Case report

Clear cells hidradenoma in a 12-year-old girl

Abstract:

Hidradenoma is an uncommon skin tumor, representing 1% of all skin tumors, and remains exceptional in children.

We report a case of a 12-year-old female patient with a mass on the anterior side of the right forearm, evolving for 2 years. The radiological evaluation did not lead to a diagnosis.

A surgical biopsy was performed, the result was a clear cell hidradenoma; therefore, an excision with a safety margin of 1 cm was performed, with a good evolution over a 12-month period.

Through this clinical observation, the authors propose a review of the literature

Key words: hidradenoma, clear cells, child, female.

Introduction:

Hidradenoma is an uncommon skin tumor, representing 1% of all skin tumors [1-2]. Clear cell hidradenoma is a rare adnexal tumor, exceptional in children, of eccrine origin, locating mainly on the trunk. Its prognosis is conditioned by the risk of frequent local and distant recurrence. We report a new observation of a hidradenoma of exceptional location on the forearm.

Case Presentation:

A 12-year-old girl, presented 2 years ago with a tumefaction of the anterior side of the middle third of the right forearm, the evolution was marked by ulceration with abundant bleeding on contact and rapid increase in size.

Examination revealed an ulcerated mass on the anterior side of the middle third of the right forearm, bleeding on contact, measuring 4*3cm (figure 1), with no other associated lesions, notably no axillary adenopathy. Biopsy was in favour of a clear cell hidradenoma.

Surgical resection was performed with a safety margin of 1 cm, removing the dermis, hypodermis and superficial fascia (Figures 2-3).

The immunohistochemical study confirmed a benign clear cell hidradenoma.

The evolution was good with a follow-up of 06 years, without local or distant recurrence.

Discussion:

Mayer was the first to describe the hidradenoma as a distinct clinical entity [3], it is a benign adnexal neoplasm of uncertain origin. The eccrine origin is the most likely. However, some studies point to an apocrine derivation. Recently, hidradenoma has been classified into two groups; those with eccrine differentiation (poroid hidradenoma) and those with apocrine differentiation (clear cell hidradenoma) [3]. Clear cell hidradenomas are by far the most common subtype and represent 95% of all cases [4-5], and 1% of primary skin tumors. The sex ratio of man to women is 1 to 2 [1-2].

The fact that oestrogen receptors are sometimes found in benign nodular hidradenomas and the female predominance suggest a possible role for these hormones in the development of these tumors [4]. The average age is between 30 and 40 years, and children are exceptionally affected. No risk factors have been identified [4].

The hidradenoma usually presents as a solitary, asymptomatic, well-circumscribed, slowly growing mass composed of lobulated, cystic or pedunculated lesions that reside in the upper or middle dermis but sometimes extend deeply into the subcutaneous tissue. They range in size from 0.5 to 3 cm and are often found on the head, face and upper limbs [6-7-8]. The color of skin above is variable.

The lesion may ulcerate, producing a clear or haemorrhagic fluid [2], as in our case where negligence had led to a 4 cm mass which subsequently ulcerated. In two of the larger series [3-9]; the hidradenoma was located in the trunk in more than half of the cases, and in the cervico-facial region in 25%. [9]
Indeed, clear cell hidradenomas rarely occur in the limbs. The most common tumors found in the limbs are fibromatosis, giant cell tumors of the tendon sheaths and lipomas [3].

The histopathological diagnosis is difficult, due to the varied clinical signs and the variable prevalence of the different components: solid, cystic, tubular lumen, clear cells and spindle cells, explaining the many histological varieties. The clear cells form remains the most common. There are several variants of clear cell hidradenoma, including the benign form, the locally aggressive tumor with tissue invasion and the malignant tumor with metastatic potential, which is mainly lymphophilic and remains rare [4].

The distinction between benign and malignant hidradenoma is quite difficult, hence the interest of immunohistochemistry. MKi67 is used to define the aggressivity of the tumor by percentage. The epithelial nature and adnexal differentiation are confirmed by other markers (keratin / EMA / CEA / S100 protein / Vimentin) [5].

Because of the high recurrence rate, surgical resection should be complete with wide safety margins, as malignant transformation may be present in other areas of the lesion [9]. In addition, hidradenoma has a recurrence rate of approximately 12% if not completely excised [10], especially in lesions with irregular peripheral margins [11].
Clinical surveillance should be regular, justified by the hypothesis of a possible benign extension to the lymph nodes.
Although most cases of hidradenocarcinoma occur de novo, it can also appear in pre-existing hidradenoma, and this potential for transformation requires clear surgical margins [12-13-14] for which there is no clear consensus.

Conclusion:

The hidradenoma is an uncommon adnexal tumor, without being extremely frequent. Immunohistochemistry remains the only examination that can differentiate benign from malignant forms due to the clinical and pathological difficulties. Surgical resection is the rule because of the risk of malignant transformation.

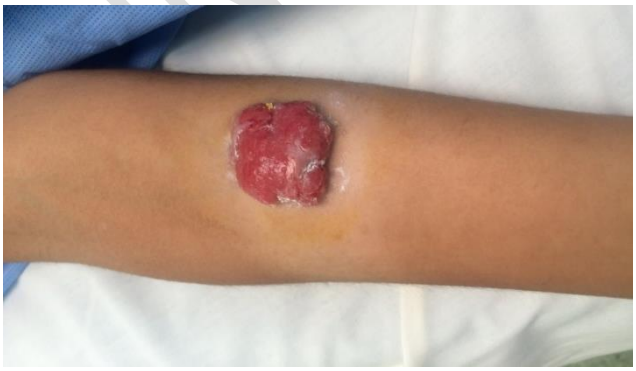

Figure1: clinical appearance of the tumor

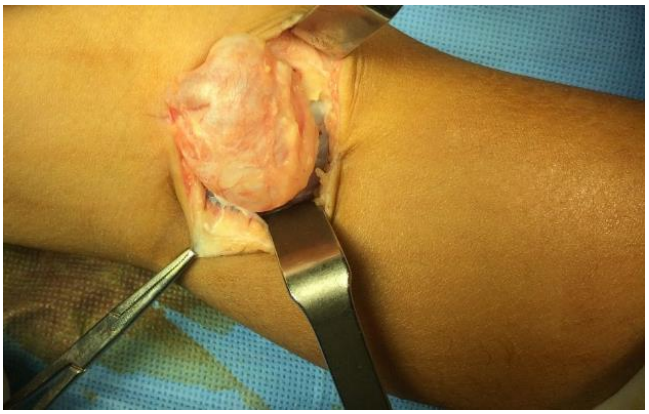

Figure 2: intraoperative appearance

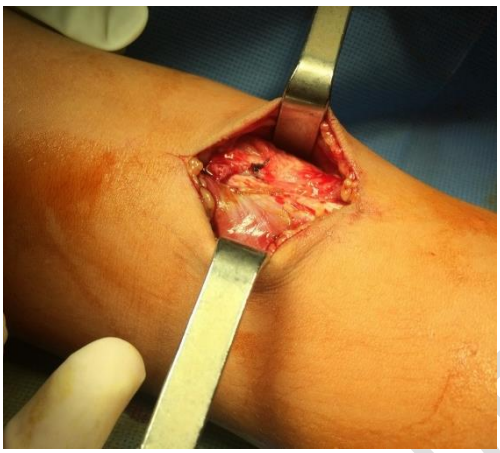

Figure 3: appearance after resection of the tumor removing dermis, hypodermis and superficial aponeurosis

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