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Generalized Eruptive Keratoacanthomas Involving the Auricles and External Auditory Canal:ACase Report and Literature Review

ZhangL[1#],SunHY[2#]andLuDK[1*]

[1]Department of Otorhinolaryngology, Hwa Mei Hospital, University of ChineseAcademy of Sciences & Ningbo Institute of Life and Health Industry, University of ChineseAcademy of Sciences, Ningbo 315000, Zhejiang, China

[2]DepartmentofOtorhinolaryngology,UnionHospital,TongjiMedicalCollege,HuazhongUniversityofScienceandTechnology, Wuhan, 430022, China

[*]Correspondingauthor:

Da-kai Lu, DepartmentofOtorhinolaryngology,HwaMei Hospital, University of Chinese Academy of Sciences&NingboInstituteofLifeandHealth Industry, University of Chinese Academy of Sciences, Ningbo 315000, Zhejiang, China, E-mail: nbldkai@163.com

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ZhangL,SunHYTheseauthorscontributeequallyto this work.

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1. Abstract

Objective: Theaimofthisstudywastoreportacaseof GeneralizedEruptiveKeratoacanthomas(GEKA)involvingtheauricles and external auditory canal and to review the relevant literature.

Methods:A patient with bilateral auricles stenosis and External Auditory Canal (EAC) atresia associated with GEKA is described. We performed a systematic review of the literature to identify and compare similar cases.

Results: This case report described a 54-year-old female patient with external auditory canal stenosis and auricle lesion as- sociated with GEKA.AComputed Tomography scan (CT) of the temporal bone.

Conclusion: Bilateral auricles stenosis and atresia of EAC associatedwithGEKAisararecase.Itprovidesanewperspective for the etiological diagnosis of acquired auricles lesion as well as stenosis and atresia of EAC in the future. The surgical strategy is necessaryforthemanagementoftheacquiredstenosisandatresia oftheEACthatassociatedwithGEKA.Furtherresearchisneeded to increase the number of clinical cases to establish whether and when the surgery is necessary for this disease.

2. Introduction

Keratoacanthoma (KA) is a common cutaneous skin tumor that originates from the hair follicles. It is characterized by unproven position on the border between malignancy and benignity1. Solitary KAis the most common form but familial multiple KA, genetically predisposed KA or sporadic multiple eruptive KA also havebeendescribed[1].GeneralizedEruptiveKeratoacanthomas (GEKA) is an extremely rare condition. Both cutaneous and mucosal are involved in GEKAwithout certain genetic background. Approximately 40 cases have been reported [2]. But so far, there isnocasereportofauditoryorganslesionassociatedwithGEKA.

Herein, we report a rare female case. To our knowledge, this is the first report describing a case of GEKAwith external auditory canalstenosisandauriclelesion.Wehopethispreviouslyunchar-

acterized clinical association may provide additional insight on management.

3. Case Report

A 54-year-old female patient transferred to the clinic of otorhinolaryngology because of congestion, swelling and lesion of bilateralauriclesinDecember2018.Herpastmedicalhistorywas

unremarkable.Sheexperiencedasuddenonsetofsomescalypap- ules located on her ears without any identified trigger. There was nohistoryofsimilarskindiseaseinherfamilyandthehistory of ear trauma, chronic inflammation and chemical exposure were all denied.Afew months, the lesion was widespread in the whole body,buttheenlargedsuperficiallymphnodesofthewholebody were not found.Histopathological examination of the specimen obtainedfromthepatient'sheaddemonstratedkeratinocyteswere surrounded by more inflammatory infiltrate and the cells become larger(Figure1A-A'').According tothetypicalsymptomsandhis- topathological examination the patient was diagnosed as GEKA after admitted to the department of dermatology in December 2016. Then she was followed up by the dermatologist for a long time.Thedermatologistgavecephalosporinantibioticsandtopical corticosteroidstopreventinfectionandlocalinflammation.How- ever,theabovesymptomsimproved.Thecongestion,swellingand lesion of bilateral auricles were aggravated, with obvious hearing lossandearfullness.Thepatientwasurgentlyreferredtotheclinic of otorhinolaryngology for further diagnosis and treatment. The physicalexaminationrevealedscleroticandmask-likefacieswith marked bilateral eyelid ectropion (Figure2 A); Auricular lesions exhibitedcongestionandswelling,erythematouspapulesandker- atoacanthoma-likenodules.Structuressuchastriangularfossa,ear nailboat,oppositetragus,oppositeearwheeldisappeared(Figure2 A',A'').Thereisnothingspecialelse.Otoscopyrevealedstenosis oftheEACandintacttympanicmembraneofbilateralear(Figure 3).Atthesametimethetuningforktestwasperformed:Rinnertest of bilateral ear was positive, Weber test sound was heard equally loudly in both ears. Pure tone audiometry revealed sensorineural hearinglossatFrequenciesof2kHZ,4kHZand8kHZ(Figure4A, B),andtherewerenotympanometryfindings(Figure4C).Atthis point,aCTscanwastakeninordertocompletetheauditorycanal evaluation(Figure 6A-A'). Furthermore,upon examination,there was no history of vertigo, tinnitus or facial nerve weakness.

Laboratoryevaluationshowedanti-thyroglobulinantibody > 500IU/ml (normal < 60IU/ml), anti-peroxidase antibody > 1300IU/ml(normal<60IU/ml).HumanPapillomavirus(HPV) DNA,Treponema pallidum and HIVantibodies were all negative on the skin lesions. The antinuclear antibody was 1:320. The an- ti-SSandanti-Roantibodywasbothpositive.ChestCTshowedno obviousabnormalityinthelungs.Thepatientissuspectedtohave a history of dry mouth and eyes, and denies the history of joint pain.Sjogren'ssyndromecouldnotbeexcludedsincesherefused to undergo further examination such as lip gland biopsy.

History of drug treatment:After diagnosed as GEKA, the patient was treated with 30mg oral isotretinoin, 0.1% tretinoin ointment externally. However, the disease continued to progress and the dose of acitretin was increased to 40 mg/day. Congestion and swelling of bilateral auricle turned better through antibiotics and anti-inflammation treatment. The occurrence of ear fullness and hearinglosswasnottakenseriously.Theauriclelesioncontinued toprogress,butnopathologicalexaminationofauriclelesionwas given. The dose of acitretin was increased to 50 mg/day, while cyclophosphamide was given by intravenous pulse therapy, with 600mgoncemonthly,6monthsasonecourseoftreatment.There werenoreportsandclinicalevidenceofauriclelesion,stenosisand atresia of EAC and hearing loss associated with the above drugs. thereisnohistoryofdrug-inducedandnoise-inducedhearingloss.

First year follow-up: On November 25, 2019, the patient visitedto the clinic of otorhinolaryngology for the first-year follow-up. She complained that the fullness and hearing loss of bilateral ear weremoreseriousthanbefore.Physicalexaminationrevealedthe mask-like facial expression and marked ectropion were the same withoneyearbefore(Figure2B).Thecongestion,swellingandle- sionoftheauriclewerealmostnodevelopment(Figure2B',B''). However, the EAC became narrower. Cerumen embolism could be seen at the external orifice of the EAC and we could not see the tympanic membrane because of the narrowed EAC. CT scan revealed minimum anteroposterior diameter of EAC (Right ear: 3.05mm.Left ear: 3.25mm) thickening of soft tissue in the bilat- eralEACespeciallynearthetympanicmembrane(Figure6B,B').

The patient was continuously given regular follow-up. Howev-er, she died of multiple organ failure syndrome on November 20, 2020.

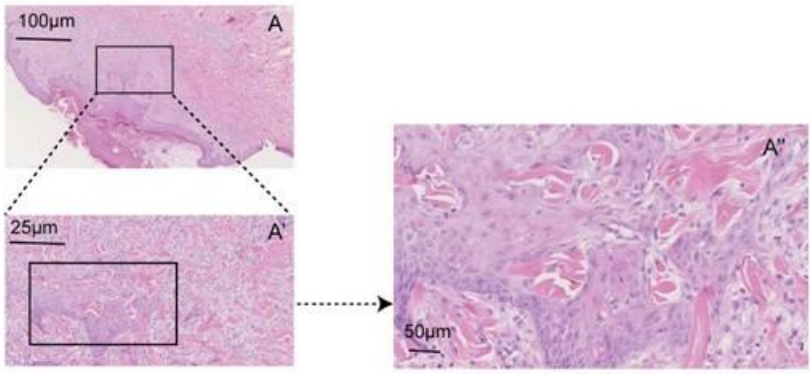


Figure1:Hematoxylin–eosinstainingofskinbiopsyspecimensobtainedfromthescalp.(A) Hyperplasiaofsquamousepitheliumwithhyperker- atosisandhypokeratosis;squamousepithelialcellsshowedinfiltrativegrowthdeepintothedermis;acentralcraterfilledwithkeratincellatypiaanda little lymphocyte infiltration **(A'-A'')** High magnification figures ofA.

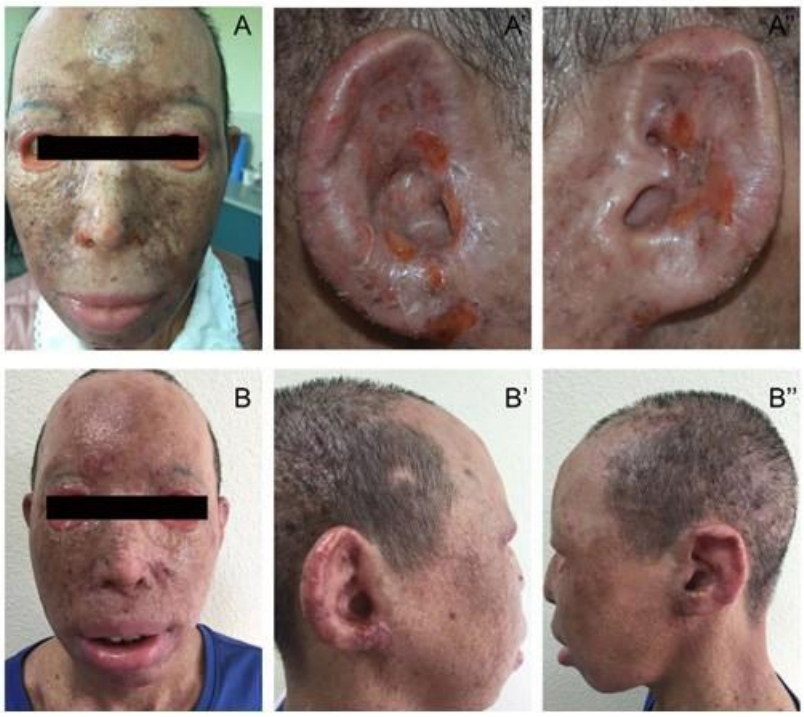


Figure 2:The appearance of the face and ears. (A-A'')At first visit. (A) Mask-like facial expression and ectropion. erythematous papules, kerato-acanthoma-likenodulesontheface.(A',A'')Bilateralauricleswereobviouslycongestedandswollen,localskinruptured.Normalstructuressuchas scaphoid fossa, triangular fossa, auricular nail boat, antiauricular tragus and opposite auricle disappeared, and the external orifice of external auditory canal was narrow. (B-B'')At 1-year follow-up. (B) Mask-like facial expression and ectropion was the same as before. (B', B'') Congestion, swelling and rupture of bilateral auricles were significantly less. Normal structures such as scaphoid fossa, triangular fossa, ear nail boat, opposite tragus and opposite ear wheel disappeared, and the right side was more obvious.

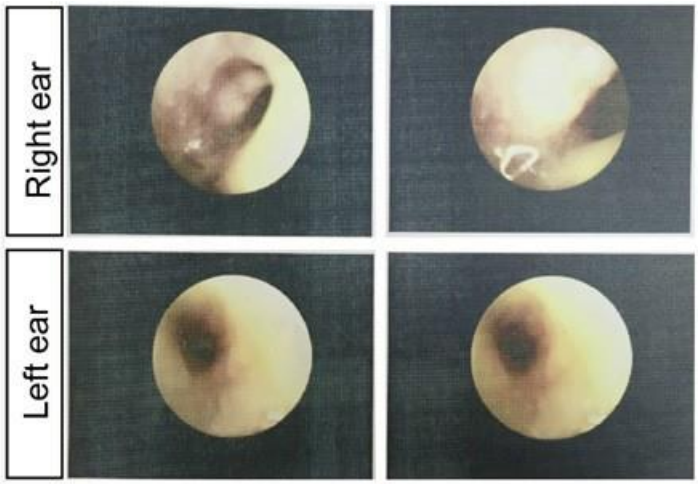
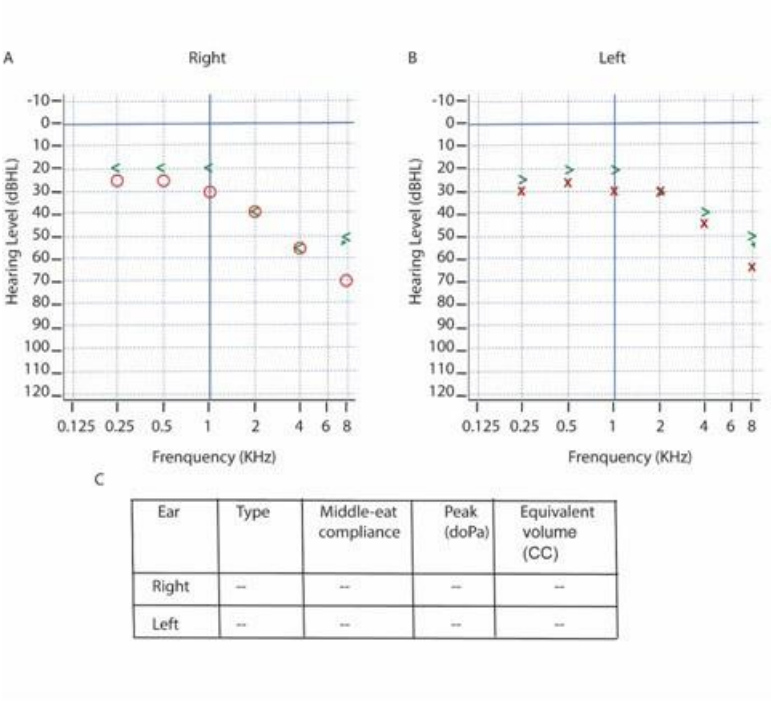


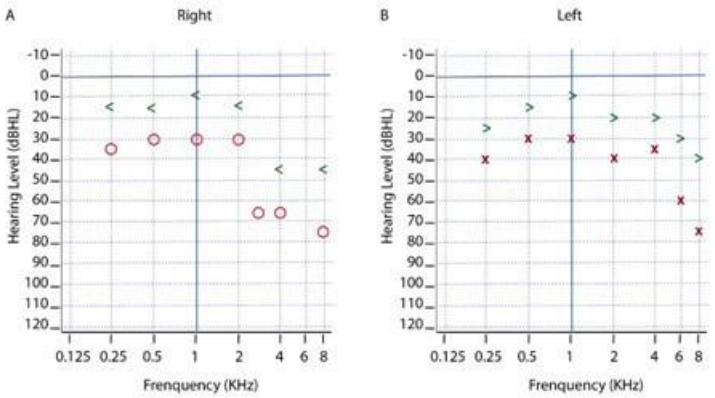
Figure3:ViewoftheOtoscopy. Externalauditorycanalandtympanicmembraneofrightandleftear.



C

Ear	Type	Middle-eat compliance	Peak (doPa)	Equivalent volume (CC)
Right	--	--	--	--
Left	--	--	--	--

Figure4:Audiometryatfirstvisit. （A,B）Puretoneaudiometry.(C)tympanometryfindings.<representsboneconduction(rightear);>represents bone conduction (left ear); ○ represents air conduction (right ear); × represents air conduction (left ear).



C

Ear	Type	Middle-eat compliance	Peak (doPa)	Equivalent volume (CC)
Right	--	--	--	--
Left	--	--	--	--

Figure 5:Audiometry at 1-year follow-up. (A, B）Pure tone audiometry. (C) tympanometry findings. < represents bone conduction (right ear); > represents bone conduction (left ear); ○ represents air conduction (right ear); × represents air conduction (left ear).

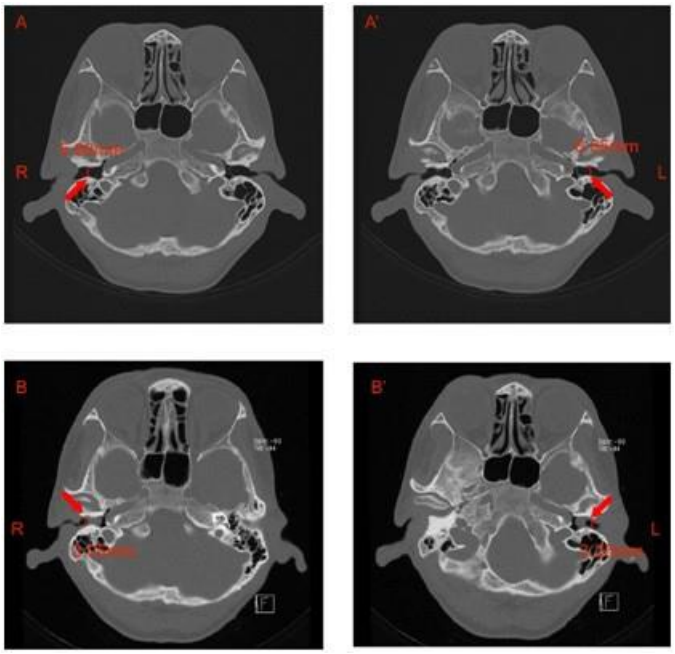


Figure 6: Axial Computed tomography scan of the temporal bone. (A, A') At first visit. (B, B') at 1-year follow-up. External auditory canal(Redarrow).Minimumanteroposteriordiameterofexternalauditory canal（redline）. R:right; L: left.

4. Discussion

The occurrence of KA in the ear is very rare. At present, only 2 casesofauricleKAhavebeenreported,bothofwhichareregional and well treated after surgical resection [3,4]. However, there is no report of GEKAin the ear and its related lesions.Therefore, it is the first case report of bilateral auricles lesion and stenosis and atresia of EAC associated with GEKA.

ManagementofAcquiredStenosisofEAC

The surgical techniques used in the management of acquired stenosisofEAChavevariedovertheyears[5,6].Intheearlyyears,it wassuggestedtowidenthebonycanalbyexcisionofthestenotic tissue.Adkins et al covered the skin-deficient canal with a transposition flap in eight cases, with no recurrence [7]. Moore et al lined the canal with a full thickness skin graft in one case that not recurrence [8]. McDonald et alused a split thickness skin graftin 22 cases, with two recurrence [9]. Bell used bilateral rotation skin flaps in 9 cases, with no recurrence [10]. McCary et al used split thickness grafts in 18 cases, with one recurrence [11]. How- ever, it seems that the use of skin flaps or grafts is not necessary in acquired stenosis, unlike acquired atresia. More recent studies havedemonstratedthatameatoplastyaloneissufficienttotreatacquiredstenosis.Oncetheanatomicalnarrowinghasbeencorrected by enlargement of the canal and excision of the thickened tissue in acquired stenosis, the condition of the ear is stable.This would suggest that the normal function of EAC is restored, enabling a normal cycle of ear cleaning and preventing poor canal patency leading to inflammatory episodes [12].

ManagementofAcquiredAtresiaofEAC

EAC atresia can be divided into congenital or acquired. Otitis externa is the most common cause of acquired EAC [13]. Males are generally more likely to be diagnosed with acquired EAC, with a male: female ratio of 2-3:113. In acquired atresia, patients'main complaint was hearing loss. Surgery was aimed at improving this deficit by restoring and maintaining the patency of the ear canal. Comparedwithsurgicaloutcomesforacquiredstenosis,thosefor acquired atresia were not good. In many cases, a hearing aid may beabetteralternative.Surgicaltechniquesforthemanagementof acquired atresia have evolved since 1966. All agree that removingthefibrousplugaloneisinadequate.Unlikeacquiredstenosis, thedenudedcanalwallshouldnotbeallowedtogranulate,asthis will lead to recurrence of the atresia; some form of canal lining is required. Different techniques have been used including transposition flaps, full thickness skin grafts and split skin grafts, but all used techniques have some degree of recurrence [13,14]. Regardless of technique used, recurrence has been seen at 6-month, 1-year,3-yearsand9-years.Thisdemonstratesthatacquiredatresia produces an instable ear canal, but whether this is secondary to the underlying disease process or due to the operative procedure, or a combination of both is still unclear. Long-term follow up is required.

In this case, erythematous papules and nodules are widely seenin the skin of the whole body, causing irreversible skin lesions, especially in exposed areas such as the face, and the failure rateof skin transplantation is high. For acquired stenosis and atresiaof EAC in the patient, the use of the above treatments may not be abletoachievebetterlong-termresults,andthepatient'sGEKAis still in progress, which brings more difficulties and challenges to our treatment.

New insights into the management of the acquired stenosis and atresia of the EAC

Nowadays,noneofthepublicationsfoundintheliteratureaddressesthepossibilityofanunderlyingsystemicetiologywhenmanag- ing this condition.Acquired atresia of the EAC is often regarded as a regional disorder and it is usually managed. However, when weconsiderthepossibilityofunderlyingsystemiccauses,thetheoreticalbasisthatsupportscurrenttreatmentswillchange,withthe goalofcorrectingunderlyingconditions,suchasimmune-mediateddiseasesorotherdiseases.Obviously,extensiveexaminationis notrequiredwhentheclinicalcauseisclear(traumatic,postoperative,recurrentotitismedia).Whenatumorissuspected,appropriateradiologicalandpathologicalexaminationsarerecommended. When the cause is unknown, we recommend a complete test and ANA screening. If the initial laboratory examination is tough, a biopsyfromtheEACisrequiredbeforesurgicaltreatment.Thepatient refuses ear biopsy, which is very regrettable for the accurate diagnosis and management of the disease [6].

5. Conclusion

Case of bilateral auricles lesion and stenosis and atresia of EAC associated with GEKAis very rare. But it provides a new insight into the etiological diagnosis of acquired stenosis and atresia of EA.Thesurgicalstrategyforthemanagementoftheacquiredstenosisandatresiaof theEACconsistsoftheexcisionofthefibrous plug,applicationofthecutaneousflapsand/ortransplantstocover the bare parts of the bone portion of the affected external canal. Eventhough,thestateoftheEACremainsinstable,anditsre-ste- nosis and re-atresia may occur.

6. Acknowledgments

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