SOCIEDADE BRASILEIRA DE DERMATOLOGIA

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CASE LETTER

Basaloid follicular hamartoma following Blaschko's lines*

Dear Editor,

A six-year-old female patient presented with hyperchromic confluent papules on the trunk and face (Fig. 1), following the embryological lines, with well-defined contours, elastic on palpation, not pruritic or painful. Hypopigmented lesions appeared at 15 days of life on the facial region, and evolved in the first year of life to hyperchromic lesions, spreading to the cervical region and trunk. The lesions have been stable and asymptomatic since that time. The mother denied comorbidities or associated symptoms. The patient does not have any family history of similar lesions, neoplasms, or autoimmune diseases.

On dermoscopy, the lesions were non-specific with a homogeneous brownish color and with structures similar to follicular crypts or openings (Fig. 2).

Histopathological examination (Fig. 3) showed a well-circunscribed, basaloid, epithelioid cell proliferation in the superficial dermis, forming strands and islets in a radial pattern. Thus, a diagnosis of multiple basaloid follicular hamartomas was made.

Basaloid follicular hamartoma (BFH) consists of the proliferation of multifocal basaloid cells, with frequent connection to the epidermis.[1] BFH lesions may present as papules, nodules, or plaques, which may be skin-colored or hyperchromic. The cells are folliculocentric and restricted to the superficial dermis. The hair follicles are distorted, with branching strands from basaloid cells.[2,3] The present case showed a distribution of basaloid cells in strands and islets with a radial pattern in the superficial dermis, which clinically followed the embryological lines.

The main differential diagnosis of BFH is basal cell carcinoma (BCC). Both consist histopathologically of basaloid strands of cells in a fibrous stroma, but the BCC is not folliculocentric and can be seen in the interfollicular dermis.[1,3]

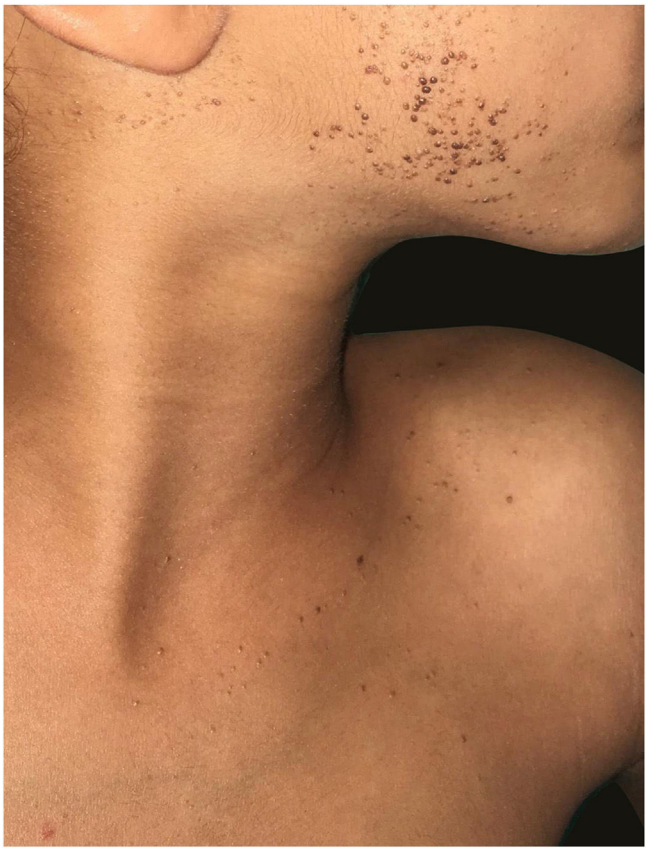

Figure 1 Hyperpigmented unilateral linear papular lesions; some are verrucous, on the face and neck.

Acquired BFH can show a linear pattern, following the lines of Blaschko - occurring due to mosaicism - or in a generalized form - commonly associated with autoimmune diseases.[3]

In the case described herein, BFH clones were distributed along the Blaschko lines, representing ectodermal development patterns, which is a rare distribution. When a somatic mutation or chromosomal nondisjunction occurs during embryogenesis, affecting an epidermal progenitor cell, the affected offspring cells proliferate and migrate along the lines of Blaschko.[4]

Currently, there is no standard treatment for BFH. Correct identification prevents patients from undergoing

* Study conducted at the Hospital Infantil Varela Santiago, Natal, RN, Brazil.

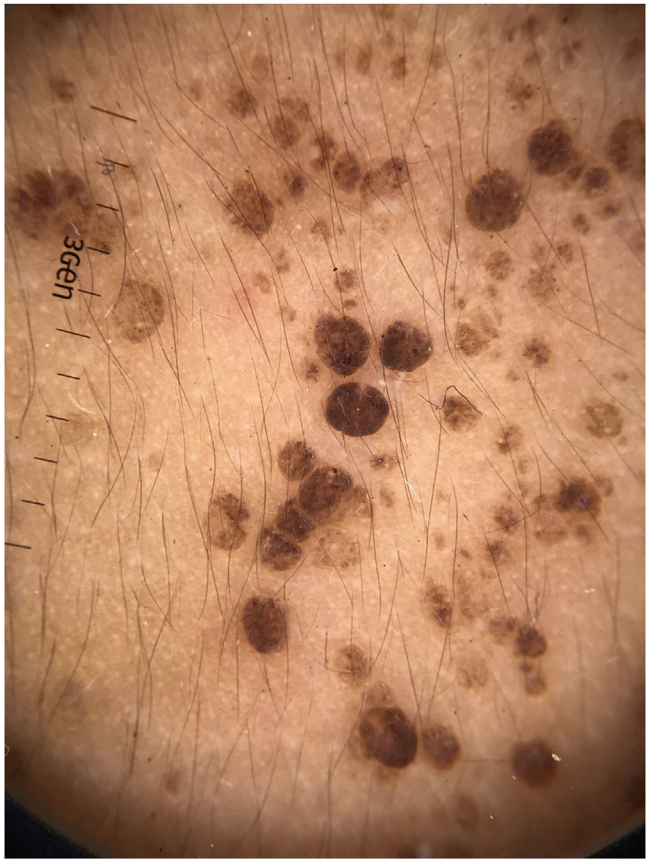

Figure 2 Dermoscopy showing brownish, crypt-like structures.

unnecessary surgery and also allows periodic monitoring to detect malignant transformations. Lesions that increase in size or change in appearance should be biopsied whenever detected. If associated with an autoimmune disease, treatment of the comorbidity may lead to the regression of the associated skin lesions.[3]

In summary, BFH is a rare type of benign skin tumor, with different presentations, which can be congenital or acquired. Its main differential diagnosis is basal cell carcinoma, and histopathology should be performed for differentiation. There is yet no standard treatment for this condition and, in most cases, it is not necessary.

Financial support

None declared.

Authors' contributions

Gabriela Martins de Queiroz: Collection, analysis, and interpretation of data; design, planning, drafting and writing of the manuscript; critical review of the literature; critical review of the manuscript; approval of the final version of the manuscript.

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Maria Clara Dantas Valle Soares: Analysis and interpretation of data; drafting and writing of the manuscript; critical review of the literature; critical review of the manuscript; approval of the final version of the manuscript.

Carlos Bruno Fernandes Lima: Analysis and interpretation of data, design and planning of the manuscript; critical review of the literature; critical review of the manuscript; approval of the final version of the manuscript.

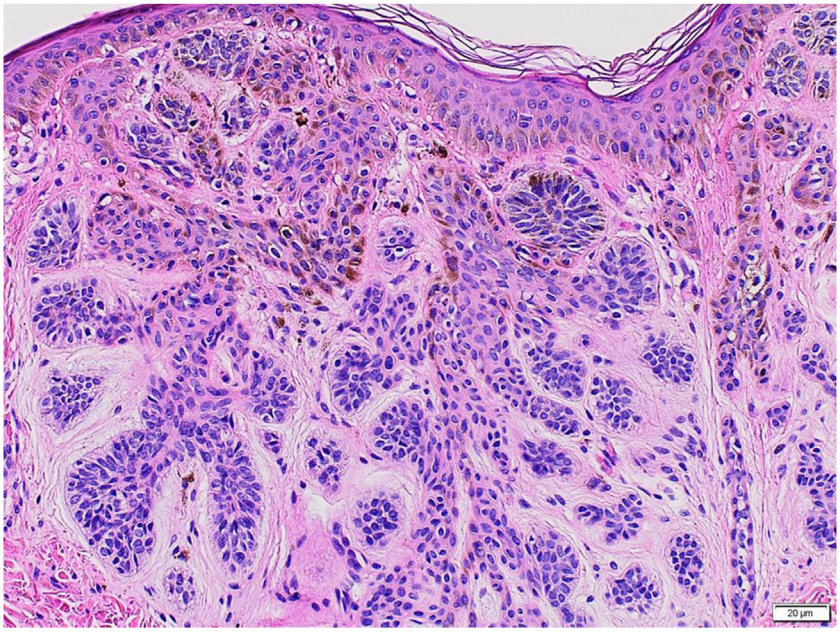

Figure 3 Proliferation of basaloid epithelioid cells in strands and islets showing a radial pattern (Hematoxylin & eosin, ×200).

Conflicts of interest

None declared.

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Received 28 August 2020; accepted 23 October 2020
Available online 21 September 2022

https://doi.org/10.1016/j.abd.2020.10.027

Congenital infantile fibrosarcoma: a rare tumor dermatologists should know about☆

Dear Editor,

A seven-month-old female patient presented with a history of a congenital, violaceous, fast-growing lesion located on the right plantar surface. Dermatological examination disclosed the presence of a firm spherical tumor, with dilated vessels on the surface, and central ulceration with friable, bleeding tissue, and hematic crusts (Fig. 1A). The child developed severe anemia (hemoglobin of 4.4 g/dL), requiring a blood transfusion. The platelet count was normal. Histopathology was suggestive of kaposiform hemangioendothelioma. Treatment with oral prednisolone (2 mg/kg/day) was started but was interrupted after one month, due to lack of a response (Fig. 1B).

Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) disclosed a well-vascularized solid mass, with the involvement of the underlying muscles and extending to the anterior aspect of the foot. Diffuse contrast enhancement was observed throughout the lesion, with no signs of arteriovenous shunts or a cluster of tortuous vessels (nidus), thus ruling out the diagnosis of a vascular tumor, including kaposiform hemangioendothelioma (Figs. 2A and 2B). A second biopsy was performed, revealing a hypercellular fusiform tumor. Immunohistochemistry was positive for vimentin and negative for CD31, CD34, factor VIII, desmin, MyoD1, myogenin, CD99 and EMA, indicating the diagnosis of congenital infantile fibrosarcoma (CIF).

The patient was submitted to neoadjuvant chemotherapy (vincristine, actinomycin-D and cyclophosphamide) to reduce tumor size (Fig. 1C), followed by amputation of the foot. There are no signs of recurrence or metastasis at five years of follow-up.

☆ Study conducted at the Hospital das Clínicas, Universidade Federal de Minas Gerais, Belo Horizonte, MG, Brazil.

CIF is a rare malignant tumor of childhood; however, it is the most common soft tissue sarcoma in children under one year of age.[1] This highly vascularized congenital tumor is difficult to clinically differentiate from vascular tumors or malformations. It may be present at birth or develop during the first five years, with approximately 80% of cases diagnosed during the first year of life.[2]

Fibrosarcomas are malignant neoplasias composed of mesenchymal fibroblasts. The infantile variant shares histopathological characteristics with adult fibrosarcoma but has a better prognosis. Although local recurrences are common, the rate of CIF metastasis is less than 10% and the ten-year survival rate is up to 90%.[3] The extremities are more commonly affected and lesions located on the trunk, head and neck are less frequent, although they are more aggressive.[1,4] Due to the risk of local recurrence, extensive surgical resection is recommended. Surgery alone shows recurrence rates of 17% to 40%. Neoadjuvant chemotherapy reduces the risk of local recurrence and metastases.[2,3,5]

The histopathological findings of CIF include the proliferation of dense fusiform cells and vascularized areas. Immunohistochemistry is positive for vimentin and, in some cases, for desmin, smooth muscle actin, and cytokeratin.[4] CIF is characterized in up to 85% of cases by a specific t(12;15) (p13:q25) chromosomal translocation encoding an ETV6-NTRK3 gene fusion.[1,3–5]

The diagnosis of CIF should always be considered in the presence of a congenital, spherical, bleeding extremity tumor in children, aiming to avoid treatment delays.

Financial support

None declared.

Authors' contributions

Luciana Baptista Pereira: Design and planning of the study; drafting and editing of the manuscript; collection, analysis, and interpretation of data; critical review of the manuscript; approval of the final version of the manuscript.