

Clear Cell Acanthoma with Atypical Location -Case Report and Literature Review-

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ABSTRACT: Clear cell acanthoma is an uncommon, benign lesion, usually present in middle-aged and elderly patients. It looks like an erythematous papule, solitary, with squama at the periphery, regularly located on the lower limbs. We present the case of an 80-year-old female, who was admitted in the Dermatology Clinic of Craiova for an infiltrative plaque, erythematous-squamous-crusty, well delimited, with sizes 1,2/1,5cm, polycyclic shape, located in the right groin. The onset of the condition was 7-8 years ago, with slow growth in size. The excision of the lesion was performed and the histopathological examination confirmed the diagnosis of clear cell acanthoma. Our case is atypical by location. Because clear cell acanthoma is difficult to be clinically diagnosed, with a wide range of lesions that make a differential diagnosis, we highlight the importance of histopathological examination for a positive diagnosis. The treatment depends on the type of lesion and on the patient's preference, usually consisting in surgical excision.

KEYWORDS: Clear cell acanthoma, histopathological diagnosis, treatment

Introduction

Clear cell acanthoma (CCA) was described for the first time in 1962. It is also known as “Degos acanthoma” or the “pale acanthoma” which is due to its histopathological features. It is an uncommon, benign lesion, most of the times, difficult to diagnose only based on the clinical aspect [1,2].

It is a rare lesion that develops like a domed, well-defined, papulae of reddish color, with a whitish border. It usually occurs as a single lesion, measuring less than 1cm in diameter, but giant shapes as well as multiple lesions have been described. It is usually located in the lower limbs, in middle-aged and elderly patients. Both genders are equally affected. Other locations are: foot, thigh, abdomen, anterior thorax. Less common localizations include nipple, lower lip, toe, hands, genitals, suprapubic region, navel, scalp [2].

Clinically, it may be difficult to be differentiated from pigmented actinic keratosis, a lentigo senilis, or even a flat and pigmented seborrheic keratosis [3,4].

Because clinically, this description coincides with a multitude of other lesions, dermatoscopy and biopsy are recommended for diagnosis purposes.

Case report

We present the case of a 80-year-old patient, female, from the rural area, who was admitted in the Dermatology Clinic of Craiova for an

infiltrative erythematous plaque, covered by a squamous scale, well delimited, with sizes 1,2/1,5cm, polycyclic shaped, located on the right groin (Fig.1,2). A written informed consent of the patient was obtained regarding the publication of these data.

The onset of the condition was 7-8 years ago, with slow growth in size during those years. Personal pathological history of the patient relieved primary high blood pressure since 1998, a left inguinal hernia operated in 1973. The patient was diagnosed with osteoporosis since 2000 and has had a left hip prosthesis in 2014.

Objective examination relieved a type III phototype according to Fitzpatrick scale, normal weight (body mass index: 22.2), having keratotic yellow-brown plaques, harsh on palpation, located on the face (actinic keratosis), xerotic and atrophic skin, with pronounced wrinkles, pigmented maculae confounding with hypopigmentation macula in the calves, mat thickened nails, varicose veins in the lower limbs.

Abdominal-pelvic ultrasound performed was without pathological findings. Laboratory analysis were within normal limits, except an increased value for erythrocyte sedimentation rate (45mm/h).

As a treatment it was practiced the surgical excision of the lesion, under local anesthesia with Xilin 1%. The surgical specimen was sent to Pathology Lab, where it was processed according to classic histopathological technique.

Histopathological examination revealed: skin with the presence of a psoriasiform hyperplasia

(Fig.3), an intraepithelial tumor proliferation with clear keratinocytes (Fig.4), hypogranulosis, parakeratosis and neutrophils throughout the epidermis and the stratum corneum (Fig.5). The blood vessels within the dermal papillae are dilated, tortuous and have a vertical orientation to the dermal papillae (Fig.6). The proliferation does not imply the hair follicles and the adnexial structures. Based on the clinical and histopathological conclusions we assumed the diagnosis of *Clear cell acanthoma*.



Fig.1. Right groin-clear cell acanthoma



Fig.2. Right groin-infiltrative plaque, erythematous-squamous, well delimited, with sizes 1.2/1.5cm, polycyclic shape

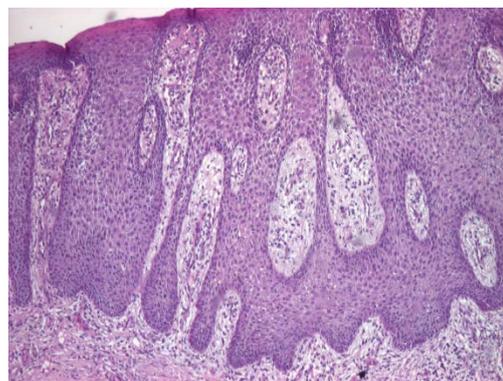


Fig.3. Psoriasiform hyperplasia, Hematoxylin-eosin staining, 4x

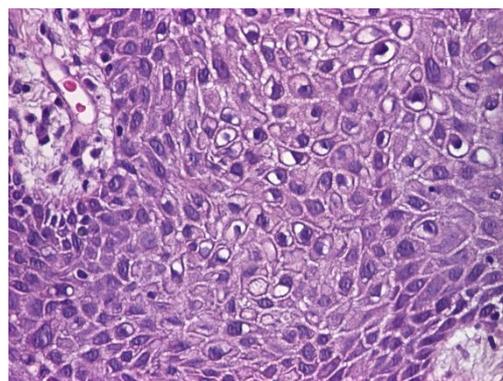


Fig.4. Clear keratinocytes, Hematoxylin-eosin staining, 20x

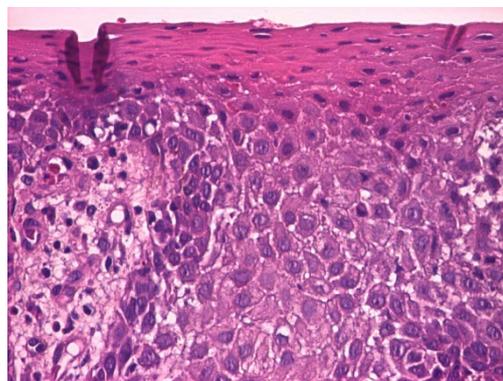


Fig.5. Parakeratosis and neutrophils throughout the epidermis, Hematoxylin-eosin staining, 20x

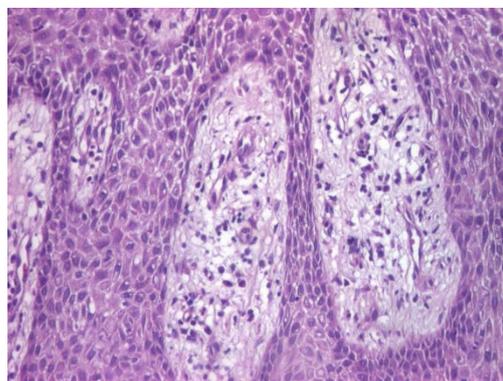


Fig.6. Blood vessels within the dermal papillae dilated, tortuous, Hematoxylin-eosin staining, 10x

Discussion

C. Fine and Chernosky described clear cell acanthoma as a "clinical hybrid" with skin-like appearance [5].

Etiopathogenesis is not known precisely. It was initially considered a benign epidermal tumor, but a more recent hypothesis defines clear cell acanthoma as a reactive inflammatory dermatosis [6].

The concept that CCA was a benign tumor was a controversy due to its unclear origins. The overall proposed sites of origin include epidermis, hair follicles, and sweat glands [7].

Ohnishi et al. considered the clear cell acanthoma as a localized form of inflammatory dermatosis because it has immunohistochemical characteristics of cytokeratin expression like in psoriasis, lichen planus, and discoid lupus [8].

Zedek et al. confirmed immunohistochemical stain similar to the chronic inflammation pattern observed in psoriasis. The emergence of clear cell acanthoma on the pre-existing active plaque psoriasis also backs up this hypothesis [9].

Other CCA-related conditions include: varicose veins, stasis dermatitis, seborrheic keratosis, bacterial dermatitis, viral infections, ichthyosis, xerosis, atopic dermatitis and insect bites. Evidence that clear cell acanthoma can be considered an inflammatory dermatosis includes clinical, histopathological, immunohistochemical and dermatoscopy appearance [2].

Clinical features

Clear cell acanthoma are generally solitary, asymptomatic, red or brown, dome-shaped papules or nodules. They may be covered by squama in the periphery or may have erosive surface. The size may vary from 3mm to 2cm and can grow slowly up to 10 years. It shows dotted vessels on the surface, easily bleeding to minor traumas [2]. Note that in our case, the CCA has a 7-8 years evolution and falls within the limits of these dimensions.

Although this is the most common aspect, there are other types of CCA:

1. *giant*, with dimensions over 4cm, located in the lower limbs, perineum, buttocks. Murphy et al. have described a case on a psoriasis plaque on the buttocks [7].

2. *polypoid*, it has between 4mm and 3cm. There have been cases on the thigh, popliteal fossa, legs, less on the scalp, neck, nipple and scrotum. Yang described polypoid CCA on a pre-existing melanocytic nevus on the neck [10].

3. *pigmentary*, also called melanoacanthoma or clear cell melanoacanthoma. It appears as brown papules [11].

4. *eruptive* (more than 20 injuries) frequently in the lower limbs, but it was also described on the hands and trunk [12].

5. *atypical*, considered by some authors a malignant form due to marked cellular atypia and mitotic reactions. A CCS in situ, which appeared on an CCA, was described. The malignant potential could be due to degenerative proliferative changes [13]. These cases have been clinically described as erythematous nodules located in the face. However, the name of benign lesion is preferred due to a lack of relapse.

6. *cystic*, described by Hamaguchi et al. in a middle-aged man, in the suprapubic region [14].

Positive diagnosis

Clear cell acanthoma is difficult to diagnose only based on the clinical aspect.

Dermatoscopically, it has features that facilitate the diagnosis, highlighting a stereotyped vascular pattern, composed of punctuated blood vessels distributed linearly in a "pearl string" configuration [15].

The diagnosis of certainty remains the *histopathological examination* which highlights: well-defined psoriasiform acanthosis with pale keratinocytes (clear cells), with high intracellular glycogen burden, PAS positivity; neutrophilic exocytosis; microarrays of parakeratosis; blood vessels dilated in the upper dermis.

The histopathological aspect in the atypical variant is as follows: cytological atypia of large nucleus tumor cells, some with mitotic reactions [16].

The histochemical study states that the phosphorylase required for glycogen degradation is absent in keratinocytes, and electron microscopy reveals glycogen granules in clear cell acanthoma [17].

Differential diagnosis

Clinically, CCA should be differentiated from psoriasis, piogenic granuloma, dermatofibroma, inflamed seborrheic keratoses, eccrine poroma, clear-cut hydradenoma, actinic keratosis, Bowen's disease, basal cell and spinocellular carcinoma, amelanotic melanoma [18].

From a histopathological point of view, the differential diagnosis is made with:

-*Bowen's disease* has cytological atypia in the entire thickness of the epidermis;

-*eccrine poroma*: lack of cytoplasmic glycogen, it has polygonal nuclei with basaloid aspect and ductal differentiation;

-*psoriasis vulgaris*: does not have well-defined lateral limits and lacks glycogen from the cytoplasm of cells, has thinner suprapapillary plates;

-seborrheic keratosis: intracytoplasmic glycogen is missing.

Treatment

If only one CCA is present, it is recommended to remove it by classic surgical excision, which we have practiced. Eventually one can call to Mohs surgery. Other treatments used are cryotherapy, curettage plus electrocoagulation, CO₂ laser.

Multiple or greater lesions have been successfully treated by cryotherapy and CO₂ laser [19,20].

We underline that a case has been published in the literature that has regressed after treatment with calcipotriol for 2 months, suggesting the inflammatory nature of clear cell acanthoma [21].

Conclusions

Clear cell acanthoma is difficult to be clinically diagnosed, with a wide range of lesions that make a differential diagnosis.

The diagnosis of certainty is based on the histopathological aspect, although the dermatoscopic examination provides some suggestive elements for a CCA.

The etiology of this lesion is unclear, and the treatment of choice is surgical excision.

Financial disclosure

None declared.

Conflicts of interest

None declared.

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