).
It is known that hair follicles in anagen (growth phase) contain matrical and supramatrical cells as well as pigmented melanocytes that give hair its color. Mitotic activity is also common. Because melanocytes are more prominent in the early anagen phase, melanocytic matricoma is suggestive of early-stage follicular differentiation during anagen; this contrasts with pilomatricoma, which is characterized by late-stage differentiation.

Clinical differential diagnosis should include pigmented basal cell carcinoma, malignant melanoma, and hemangioma. Histopathological differential diagnosis, in contrast, should include matrical carcinoma with prominent melanoacanthoma. Histopathological differential diagnosis, in contrast, should include matrical carcinoma with prominent melanocytic hyperplasia, a poorly defined, multinodular tumor that penetrates deeper layers and contains mitotically active cells and areas of necrosis. It has been suggested that matrical carcinoma with prominent melanocytic hyperplasia might in fact be a malignant form of melanocytic matricoma; this variant of carcinoma is a poorly defined, multinodular tumor that penetrates deeper layers and contains mitotically active cells and areas of necrosis. It has been suggested that matrical carcinoma with prominent melanocytic hyperplasia might in fact be a malignant form of melanocytic matricoma; this variant of carcinoma is a poorly defined, multinodular tumor that penetrates deeper layers and contains mitotically active cells and areas of necrosis.

Pilomatricoma occurs in young people as a cystic neoplasm, is firm to the touch, and is located in the deep dermis or subcutaneous tissues; it is often accompanied by calcification and granulomatous reactions. Pigmented pilomatricoma does not have prominent melanocytic hyperplasia, contrasting with the marked proliferation of pigmented dendritic melanocytes in melanocytic matricoma.

Ever since melanocytic matricoma was first described by Carlson et al, there has been some discussion about whether there is sufficient clinical and pathologic evidence to support the hypothesis that it is a separate entity from matricoma.

### References


### Table. Published Case Reports of Melanocytic Matricoma

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age, y</th>
<th>Sex</th>
<th>Dermatologic History</th>
<th>Diameter, cm</th>
<th>Color</th>
<th>Site of Lesion</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>66</td>
<td>M</td>
<td>Basal cell carcinoma</td>
<td>0.8</td>
<td>Black</td>
<td>Left chest region</td>
</tr>
<tr>
<td>2</td>
<td>80</td>
<td>M</td>
<td>Sun damage</td>
<td>0.5</td>
<td>Purple</td>
<td>Forearm</td>
</tr>
<tr>
<td>3</td>
<td>62</td>
<td>F</td>
<td>Sun damage</td>
<td>0.6</td>
<td>Hyperpigmented</td>
<td>Bridge of nose</td>
</tr>
<tr>
<td>4</td>
<td>78</td>
<td>M</td>
<td>Basal cell carcinoma</td>
<td>0.4</td>
<td>Black-purple</td>
<td>Left preauricular area</td>
</tr>
<tr>
<td>5</td>
<td>69</td>
<td>M</td>
<td>Sun damage</td>
<td>0.5</td>
<td>Black-brownish</td>
<td>Right cheek</td>
</tr>
<tr>
<td>6</td>
<td>66</td>
<td>F</td>
<td>Hyperpigmented</td>
<td></td>
<td></td>
<td>Right shoulder</td>
</tr>
<tr>
<td>7</td>
<td>70</td>
<td>M</td>
<td>Basal cell carcinoma</td>
<td>1.5</td>
<td>Black</td>
<td>Back of right hand</td>
</tr>
<tr>
<td>8</td>
<td>82</td>
<td>M</td>
<td>AK, SCC, and BCC</td>
<td>0.5</td>
<td>Black-purple</td>
<td>Right preauricular area</td>
</tr>
<tr>
<td>9</td>
<td>76</td>
<td>M</td>
<td>Brown-purple</td>
<td>2</td>
<td>Black</td>
<td>Top part of back</td>
</tr>
<tr>
<td>10</td>
<td>2</td>
<td>M</td>
<td></td>
<td>2</td>
<td>Black</td>
<td>Tail</td>
</tr>
<tr>
<td>11</td>
<td>66</td>
<td>M</td>
<td>Actinic keratosis</td>
<td>0.2</td>
<td>Black</td>
<td>Bridge of nose</td>
</tr>
</tbody>
</table>

Abbreviations: AK, actinic keratosis; BCC, basal cell carcinoma; F, female; M, male; and SCC, squamous cell carcinoma.

*Present case.*
LETTERS TO THE EDITOR

Bilateral Congenital Triangular Alopecia Associated With Congenital Heart Disease and Renal and Genital Abnormalities

E. León-Muiños, a B. Monteagudo, b J. Labandeira, c and M. Cabanillas a

aServicio de Pediatría and bServicio de Dermatología, Complejo Hospitalario Arquitecto Marcide-Nova Santos, Ferrol, La Coruña, Spain
cServicio de Dermatología, Complejo Hospitalario Universitario, Facultad de Medicina, Santiago de Compostela, La Coruña, Spain

To the Editor:

Congenital triangular alopecia, also known as temporal triangular alopecia or Brauer nevus, is a nonscarring circumscribed permanent and asymptomatic alopecia that was first described by Sabouraud in 1905. It is usually found on the frontotemporal area and affects only 1 side of the head.

Histopathology of the affected area reveals reduced hair follicle size, although hair density remains normal, with no other significant abnormalities. Diagnosis is usually clinical. Other causes of nonscarring circumscribed alopecia must be ruled out, especially alopecia areata, with which it is often confused. In the literature, there are reports of different conditions that coexist in patients with congenital triangular alopecia. We present the association between bilateral congenital triangular alopecia and a multiple malformation syndrome.

The patient was a 7-year-old boy with a history of congenital heart disease involving a perimembranous ventricular septal defect and an atrial septal defect with no hemodynamic consequences. He also had a history of left hydronephrosis, subcoronal hypospadias, Wormian bones, and recurrent bronchiolitis. The patient was referred because of the presence on the scalp of 2 areas with finer, lighter-colored hair, which his parents remembered as being there since birth. There had never been total hair loss in the area, and the patient

---

Figure 1. Oval alopecia plaque on the right temporal region.

Figure 2. Congenital triangular alopecia on the left frontotemporal region reaching the hairline.