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Case Report

Scrotal Kerathoacanthoma : Exceptionnal Case

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Abstract

Keratoacanthoma (KA) is a common butunder reported skin tumor developed from pilosebaseous glands. Its main clinical feature is a spontaneous regression after rapid growth. Some authors put KA on the border between benignity and malignity because of a rare but possible transformation into a real squamous cell carcinoma (SCC), in addition to similar pathological features between the two entities. KA is clinically a 1 to 2 cm nodule with a central crater filled with horny material located on sun exposed skin. The diagnosis of KA is set on a clinicopathological confrontation. The treatment is based on surgical excision of the lesion to prevent a malignant transformation. The prognosisis excellent with extremely rare recurrence cases.

Here in we report an exceptionnal case of giant scrotal keratoacanthoma in a 62yearsold patient. The diagnosis was based on clinical and pathological exams. Surgical excision was performed and the postoperative course was uncomplicated. Through this clinical case and a literature review, we analyze the ethiopathogenesis, diagnosis and therapeutic care of this under reported pathology.

Introduction

Keratoacanthoma (KA) is abenign skin tumor developped from pilosebaceous glands and it is pathologially similar to squamous cell carcinoma (SCC). It is generally located on sun exposed skin, and scrotum remains an exceptional location. KA is characterized by rapid growth over a few weeks to months, followed by spontaneous resolution over 4-6 months. But in some rare cases KA can reach important size before involution which leads to giant KA. Moreover, it reportedly progresses, although rarely, to invasive or metastatic carcinoma; therefore, aggressive surgical treatment is often advocated.

We report a case of giant keratoacanthoma of the srotum in a 62 years old man in view of itsrarity. From this observation, we present the clinical and pathological aspects and report our therapeuticstrategy.

Observation

A 62-years-old man with a history of cholecystectomy in 2005, presented with a tumor on the left side of his scrotum, which had been evolving for almost 9 months. This lesion had fastly grown during the last months and caused discomfort. Physical examination revealed a rounded, roughly symmetrical vegetative tumor measuring 3,5 cm in diameter, not-adhering to deep structures, painless, and without any signs of infection or haemorrhage or necrosis (Figure 1). Moreover, the tumor had central crater with whitish punctate deposits. Total surgical excision of the lesion was performed under spinal anesthesia (Figure 2).We took care to keep 5mm of margin of safety to avoid any subsequent recidivism. Macroscopically, the tumor were non encapsulated, with whitish fibrous tracts alternating with yellowish tissue of fatty consistency (Figure 3). The histological study revealed a benignproliferation composed of collagen and elastic fibers, associated with some fibroblasts and fat cells. The elastic fibers were fragmented into globules or had a linear arrangement (Figure 4). Surgical margins were negative was without complications, and the patient was discharged 1days after surgery.

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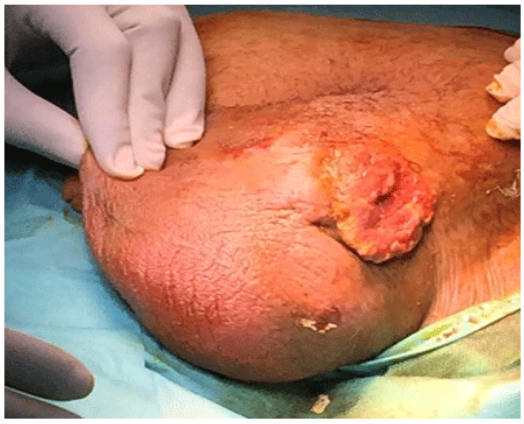

Figure 1. Vegetative tumor measuring 3,5 cm in diameter

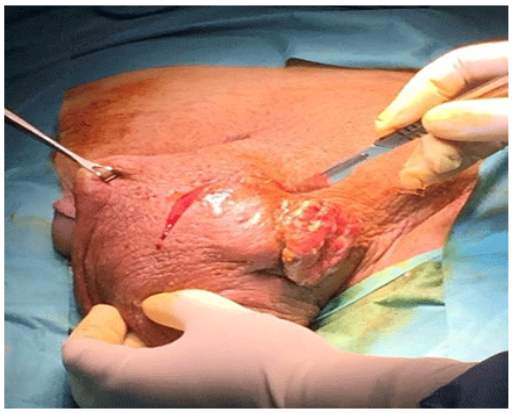

Figure 2. Total surgical excision of the lesion

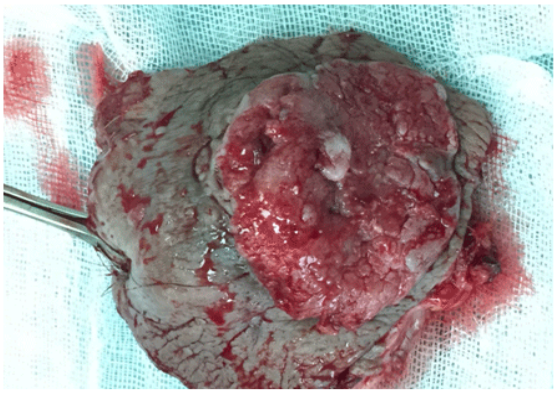

Figure 3. Macroscopic aspect of the specimen (giant KA).

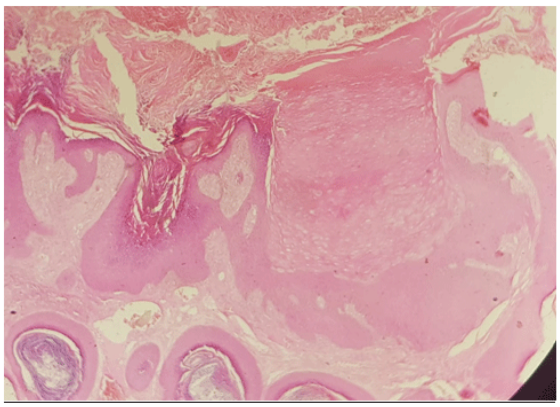

Figure 4. Microscopic aspect of Kerathoacanthoma

Discussion

First described in 1889 by Hutchinson [1], KA is also called « molluscum sebaceum », « regressing tumor » or « self healing squamous cell carcinoma » [2,3]. It is an epithelial benign tumor developped from the pilosebaceous follicle which is characterized by a sudden onset and rapid enlargement then a spontaneous regression[4,5]. Its histological patterns are often suggestive of a squamous cell carcinoma (SCC). Therefore, all the interest lies in the distinction with the highly malignant de novo type of SCC which has a high metastatic potential [6].

KA affects most likely men between 60 and 70years old especially in sun-exposed areas [7]. Its true incidence is underestimated probably because of misdiagnosis as a SCC, or because of a spontaneous regression before the diagnosis. Moreover, the pathogenesis of KA remains insolved and is still open to controversy. Many factors seem to promote its occurrence.

On the one hand, multiple etiologies have been suggested including ultraviolet radiation, exposure to chemical carcinogens, immunosuppression, use of BRAF inhibitors, geneticpredispositionincludingp53 or H-Ras mutations, viral exposure including human papillomavirus (HPV), and recent trauma (includingTatoo) or surgery to the location. On the other hand, KA can be commonly associated with syndromes such as autosomal dominant Muir-Torre syndrome, autosomal dominant Ferguson-Smith syndrome, autosomal recessivexerodermapigmentosum, X-linked dominant in continentiapigmenti,autosomal dominant Witten-Zak [8, 9, 10,11, 12].

KA is most often a solitary lesion with a clinical history of rapid growth over 4 to 5 weeks, with resolution after 6 months [13]. It is typically located on face and arms because of sun exposure. Nevertheless, the rare solitary KA located on the mucous membranes (buccal, conjunctival and vulva) challenges this theory [14, 15].

It is clinically presented as an erythematous-squamous papule that often goes unnoticed in the initial stage; then a symmetrical well-

defined nodule is formed in a few days. It is usually a 1 to 2cm nodule. Giant KA is a rare variant of KA with a size exceeding 2 cm [16,17]. Our case is even more exceptional not only by its size but also by its location due to the occurrence of keratoacanthoma on the scrotum in a patient who does not practice nudism.

The nodule is composed by a peripheral bead covered with a stretched shiny and telangiectatic epidermis that circumscribes a central crater filled with horny material. Then, keratoacanthoma regresses spontaneously by subsidence of the peripheral bead and elimination of the central horny plug, leaving an atrophic scar often traversed by grains of milium. Extensive ulcerative progression is possible, especially in the nose and eyelids. Metastasis have not been reported, which distinguish KA from the SCC [18,19].

XerodermaPigmentosum and nevussebaceous of Jadassohn) [20].

Dermoscopy of KA shares some features in common with SCC and cannot distinguishes between the two [21,22]. Typically, dermoscopy of KA appears as a central keratin plug surrounded by vessels in crown, with haemorrhages [23].

With regard to anatomopathological diagnosis, the experts recommend focusing more on the global architecture of the lesion than its cytological characteristics. Thus it is therefore essential to perform a biopsy of the entire lesion comprising both the central crater but also the peripheral bead. Around the central crater filled with keratin, the peripheral pearl representing the proliferation of keratinocytes completes the typical image of KA. However, some eosinophilic or neutrophilic epithelial microabscesses have been reported in the literature. The main obsession of the pathologist remains the ignorance of asquamous cell carcinoma. However, unlike SCC, KA is characterized by a symmetrical general organization and an abrupt transition between the tumor mass (connection in bill or spur) and the normal epidermis. Moreover, in almost all cases, the tumor does not extend into the dermis until the eccrine glands [4]. Finally it is important to remember that there is no dysplasia and that the mitotic index is low.

We attempted to collegiate in a table the main characteristics of each of the two entities (KA and SCC) in order to better differentiate them (table 1).

Table 1: The main characteristics of each of KA and SCC

Clinicalfeatures	Keratoacanthoma	WelldifferentiatedSquamouscellcarcinoma of the skin
Anamnesis	-Appearancein lessthan10 weeks	-Progressive appearance
Clinical aspect	-Evolution in 4 phases (proliferation, maturation, involution, healing) -Important size (10-20 mm) -Healthyperilesional skin	-Slow and continousevolution -Variable size -Oftenmodifiedperilesional skin
Evolution	-Benign	-Local destruction, recurrences, metastasis
Histologicalfeatures		
General architecture	-Symmetrical exophytic tumor -Predominent orthokeratosis -Transitions in beak between the tumor and the normal skin -Well delimited tumor -No rupture of the basement membrane -No necrosis -Rare overflow of eccrine glands -Rare vascular emboli or perineural invasion	-Exo / endophytic tumor -Predominant parakeratosis -Frayed tumor limits -Rupture of the basement membrane -Frequent necrosis -Frequent overflow of eccrine glands -Vascular emboli and frequent perineural invasion
Cytology	-Possible atypias and mitosis -Glassy appearance of keratinocytes -Micro abscess with neutrophils	-Numerousatypias andmitosis -Variable keratinocytes aspect -Unfrequent micro abscesswithneutrophils

Moreover, as KA is biologically unstable, some KA can be transformed into real SCC with a metastatic potential although it's rare [24-25].

Controversies remain about the management of the solitary KA. Some authors advocate the attitude of the « wait and see » due to its spontaneous regression [26,27]. However, the keratoacanthoma can reach several centimeters before regressing, leaving an unsightly scar that can some times bed is figuring.

The treatment of choice remains the complete surgical excision with 4 mm margins. It usually provids a good esthetic out come and allows histological study of the complete lesion which is an optimal specimen for the pathologist to make an absolute distinction of this lesion from SCC or other aggressive entity. Lesions less than 2 cm on the extremities may be treated with electrodessication and curettage [28]. Nonsurgical interventions have been limited to case reports and retrospective reviews consisting of topical 5% imiquimodcream, topical 5% 5-fluorouracil (5-FU) cream, intralesional methotrexate injections, intralesional bleomycin, intralesional 5-FU, and oral isotretinoin. Intralesional 5-FU have been administered at a dose of 40 to 75 mg weekly for 3 to 8 weeks. [29- 30]. What ever molecule isused, the intra lesional therapy is more indicated before surgery to reduce the tumor mass and get a better out come [31].

Wharever the treatment used for KA, clinical follow up is required for patients after KA removal due to the recurrences that occure in 1 to 8%. They should be advised to avoid provoking factors, including intense and prolonged ultraviolet light exposure, and to perform self-evaluation in all predisposed areas.

Conclusion

KA is a benign skin tumor with a rare but real malignant potential. It'soften developped on sun exposed skin but scrotal localization calls into question UV light as a risk factor. In KA, diagnosis should be based on clinical and pathological correlation, and its main differential diagnosisis the SCC. This justifies the use of surgical tratment of KA. How ever, a correct diagnosis discourages over treatment, and provides more treatment approaches such as topical drugs.

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