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Case Report

The diagnosis of rare skin lesions in ear, nose, and throat: the clues lie in clinical examination

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ABSTRACT

Here we present 2 case reports of 2 relatively rare skin lesion that can be encountered in ENT practice, and touch upon the basic clinical evaluation that is mostly overlooked in favour of investigations to reach a diagnosis, since in many cases, investigations fail to give a clear result. The first case is a pilomatricoma, which can be tricky differential diagnosis to reach in the preoperative phase, with the lesion not having any clear differentiating characteristics on imaging. But the answer to a preoperative diagnosis might lie in the "Tent Sign", bluish discoloration of the skin, or calcium extrusion from the lesion. The second case is a keratoacanthoma, which, waiting long enough should regress on its own. But as watchful waiting is not a feasible line of management, the distinct crateriform pattern should be looked for in lesions on sun exposed areas, and if present, instead of just an incisional biopsy or wedge biopsy, complete surgical excision with clear margins and intact architecture should be planned.

Keywords: Pilomatricoma, Keratoacanthoma, Dufourmental flap

INTRODUCTION

Pilomatricomas are benign superficial tumours of the skin, mostly occurring in the head and neck region in young adults. They are frequently misdiagnosed as they are relatively unknown to otorhinolaryngologists, despite defining clinical features such as a solitary, slow growing course and presenting as firm, painless skin lesions, often with either a bluish discolouration or with calcium crystals seen being extruded from the skin surface.[1] Preoperative investigation in the form of imaging or fine needle aspiration cytology (FNAC) often show inconclusive results resulting in misdiagnoses.[2] Keratoacanthomas on the other hand, are asymptomatic crateriform lesions found in fair skinned people on sun exposed areas of the body, which are frequently misdiagnosed as squamous cell carcinomas.[3]

Although seen in the dermatology speciality, ENT surgeons do also occasionally see these lesions in their practice. Over reliance on imaging and preoperative cytology has slowly overtaken basic clinical examination. In cases the ENT surgeons see in day-to-day practice, although clinical examination is given adequate importance, this case report aims to highlight the basic signs and symptoms that can aid in reaching a preoperative diagnosis in cases of problematic skin lesions that are not commonly seen by ENT surgeons, in places where imaging and cytology can fail to provide results, thus aiding in surgical planning and treatment.

Case 1

A 35-year-old female presented with a 2x2 cm swelling just below and behind the left angle of mandible since the last 5 months (Figure 1).

The size of the swelling had been gradually progressing since the last few months. The swelling was firm, tender, mobile in all axes and puerile discharge stained. There

was no other similar lesion any anywhere else on the body. An ultrasonography (USG) surface scan showed non-specific features of an anechoic oval lesion with internal shadows, features suggestive of an abscess, and an FNAC done was inconclusive, showing features of inflammation. We decided to undertake complete excision of the mass instead of an incision and drainage because clinically the features did not suggest an abscess. Since the excision of the mass resulted in slight straining of the skin on turning the head to the opposite side, a rhomboid flap (Dufourmental type) was used for the closure of the defect (Figure 1).

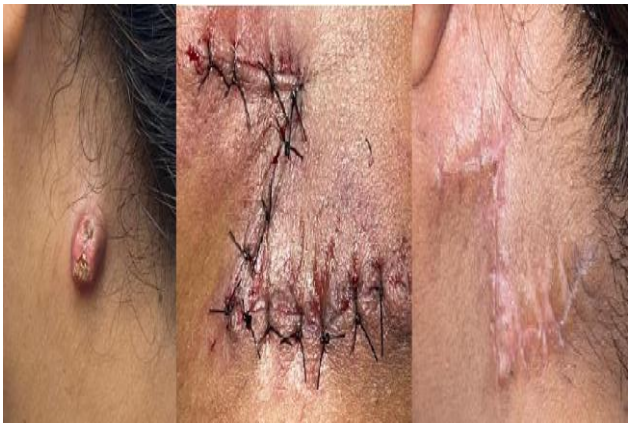

Figure 1 (Left to right): Superficial skin lesion with ulceration and calcium crystals; rhomboid flap (Dufourmental type); day 10 post operative picture.

A post operative histopathological examination (HPE) showed a partially cystic lesion lined by stratified squamous epithelium with central ghost cells and multiple foci of calcification. Hence a diagnosis of pilomatricoma was arrived at (Figure 2).

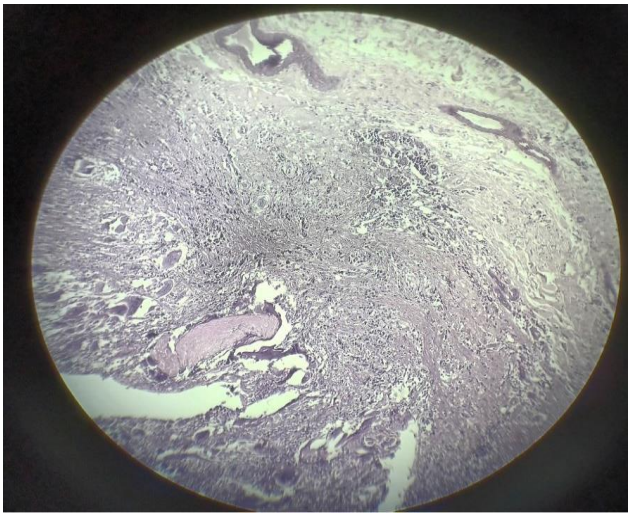

Figure 2: Histopathology of ghost cells, foreign body-type giant cells and calcification.

At 1 year of follow up, the patient has not yet had any recurrence.

Case 2

A 62-year-old male presented with a 1.5×1.5 cm lesion on the tip of the nose, progressively increasing in size since 2 months (Figure 3). The lesion was insidious in onset, gradually progressive, with no past history of trauma or radiation exposure. The lesion was firm and non-tender, and blackish on the sides with a pinkish crateriform centre. An FNAC of the lesion was inconclusive, so an excision biopsy was done.

Figure 3 (Left to right): Lesion at tip of nose with crateriform centre; post operative picture after excision.

HPE revealed keratin containing epidermal invagination with acanthosis, patchy hypergranulosis, horn cysts and pseudo horn cysts. glassy keratinization, loss of polarity, nuclear atypia, foci of histiocytes and giant cell reaction were also noted, leading to a diagnosis of keratoacanthoma (Figure 4).

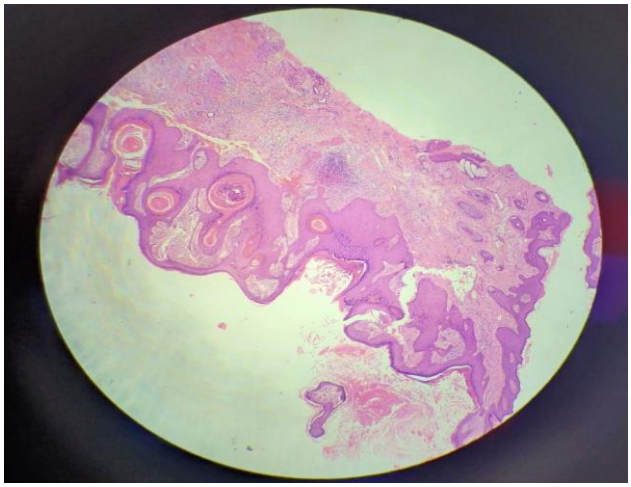

Figure 4: Histopathology of acanthosis and horn cysts.

The patient did not have any other lesions anywhere else on the body, and has not had a recurrence after 6 months of follow up.

DISCUSSION

Pilomatricomas earlier known as pilomatrixomas are benign, slow growing, superficial skin lesions. They are said to occur before the age of 20 years in most cases, the head and neck region being the predisposed site for their occurrence.[1] Originally described in 1880 by Malherbe as a benign tumour of the sebaceous glands (a "calcifying epithelioma of Malherbe"), it underwent modifications in nomenclature, first by Dubreuilh and Cazenave and then by Forbis and Helwig (who termed it as a "pilomatrixoma" to convey the histogenesis of the tumour).[4-6] "Pilomatrixoma" as a term was re-iterated by Jones and Campbell.[7] Finally, the term "pilomatricoma" was used by Hermann Pinkus, as was pointed out by Arnold Jr.[8] In 1973, Friedrich showed that pilomatricomas have an incidence of about 0.12% among skin tumours (170 out of 1,40,000 skin tumours).[9] The peak presentation is accepted to be the first 2 decades of life, with a second peak between 50 and 65 years.[2,10] Sex predilection ranges from 1.5:1 (female: male) to equal in both sexes.[1,10] Occurrence is not affected by either occupation or geographical location. It also does not seem to have any hereditary linkage.[6] The majority of cases are seen in the head and neck region (neck> cheeks> scalp> brow> periorbital area), followed by the upper extremities, trunk and lower extremities.[1] They present as solitary, slow growing, firm, painless skin lesions, often with either a bluish discolouration or ulceration of the skin. Although predominantly solitary, 2-10% cases present with multiple lesions, which may be associated with Gardner Syndrome, Turner Syndrome, Steinert disease, myotonic dystrophy and sarcoidosis.[1] Activating β catenin gene mutations have been described in association with this tumour, and its locus has been mapped to CTNNB1 gene on 3p22-p21.3.[11] In superficial lesions, calcium crystals may be seen being extruded from the skin surface. Around 20% of the lesions are detected by palpation as they may be deeply subcutaneous.[10] Of particular clinical aid is the "tent sign" described by Graham and Merwin, where flattening a portion of the tumour with angulation, it produces a flat facet akin to a tent, which may be due to calcium deposits.[12]

Investigations include a baseline ultrasonography (USG) and an FNAC. Imaging studies tend to be inconclusive, with non-specific findings such as "complicated skin appendage lesion", although the calcification may help in the diagnosis. Overall, imaging studies are not considered helpful.[2] A confident diagnosis on FNAC can be made if a combination of basaloid cells, ghost cells, nucleated squamous cells, and foreign body-type giant cells are seen in a superficial skin nodule, especially in a child or young adult. Other features that may be present include multinucleated giant cells and keratin clumps.[13] An accurate diagnosis is established in 0-30% of cases, and differentials that can be considered include sebaceous cyst, dermoid cyst, foreign body reaction, calcified lymph node, fat necrosis, nonspecific cyst, molluscum contagiosum and cartilage.[1,11] The management entails complete surgical excision of the lesion, with a few authors preferring incision and curettage.[2] Recurrences after complete excision can be seen in 0-3% of cases, and a malignant transformation can be suspected in cases of repeated localizes recurrences. Although rare, has been reported in around 80 cases worldwide.[1,2]

A Dufourmental flap is a modified rhomboid flap with a broader pedicle. This was done to improve the cosmetic outcome; and relieve the tension at the area of excision. This flap has been used for the closure of almost any size, with Borges believing this flap is preferable to primary closure in facial reconstructions, even for small lesions. This was because the flap wastes much less normal tissue and the suturing can be done with much less tension.[14] This leads to lower cases of wound dehiscence and overall lesser complications when compared to primary wound closure.[15]

A case series of 21 cases by Mikiko et al showed aesthetically superior results and no post operative complication in 100% of cases of small (<12 mm) facial lesions using pedicled flaps in place of simple fusiform excision. They have also hence advocated the use of flaps in place of simple excision of small benign tumour of the face to achieving physiological and aesthetically superior results.[16] Similarly, Li et al used local flap reconstruction in cases as small as 0.9 cm for aesthetically superior results, and had a satisfactory outcome in 41 of the 48 patients in their study.[17] Osman had also described the versatile nature of the Dufourmental flap, being used all over the body for lesions ranging from 1 cm to 14 cm with exquisite results.[18]

Keratoacanthoma, fist described by Sir Jonathan Hutchinson in 1889, with names such as ''molluscum sebaceum,'' ''pseudotumor,'' ''regressing tumor,'' and ''self-healing squamous cell carcinoma'' (SCC) being later given to this entity, to reflect the controversies surrounding it.[3,19] The now accepted name keratoacanthoma (KA) was first proposed by Freudenthal in the 1940s, as was commented on by Dr Arthur Rook.[20,21] Its true incidence is often incorrect due to being misdiagnosed as squamous cell carcinoma, but it is estimated to be around 105/100,000 in Australia, more in fair skinned people, with a peak around 65-71 years of age and men being more affected than women.[3] It is seen more commonly on sun exposed areas of the body, with UV light being considered important for its development. Other factors contributing to its formation include x ray or megavolt radiation, CO2 ablation therapy, chemicals and an immunocompromised state. It is assumed to originate from hair follicles, implying that it may be the benign counterpart of follicular SCC rather than ordinary SCC. Multiple varieties of keratoacanthoma exist, each with their own subtypes, like solitary (subdivided into typical, giant, subungual, mucosal or centrifugum) and multiple (Ferguson Smith type, Muir-Torre, Witten and Zak type etc).[3, 22] It has also been assumed that KA might

be an SCC deficient in blc-2 expression, and expression of p27 in ka might be an important factor. Wnt pathway is also responsible for its proliferation, along with H-ras mutations also being commonly seen in KAs. Solitary Kas are normally 1-2cm in diameter and 0.5mm thick and their natural history is mapped out in 3 stages: a proliferative phase (skin collared papule with an acanthotic hyperproliferating epidermis), mature phase (typical prominent dome shaped symmetric nodule with a keratin plug in its centre. Epidermal lips rise around both sides and cover partially the top of the crater) and a regression phase lasting about 12 weeks.[3,22,23] Although not clearly understood regression may be caused by cytotoxic T cells by release of granzyme B, keratinocyte apoptosis or by mechanisms related to normal hair follicle cycle.[24]

The diagnosis is based on 3 key factors: rapid development of a crateriform lesion, triphasic evolution and HPE of a lesion with intact architecture (which is why cytology is not sufficient for a diagnosis). Theoretically, KA can be monitored for spontaneous regression, but it may be difficult to predict the maximum size before regression, and may leave an unsightly scar. Hence the definitive management consists of surgical excision with clear margins and preserving the architecture for accurate post operative HPE diagnosis.[25] Other therapeutic options range from curettage with electrodesiccation, radiotherapy, laser and cryosurgery to intralesional and topical 5-fluorouracil, intralesional bleomycin or methotrexate, systemic retinoids, cyclophosphamide and 5-FU.[23] It has a post-surgical recurrence rate of 3-5%.[24]

Whether KA is a benign or malignant lesion is still debated, with its occasionally seen malignant behaviour, and its unknown potential for metastasis. On the other hands KAs do not display prominent mitosis or cytological atypia as observed in malignancies.[22]

CONCLUSION

Pilomatricomas can be tricky differential diagnosis to reach in the preoperative phase, with the lesion not having any clear differentiating characteristics on imaging. But the answer might lie in the tried and tested methods of thorough clinical examination. Giving ample importance to minor details such as the “Tent Sign,” bluish discoloration of the skin, or calcium extrusion from the lesion will not only aid in the diagnosis but prevent the patient from undergoing the financial burden of an imaging study that will not provide much further details, while also speeding up the process of definitive surgical management. Keratoacanthomas, on the other hand have the advantage of being a triphasic lesion, which, waiting long enough should regress on its own. But as watchful waiting is not a feasible line of management, the distinct crateriform pattern should be looked for in lesions on sun exposed areas, and if present, instead of just an incisional biopsy or wedge biopsy, complete surgical excision with clear margins and intact architecture should be planned. Hence, in the era of high-end investigations, wherever there is a doubt, one should always remember the importance of clinical examination.

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REFERENCES

1. Lan MY, Lan MC, Ho CY, Li WY, Lin CZ. Pilomatricoma of the head and neck: a retrospective review of 179 cases. Arch Otolaryngol Head Neck Surg. 2003;129(12):1327-30.
2. O’Connor N, Patel M, Umar T, Macpherson DW, Ethunandan M. Head and neck pilomatricoma: an analysis of 201 cases. Bri J Oral Maxillofacial Surg. 2011;49(5):354-8.
3. Kwiek B, Schwartz RA. Keratoacanthoma (KA): An update and review. J Am Academy Dermatol. 2016;74(6):1220-33.
4. Malherbe A, Chenantais J. Note sur l’e´pithe´lioma calcifie´des glandes se´bace´es. Prog Med. 1880;8:826-37.
5. Dubreuilh W, Cazenave E. De l’epithelioma calcifie: etude histologique. Ann Dermatol Syphilol. 1922;3:257-68.
6. FORBIS R, Helwig EB. Pilomatrixoma (calcifying epithelioma). Arch Dermatol. 1961;83(4):606-18.
7. Jones PG, Campbell PE. PILOMATRIXOMA: A not uncommon Hamartoma of Infancy and Childhood. J Paediatr Child Heal. 1969;5(3):162-6.
8. Arnold Jr HL. Letter: pilomatrixoma. Arch Dermatol. 1977;113:1303.
9. Moehlenbeck FW. Pilomatrixoma (calcifying epithelioma): a statistical study. Arch Dermatol. 1973;108(4):532-4.
10. Julian CG, Bowers PW. A clinical review of 209 pilomatricomas. J Am Academy Dermatol. 1998;39(2):191-5.
11. Kumaran N, Azmy A, Carachi R, Raine PA, Macfarlane JH, Howatson AG. Pilomatrixoma—accuracy of clinical diagnosis. J Pediatr Surg. 2006;41(10):1755-8.
12. Graham JL, Merwin CF. The tent sign of pilomatricoma. Cutis. 1978;22(5):577-80.
13. Wang J, Cobb CJ, Martin SE, Venegas R, Wu N, Greaves TS. Pilomatrixoma: clinicopathologic study of 51 cases with emphasis on cytologic features. Diagnostic Cytopathol. 2002 Sep;27(3):167-72.
14. Borges AF. The rhombic flap. Plastic Reconstructive Surg. 1981;67(4):458-66.

15. Kang AS, Kang KS. Rhomboid flap: Indications, applications, techniques and results. A comprehensive review. Ann Med Sur. 2021;68:102544.
16. Ooshima M, Kochi M, Morioka D, Miyamoto T, Tsuda T, Kadomatsu K. Bipedicled mini VY advancement flap for skin defect of the face. Modern Plastic Surg. 2018;8(02):15.
17. Li ZR, Jiang Y, Zhang JY, Su YW, Hu JZ. Modified OT advancement flap for reconstruction of skin defects. Int J Clin Experimental Pathol. 2017;10(9):9158.
18. Aydin OE, Tan O, Algan S, Kuduban SD, Cinal H, Barin EZ. Versatile use of rhomboid flaps for closure of skin defects. Eurasian J Med. 2011;43(1):1.
19. Hutchinson J. Morbid growths and tumors: 1: the ‘‘crateriform ulcer of the face’’ a form of acute epithelial cancer. Trans Pathol Soc London. 1889;40:275-81.
20. Musso L, Gordon H. Spontaneous Resolution of a Molluscum Sebaceum. Proceedings of Royal Society Med. 1950;43(11):838-9.
21. Rook A, Whimster I. Keratoacanthoma–a thirty year retrospect. Brit J Dermatol. 1979;100(1):41-7.
22. Gleich T, Chiticariu E, Huber M, Hohl D. Keratoacanthoma: a distinct entity? Experimental Dermatol. 2016;25(2):85-91.
23. Schwartz RA. Keratoacanthoma: a clinico-pathologic enigma. Dermatol Surg. 2004;30(2 Pt 2):326-33.
24. Ko CJ. Keratoacanthoma: facts and controversies. Clin Dermatol. 2010;28(3):254-61.
25. Tisack A, Fotouhi A, Fidai C, Friedman BJ, Ozog D, Veenstra J. A clinical and biological review of keratoacanthoma. Brit J Dermatol. 2021;185(3):487-98.

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