

A skin tumor in a young girl

A nine-year-old girl attended our clinic with a slowly-enlarging nodule on the left leg. The lesion had arisen one year ago, gradually enlarged and ulcerated from two different areas. She did not report trauma to the location of the lesion previously. On dermatological examination, a 3.5 × 2 cm, firm, non-tender, erythematous tumoral lesion was seen on the left leg. Its surface was smooth and two ulcerated areas were observed [Figure 1]. There was no lymphadenopathy in the left inguinal region. The patient was otherwise in good health and routine laboratory tests were normal. The tumor was totally excised. Histopathological examination revealed a circumscribed tumor centered in the dermis connected with the epidermis. The tumor was composed of clear and polyhedral cells. Focally, there were cystic spaces and a few duct-like structures [Figures 2a, b and 3]. Histochemically, the cytoplasm of clear cells and the lumen of duct-like structures showed PAS-positive, d-PAS-resistant staining.



Figure 1: Erythematous tumoral lesion and fistulated areas were seen on the left leg

WHAT IS YOUR DIAGNOSIS?

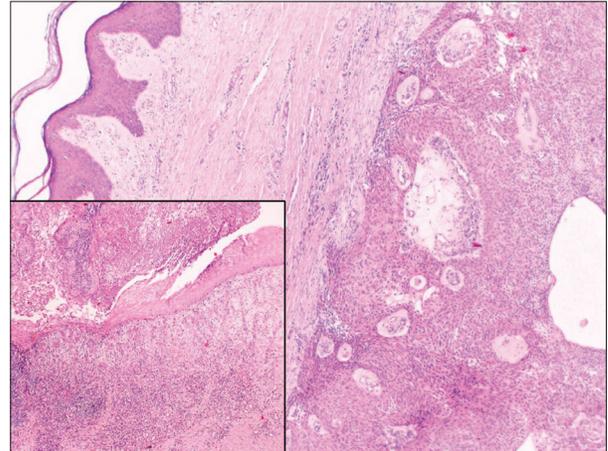


Figure 2: (a) Tumor cells, focal cystic changes and a few duct-like structures are seen (H and E ×40). (b) (small figure) The connection with the tumor's cells and surface of the epidermis (H and E, ×40)

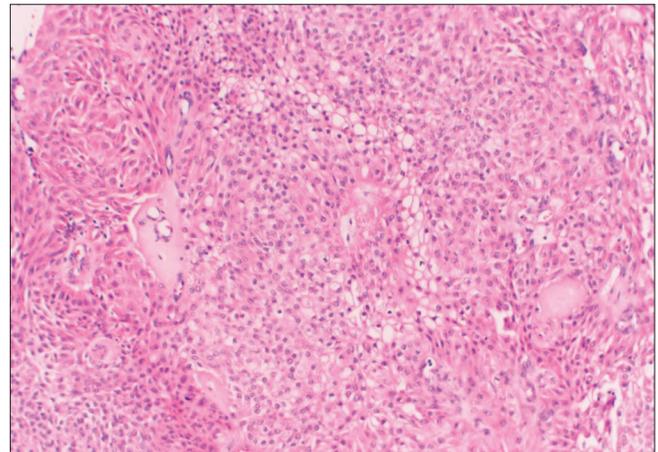


Figure 3: The tumor is comprised of clear and polyhedral cells, some of which appear fusiform (H and E, ×100)

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DIAGNOSIS

Clear cell hidradenoma.

DISCUSSION

Hidradenoma is an asymptomatic, uncommon benign tumor that grows slowly. It presents more often on the upper body. It usually appears between the fourth and the eighth decade of life, with a peak incidence in the sixth decade.^[1,2]

Its origin is uncertain. Most of the authors consider these tumors to be of eccrine origin, but there are some reports supporting an apocrine derivation. Clear cell hidradenoma is the most frequent type of hidradenoma.^[3] Clear cell hidradenoma is referred by various terms, such as nodular hidradenoma, eccrine acrospiroma, solid-cystic hidradenoma, clear cell acrospiroma, clear cell myoepitelioma, and eccrine sweat gland adenoma.^[1,4]

Clear cell hidradenoma is usually an asymptomatic, benign, intradermal, rare skin tumor that grows slowly. These tumors are variable in size with a small tendency to ulcerate and have a low malignant potential. They usually present as solitary, skin-colored or red lesions. Women are affected more often than men. The lesions are seen most frequently on the scalp, face, thorax and abdomen.^[5]

Differential diagnosis includes hemangioma, glomus tumor, cutaneous lymphoma, dermatofibrosarcoma protuberans, leiomyoma, follicular cyst, tricholemmoma and sweat gland tumor.^[1,2] Histopathological examination of the lesion provides diagnosis of clear cell hidradenoma. Histopathologic examination shows encapsulated and well-circumscribed tumor, variably-sized nests and nodules of neoplastic epithelial cells, with small ductular lumens, confined to the upper dermis. Tumor cells are small, monomorphous and polyhedral. Clear cell change and/or squamous metaplasia may be prominent. Focal apocrine components may also be present.^[4]

Clear cell hidradenomas should be surgically removed because they have a high local recurrence rate. Safety margins should be considered during resection because of the potential of malignant transformation.^[2,5]

Faulhaber *et al.*, reported a case of clear cell hidradenoma in a girl in the first decade of life.^[2] After that, few cases of clear cell hidradenoma appearing in the first decade have been reported.^[1,2,5] In those cases, localizations of the lesions were chest, shoulder and upper extremities. Our case is the first in the literature of a child patient who had a hidradenoma lesion on a lower extremity. Although the exact number of patients with clear cell hidradenoma is not known because of the different designations under which this tumor has been reported, there are few patients in literature reported as clear cell hidradenoma. It is important to consider clear cell hidradenoma as a rare differential diagnosis of cutaneous tumors even in young children, since in some cases a malignant transformation has been observed.

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