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Pediatric keratoacanthoma - A rare tale of keratin plugs in a bumpy dome - Case report

[1]Dr.MS Manasa, Post graduate, Ramaiah Medial College, Bengaluru.

[2]Dr.Vara Prasad BM, Assistant Professor, Ramaiah Medial College, Bengaluru.

[3]Dr. Clement Wilfred, Professor, Ramaiah Medial College, Bengaluru.

Corresponding Author: Dr. Vara Prasad BM, Assistant Professor, Ramaiah Medial College, Bengaluru.

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Abstract

Background: Keratoacanthoma is a cutaneous neoplasm with rapid growth. Its cclinical and histological features are very similar to those of squamous cell carcinoma. Differentiating keratoacanthoma from conventional invasive squamous cell carcinoma poses a great challenge due to its similar appearance. It usually arises on sun-exposed surfaces and is characterized by firm, symmetrical, dome-shaped nodules with a horn-filled crater in its center and it has a tendency for spontaneous regression.

Case presentation: 5-year-old child presented with multiple asymptomatic dark-colored spots over both the cheeks which gradually progressed to involve entire face within one year. These dark spots also appeared on both eyes at the age of two years. Similar skin lesions were noted over photo exposure body parts by the age of four years. Wide local excision of the lesion was done in view of clinical suspicion of malignancy.

Conclusion: Keratoacanthoma is an epithelial tumor with morphological characteristics similar to those of ssquamous cell carcinoma, so a correct diagnosis of keratoacanthoma based on clinicopathological findings facilitates appropriate treatment and provides a wide spectrum of available therapies.

Keywords: Keratoacanthoma, squamous cell carcinoma.

Introduction

Keratoacanthoma was first described in 1889 by Sir Jonathan Hutchinson as "crateriform ulcer of the face" [1]. It develops most often in the older age groups, particularly in the sixth and seventh decades, but it is rare in childhood and they also have male preponderance. Keratoacanthomas (KA) are common benign epidermal tumors characterized by the rapid development of a firm, symmetrical, dome-shaped nodules with a horn filled crater in its center and a tendency for spontaneous regression. Solitary KA remains the most common form with its three distinct stages: Proliferation, maturation,

and spontaneous involution. Typical cases reach their full size of 1.0-2.5 cm within 6-8 weeks, followed by spontaneous involution within less than 6 months [2]. It usually arises on sun-exposed surfaces. Its cclinical and histological features are very similar to those of squamous cell carcinoma [3]. Differentiating keratoacanthoma from conventional invasive squamous cell carcinoma poses a great challenge due to its similar appearance [4].

Case presentation

5-year-old child presented with multiple asymptomatic dark-colored spots over both the cheeks which gradually progressed to involve entire face within one year. These dark spots also appeared on both eyes at the age of two years. Similar skin lesions were noted over photo exposure body parts by the age of four years. Wide local excision of the lesion was done in view of clinical suspicion of malignancy. Microscopic examination from the nodule shows skin composed of epidermis and dermis. Epidermis shows tumours composed of squamous cells invaginating deep into the dermis and forms a crater. These squamous cells at the periphery of the lesion shows cytological atypia and abundant eosinophilic cytoplasm. The center of the crater shows keratin pearl formation. Dermis shows dense lymphoplasmacytic infiltrate along with giant cells of multi nucleate type. The lesion was well delineated from the underlying subcutaneous tissue and there is mitosis only limited to basal layer. There is no perineural invasion, deep infiltrative growth and marked atypia noted, hence the diagnosis of keratoacanthoma was given.

Discussion

Kkeratoacanthoma is a rapidly growing skin tumor which most often presents as single, nodular, round, firm lesion with a central depression filled with keratin [3]. Lesions are located in greater exposure to ultraviolet radiation and most commonly found on face and upper limbs. Predisposing or provoking conditions and factors that promote mmultiple kas can also appear in the context of rare genetic disorders that predispose to carcinogenesis, such as xeroderma pigmentosum and Muir-torre syndrome(MTS).Iatrogenic KA induced by drugs or medical procedures.With the rapidly growing number of aesthetic and antiaging procedures, the risk of inducing KA on sun-damaged skin by laser procedures (mostly resurfacing, including fractional laser), chemical peels and fillers has to be considered.[5]

Exophytic lesions with a central keratin-filled crater are difficult to diagnose clinically. Kerato acanthoma and squamous cell carcinoma shares some features so they cannot be confidently differentiated by dermoscopy So, Histopathological examination remains as the gold standard for the diagnosis of KA [2]. Diagnosis of the KA is based on 3 principles: typical clinical presentation of a crateriform tumor, rapid (weeks to months) growth with a triphasic course, and histopathological examination of a suitable biopsy specimen.[5]

Kkeratoacanthomas are Ex endophytic lesions with an invaginating mass of keratinizing, well-differentiated squamous epithelium at the sides and bottom of the lesion. There is a central keratin-filled crater that enlarges with the maturation and evolution of the lesion [1]. Another key feature is the lipping (also known as buttressing) of the edges of the lesion that overlap the central crater, giving it a symmetrical appearance. In some lesions, a keratotic plug overlies discrete infundibula, and a central horn-filled crater is not formed. A well-formed crater containing keratin is present in most regressing (involuting) lesions. The component cells have a distinctive eosinophilic hue to their cytoplasm and as they mature toward the center of the islands of squamous

epithelium, they can become quite large. Eosinophils and neutrophils may be prominent, and these may extend into the epithelial nests to form small micro abscesses [1]. Perineural invasion of keratoacanthoma is rare. It has a greater potential for aggressiveness. When perineural invasion is extended, the prognosis can correspond to that of squamous cell carcinoma with perineural infiltration, so a closer follow-up of the patient is recommended [4]

Histological features that favor diagnosis of keratoacanthoma over squamous cell carcinoma are: KA is an Ex endophytic lesion with a central horn filled crater and with overhanging lips of epithelium were as SCC is predominantly endophytic with no horn filled crater in it [6]. Volume-weighted mean nuclear volume is higher in keratoacanthomas than in squamous cell carcinomas [1]. There is lack of anaplasia and absence of stromal desmoplasia in KA, were as the presence of stromal desmoplasia is noted in SCC [1,6]. The presence of intraepithelial elastic fibers and intracytoplasmic glycogen has also been said to favor the diagnosis of keratoacanthoma [1]. In keratoacanthoma, the subcutaneous tissue is preserved. In SCC, there is an invasion in the interface of the underlying subcutaneous tissue.[3]

Conclusion

Keratoacanthoma is an epithelial tumor with morphological characteristics similar to those of ssquamous cell carcinoma.[3] Many studies addressing the problem of clear histopathological differentiation between SCC and KA support the concept of the peculiarity and importance of KA as a precise diagnosis. In keratoacanthoma, diagnosis should be based on clinical and pathological correlation. A correct diagnosis of KA discourages overtreatmentt [5]. "Overcall" can have as serious consequences for the patient as an "under call." [1]

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Legends Figures

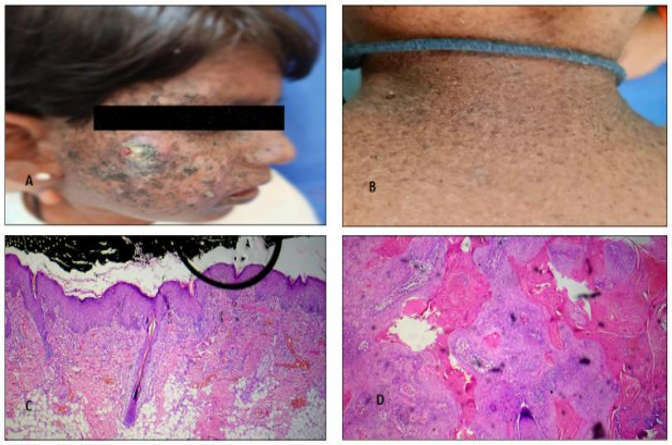

FIGURE (A & B) Gross specimen showing multiple dark colored spots of varying sizes which is present on the cheek than gradually progressed to entire body. (C & D) Microphotograph showing skin with epidermis with a tumor invaginating deep into the dermis and forms a crater with keratin pearl formation(H&E,10X).