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PHOTOQUIZ

A firm purple nodule on a child's abdomen

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CASE PRESENTATION

A 9-year-old girl presented with a 5-month history of a painless nodule on her left lower abdomen. It was initially described as "like a nipple," but then became larger and darker. It had bled once following minor trauma. There were no systemic symptoms and no past medical history.

On examination, there was a 15 × 5 mm firm dark purple nodule on the left lower abdomen (Figure 1). Whitish streaks were visible within the middle of the nodule on dermoscopy. The rest of the cutaneous examination was normal and there was no lymphadenopathy. Full blood count, coagulation screen, renal function, and liver function were normal. An incisional biopsy (Figures 2 and 3) was performed.

WHAT IS YOUR DIAGNOSIS?

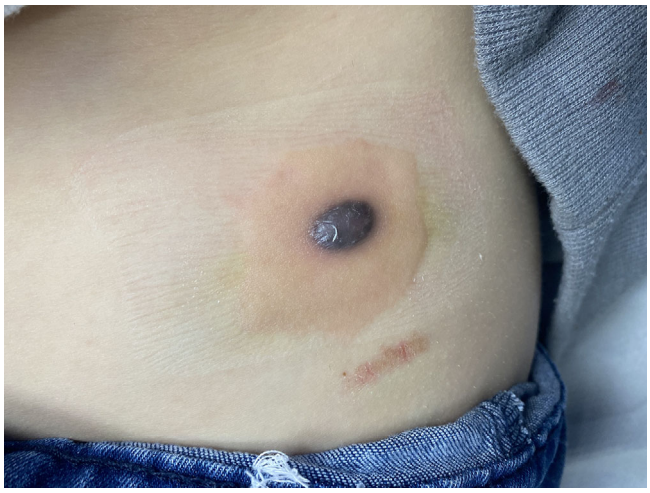

FIGURE 1

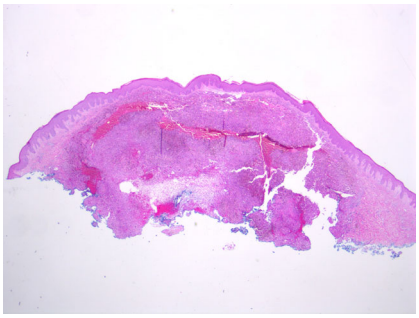

FIGURE 2

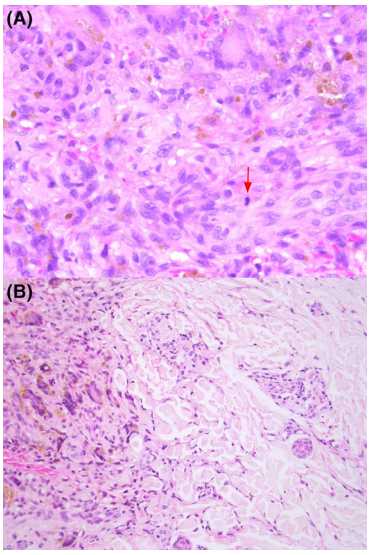

FIGURE 3

Diagnosis: Aneurysmal fibrous histiocytoma (aneurysmal dermatofibroma)

Histological sections revealed a dermal lesion with a Grenz zone separating it from the overlying epidermis. The lesion was composed of bland spindle cells and a histiocytic proliferation with entrapment of dermal collagen and pseudovascular spaces. There were prominent multinucleated giant cells scattered throughout the lesion together with hemosiderin laden macrophages, and occasional mitoses (arrow, Figure 3).

Magnetic resonance imaging showed a residual 12.5 cm × 5 cm area of altered signal intensity, on T1- and T2-weighted imaging in the skin and the subcutaneous fat of the left anterolateral abdominal wall, adherent to the fascia of the left external oblique muscle. The lesion enhanced following gadolinium administration. No lymphadenopathy was present.

The patient was referred to our plastic surgery colleagues for excision of the lesion. The histology of the excised lesion was consistent with aneurysmal fibrous histiocytoma (AFH). After 2 years of follow up there has been no recurrence.

DISCUSSION

AFH is a rare variant of the common cutaneous soft tissue lesions, dermatofibromas, representing less than 2% of total cases.[1] AFH are mesenchymal tumors that present as blue, red or purple papules and are often found on the lower extremities.[1] Accelerated enlargement of AFH due to hemorrhage in a chronic lesion is frequently seen. They are usually larger than typical dermatofibromas, and significant subcutaneous extension can be seen, as in this case.[1] Multiple AFH in one patient has been reported.[2]

AFH usually develops in middle-aged adults (median age at diagnosis 37 years), with a slight female preponderance.[1] All types of dermatofibromas are more commonly seen in adults and are rare in young children, although the aneurysmal variant may be underrecognized in children.[3]

Histologically, AFH is usually dermal but can extend into superficial adipose tissue, or even to the subcutis and skeletal muscle. Epidermal acanthosis may be present. Blood-filled spaces are commonly seen, lined by histiocytes, siderophages, foam cells, fibroblasts, and occasionally giant cells. The surrounding stroma contains abundant capillaries with interstitial hemorrhage and hemosiderin deposition. A focal storiform arrangement of fibroblasts and histiocytes may be seen. Focal positivity for smooth muscle actin and Factor XIIIA may be seen in lesional cells, which are usually negative for desmin, S100, CD34, and CD31.

Important histological differentials for AFH include angiomatoid fibrous histiocytoma, angiosarcoma, Kaposi sarcoma, spindle cell hemangioma, and malignant melanoma.[1] The distinction from malignant or borderline tumors can be challenging because of histological features such as architectural distortion due to extensive hemorrhage and frequent mitosis. In particular, angiomatoid fibrous histiocytoma, a distinct entity of intermediate biologic potential, can be seen in children. However, these are located exclusively in the subcutaneous tissue and lower dermis, in contrast to AFH which is entirely or predominantly located in the dermis. In addition, a thick tumor capsule and lymphoid aggregates are typical of angiomatoid fibrous histiocytoma, but are not seen in AFH. Immunohistochemistry can be helpful in differentiating AFH from other diagnoses. In this case, S100 (a melanocytic marker) and CD31 (a vascular marker) were negative in tumoral cells, thus excluding melanoma (S100 positive), angiosarcoma, Kaposi sarcoma, and spindle cell hemangioma (CD31 positive).

Given the rapid changes that are frequently reported in AFH, the clinical differential diagnosis for AFH is similar to the histological differential, including melanoma, nodular Kaposi's sarcoma, and angiosarcoma. The most frequent dermoscopic features of AFH include a central bluish or reddish homogenous area with white structures and a peripheral delicate pigment network with vascular structures of variable degrees. Dermoscopy cannot definitely rule out melanoma, as the pigment network, homogenous areas, regression structures (white areas), and atypical vascular structures are frequent in melanoma. Nodular Kaposi's sarcoma typically presents simultaneously with patch and plaque stages. Angiosarcoma is usually seen on the face or scalp of older adults. Histopathological assessment is recommended in all cases to differentiate between these entities.[4]

The recurrence rate for AFH post excision may be up to 20%,[5] much higher than the typical form of dermatofibroma (<2%). The high rate of recurrence is likely due to incomplete excision of large primary tumors, rather than a biological difference.[5,6] Although a benign entity, AFH can metastasize in exceptional cases (<1%),[7] most commonly to lungs and lymph nodes.[5] Therefore, complete surgical excision is recommended. Moh's surgery has been recommended given its precise margin control,[8] although this modality may be limited in the pediatric population due to the need for strict patient cooperation.

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