

## CASE REPORT

# Giant Keratoacanthoma of Pinna: A Rare Presentation

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## ABSTRACT

**Aim:** The present case describes a rare and large size of giant keratoacanthoma (KA) of pinna.

**Background:** Keratoacanthoma is a fast growing benign cutaneous tumor resembling closely to well-differentiated squamous cell carcinoma (SCC) on clinical and histopathological examination (HPE).

**Case report:** A 70-year-old male presented with a rapidly growing mass on the left ear for 9 months. Tumor was excised and sent for HPE, which revealed well-differentiated SCC with focal features of a KA.

**Clinical significance:** Differentiation of KA from SCC has been a major challenge for dermatologists, especially at setups with unavailability of molecular studies. So, if the tumor is giant, nonregressing in size especially on sun-exposed sites in an elderly patient, always think of SCC and treat it by surgical excision rather than watching for a spontaneous resolution.

**Keywords:** Excision, Giant, Keratoacanthoma, Squamous cell carcinoma.

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**Conflict of interest:** None

## INTRODUCTION

The term keratoacanthoma (KA) was coined by Freudenthal in 1940 in view of considerable acanthosis seen on the histopathological examination (HPE).<sup>1</sup> Giant

KA is a clinical variant of KA, where the size of the tumor exceeds 20 to 30 mm.<sup>2</sup> Similar to other forms of KA, it may regress spontaneously or can have an aggressive course to behave like squamous cell carcinoma (SCC). It is said that KA arises from pilosebaceous unit and due to this, most KAs are found in hair-bearing areas. Furthermore, immunohistochemical markers of KA and outer root sheath of hair follicle are similar, and biological evolution of KA corresponds with the hair follicle cycle.<sup>3</sup> The KA has also been seen in areas free of hair follicle like glans penis and conjunctiva, which creates suspicion regarding the exact site of origin.<sup>4,5</sup> The hallmark for diagnosis of KA is the spontaneous resolution of the lesion, which reacquires Griffith's wait and watch principle for management.<sup>6</sup> But this principle cannot be generalized due to a definite risk of postinvolvement scarring and malignant transformation. Due to poor differentiation of KA from SCC by clinical, HPE, or even by immunohistochemical stains, most dermatologists use the term "SCC, KA type."<sup>7</sup> Accordingly, multiple therapeutic modalities have been individualized for the treatment of KAs, but wide excision of tumor is the treatment of choice for the majority of solitary KAs including the giant variants.<sup>3</sup>

## CASE REPORT

A 70-year-old male presented with a large, solitary, dome-shaped growth on right pinna that had been rapidly growing for the past 9 months. The lesion was asymptomatic and further there was no history of any trauma or previous lesion at the same site. On examination, there was a single giant, pinkish-red dome-shaped tumor of 5 cm size involving helix of the left pinna. The tumor had a crater in the center filled with black keratin plug with the smooth shiny surrounding border (Fig. 1). The lesion was firm in consistency, nontender, and there was no evidence of invasion into deeper structures. Lymph nodes were not palpable in surrounding area and neck. Routine investigations and workup to rule out mycotic etiology were within normal limits. Fine needle aspiration cytology was not fruitful. Wide excision of tumor with adequate skin margins was done and the specimen was sent for HPE with clinical possibilities of KA, SCC, and mycotic infection. Histological examination showed cellular anaplasia, increased mitotic activity admixed with lymphocytic infiltrate, and no stromal invasion

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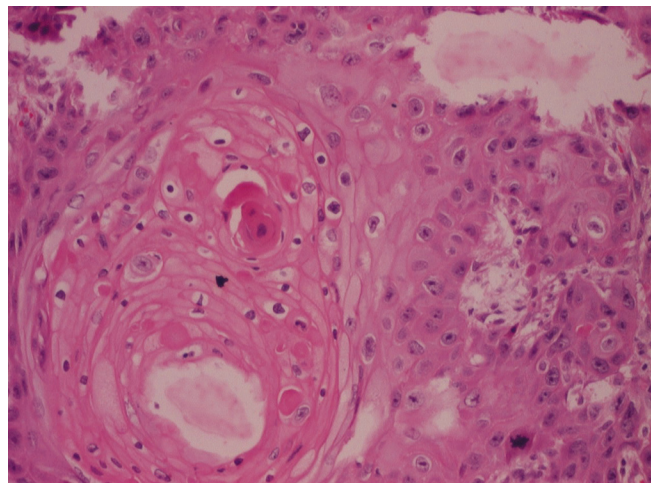
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**Fig. 1:** Giant KA type of SCC on left ear in a 70-year-old male



**Fig. 2:** Histopathological examination view well-differentiated SCC with focal features of KA

suggesting well-differentiated SCC with focal features of a KA (Fig. 2).

## DISCUSSION

The KA is an antique benign squamous epithelial neoplasm, which can progress into malignancy.<sup>7</sup> Once KA was a totally different entity from SCC due to its unique morphology, biological behavior, and immunohistochemistry. Weedon et al<sup>8</sup> reviewed 3,465 cases of KA retrospectively over a period of 14 months and found that 200 cases had development of SCC within the lesion. Terminology, classification of KA, and other types of SCC with crateriform architecture have not been clarified in the literature. The KA can be histopathologically classified into six categories: (1) KA (well-developed stage), (2) KA-like SCC, (3) KA with malignant transformation, (4) SCC (crateriform), (5) crateriform SCC arisen from actinic keratosis, and (6) crateriform Bowen's disease. Although the true characteristics of KA-like SCC remain still unclear but assumptions say that either it is one form of KA with malignant transformation or it is one step in the evolution of KA, or it is a borderline lesion between KA and invasive SCC.<sup>7</sup>

The evolution of KA is a three-stage process as described by Schwartz<sup>9</sup>: Proliferation or rapid growth phase, maturation phase (development of central keratinous core), and finally involution by necrosis and scarring. It attains maximum size in 4 to 12 weeks and after that it undergoes spontaneous involution usually over a period of 2 to 4 months, but sometimes the lesion can persist even for 1 year. The mechanism for regression has been explained by Paterno's "Regression Theory",<sup>10</sup> which hypothesizes that an individual's immune response elevates the number of T cells which then express granzyme B in local tissue. It is the higher number of CD8 T cells that kill tumor cells and lead to the regression of

the neoplasm in KA as compared with SCC. However, tumor does not involute always and it may transform into malignancy or can have aggressive course with perineural, perivascular involvement, and even metastasis to regional lymph nodes.<sup>11</sup> Malignant transformation has been reported in 25% cases, and this rate is higher if patient is elderly and tumor is present on photo-exposed site.<sup>12</sup> For this reason, KAs should be treated empirically as SCC rather than waiting for spontaneous resolution.

Etiology of KA still remains obscure but factors implicated in SCC like actinic rays, human papillomavirus, trauma, genetic factors, and immunocompromised status play some role. Both KAs and SCC are usually seen on sun-exposed areas of lighter-skinned individuals with peak incidence in the fifth decade.<sup>13</sup> Although KA appear as a solitary lesion, multiple tumors have been seen in various syndromes like Muir-Torre, xeroderma pigmentosum, and nevus sebaceous of Jadassohn.

Cribier et al<sup>14</sup> proposed five histological criteria to differentiate KA from SCC, which include epithelial lip (marginal epithelial buttresses) and sharp demarcation between tumor and stroma that suggest KA while cellular anaplasia, pleomorphism, and significant mitotic activity favor the diagnosis of SCC. In the present case, features of SCC were more prominent with only focal findings of KA in the form of lymphocytic infiltrate and noninvasion. Treatment modalities implicated are surgical excision, Mohs micrographic surgery, electrodesiccation and curettage, intralesional corticosteroids, topical and intralesional 5-fluorouracil, systemic retinoids, podophyllin, radiation therapy, and interferon alfa and methotrexate. Surgical excision remains the treatment of choice for giant KA and same principle was followed in our case. Advantages of this modality include rapid treatment, definitive HPE, prevention of local invasion, and minimization of scarring, which may be significant

in postinvolution. Recurrence rates have been reported in 4 to 8% of cases after surgical excision.<sup>6</sup> The KA is believed to have a good prognosis; however, it has been reclassified as SCC-KA type to reflect the difficulty in histological differentiation, as well as the uncommon but definite risk of transformation into SCC.

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