

CASE REPORT

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# Synchronous bilateral idiopathic external auditory canal cholesteatoma: a case report and review of the literature

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## Abstract

**Background** External auditory canal cholesteatoma (EACC) is a rare pathological condition representing 0.1% of all new otologic cases. Bilaterality manifests in 10% of EACC cases. Similar disease processes include keratosis obturans (KO) and benign necrotizing otitis externa (BNOE). Diagnostic differentiation may not always be unexacting but does however influence management and eventual outcome. This study aims to briefly review the literature, describe an unusual case of synchronous bilateral primary EACC, and simplify the diagnostic challenges surrounding this disease.

**Case presentation** A 66-year-old man with no relevant family history, no concurrent or intercurrent illnesses, having undergone no relevant surgery, and had visited the Department of Otorhinolaryngology regarding right-sided otorrhoea and aural pruritis 9 years ago. At the time, he was diagnosed clinically with KO. Nine years later after having defaulted follow-up, he presented again with the same symptoms. Oto-microscopy now revealed excavation of the posterior canal wall, keratinous desquamation, and an intact tympanic membrane. Oto-endoscopy demonstrated extension posteriorly into the mastoid segment of the temporal bone. Computed tomography evinced a soft tissue attenuating mass within the excavated sub-adjacent bone, with extension into the mastoid air cell system. The contralateral ear had a smaller epithelial defect of the canal floor with underlying tympanic plate erosion. A closed mastoidectomy with the reconstruction of the posterior canal wall was performed on the right ear. The left ear was managed conservatively with micro-suctioning and aural toilette. Six months post-surgery, however, the reconstructed posterior canal wall underwent necrotic breakdown. The contralateral lesion gradually progressed into a Naim et al. (2005) Stage III EACC.

**Conclusion** This case illustrates the diagnostic challenge presented by EACC and is unusual regarding its bilaterality. The importance of diagnostic differentiation, both in the management of this disease and in the prevention of its progression, is highlighted. The idea of oto-endoscopy as a routine clinical tool is introduced, and endoscopic images are compared to high-definition computed tomography scans of the affected temporal bones.

**Keywords** Case report, External auditory canal, Cholesteatoma, Literature review

## Background

External auditory canal cholesteatoma (EACC) is a rare disease with an estimated incidence proportion amongst new otologic patients of 0.1–0.5% [1, 2], and an annual incidence rate amongst the general population of 0.15 cases per 100,000 individuals [2], compared to 9.2 per 100,000 for middle-ear cholesteatoma [3].

EACC was first described by Toynbee in 1850 [4], who investigated a lesion of the external auditory canal (EAC)

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which he considered to be molluscum contagiosum. Schofield introduced the term EACC in 1893 [5]. In 1943, Altmann and Waltner concluded that the EACC results from the invasion of a sequestered portion of the osseous EAC by keratinizing stratified squamous epithelium [6]. There remained confusion surrounding its relationship to other processes of the EAC, especially keratosis obturans (KO), until Piepergerdes et al. made the clinical distinction between the two in 1980 [7]. In 1984, Naim et al. demonstrated the histopathological differences between EACC and KO, the distinguishing feature of EACC being the focal invasion of necrotic bone by squamous epithelium, in comparison to the circumferential accumulation of desquamated keratin within a uniformly expanded EAC as seen in KO [8]. It is thus the focal nature of EACC that distinguishes it from KO [7]. For every case of EACC, there are four or five cases of KO [1].

### Histopathology

The histopathology of EACC is characterised by hyperproliferative keratinizing squamous epithelium with an intact basement membrane and increasing accumulation of stromal inflammatory cells (mast cells, lymphocytes, monocytes) [9]. Periosteitis accompanies higher grades of EACC and results in dull otalgia. Invasion of EACC into deeper mesenchymal tissue results in necrotic osseous excavation. Standard haematoxylin and eosin staining demonstrate sporadic epithelial cones, which are extensions of the basal cell layer into the subepithelial connective tissue [9]. This osteonecrosis and overlying focal loss of epithelium are pathognomonic of the diagnosis [10]. Osseous erosion is thought to be caused by proteolytic enzymes produced within the matrix lining. This may be hastened by superimposed infection with ulceration and reactive granulation tissue formation [11]. The keratin sac contains sequestered bony fragments, and random keratin is found within the EAC lumen [8, 12]. Further

extension may involve the middle ear, mastoid, sigmoid sinus, temporomandibular joint, or fallopian canal [1, 2, 13–15]. Albeit rare, posterior cranial fossa extension with resulting intracranial abscess has been reported [16].

Naim et al. sought to classify the different stages of EACC [17] based on the depth of invasion of the lesion (Table 1). The evolutionary continuum begins with hyperproliferation of the epithelium, which becomes inflamed and results in periostitis, eventually denuding the underlying bone. Denudation leads to necrobiosis and excavation, leaving an osseous defect which collects keratinous debris, and can ultimately involve neighbouring anatomical structures.

### Etiopathogenesis

The etiopathogenesis of EACC remains unclear [7–9, 18–20]. In keeping with earlier studies [6, 21], Sakamoto and Kitahara suggested that any disease of the osseous EAC could be a potential cause of EACC [22].

Holt divided EACC into five groups based on its etiopathogenesis. In order of prevalence, these are the following [13]: primary/spontaneous, postsurgical, post-traumatic, secondary to acquired EAC obstruction, and secondary to congenital EAC stenosis [23].

Although there have been cases reported in children and young adults [24, 25], primary/spontaneous EACC is generally regarded as a disease of the elderly. It results from a reduction in the migratory capacity of the EAC epithelium causing disruption to the self-cleansing mechanism and leading to retention of desquamated epithelium, i.e. ‘keratinization in situ’ [2, 13, 17, 26]. The accumulation of desquamated epithelium predisposes to inflammation, which in turn results in epithelial hyperproliferation [9]. Once this stage is established, active in-growth and bone resorption can occur [9, 23, 26].

Although recently challenged [27], this theory is supported by the findings of Makino and Amatsu, who

**Table 1** Naim et al. classification of EACC [17]

Stage I	Epithelial hyperplasia
Stage II	Hyperplastic epithelial inflammation with surrounding periostitis. Superimposed infection causes purulent otorrhoea
Subclass IIA	Intact epithelium
Subclass IIB	Epithelial defect with osseous denudation
Stage III	Necrobiotic excavation of the osseous EAC which accumulates keratin
Stage IV	Extension into adjacent anatomical structures
Subclass IVM	Mastoid
Subclass IVS	Skull base with involvement of the sigmoid sinus
Subclass IVJ	Temporomandibular joint
Subclass IVF	Facial nerve

EACC external auditory canal cholesteatoma, EAC external auditory canal

found that the epithelial migratory rate in a normal ear is highest along the inferior EAC, at approximately 142  $\mu\text{m}/\text{day}$  [1, 26]. These authors observed a markedly reduced migratory rate in this region of patients with EACC, likely attributable to a reduction in blood supply to this already tenuously perfused area [26]. In response to hypoxia, normal EAC skin may induce uncontrolled angiogenesis, predisposing to EACC formation [17, 26]. Hypoxic angiogenesis has been confirmed by studies of the angiogenic factors: hepatocyte growth factor/scatter factor (HGF/SF) and vascular endothelial growth factor (VEGF) and the tyrosine-kinase c-Met receptor, all of which are strongly expressed in specimens of EACC, induction of which results in an enhanced proliferation rate of these specimens [28, 29]. Adamczyk et al. also analysed the biological behaviour of EACC using the expression and distribution of the biomarkers transforming growth factor- $\alpha$  (TGF- $\alpha$ ), epidermal growth factor-receptor (EGFR), and the proliferation marker MIB 1, in comparison to that of normal EAC skin [9]. It has long been established that TGF- $\alpha$  stimulates the growth of several types of epithelial tissues [30]. Mediated by TGF- $\alpha$  binding, the EGFR controls these cellular activities [31]. The authors demonstrated overexpression of EGFR and TGF- $\alpha$  as well as higher mitotic activity in the EACC epithelium, evidenced by a significantly higher number of MIB-1-positive cells, reflecting a hyperproliferative state. These results suggest that a chronic inflammatory process underlies EACC, potentially altering keratinocyte proliferation [9].

Surgery and trauma to the EAC may cause entrapment of squamous epithelium during the healing process, or predispose to periostitis, with subsequent invasion and proliferation of the squamous epithelium [23, 24, 32–34].

In some cases, the occurrence of EACC has been linked to branchial arch anomalies, which result in the retention of epithelial masses [19, 35].

### Clinical features

A recent meta-analysis of published case series concluded that the most common presenting symptoms of idiopathic EACC are unilateral otorrhoea and mild to moderate otalgia [18]. The otorrhoea is thought to be related to an associated localised infection by a variety of organisms, most commonly *Pseudomonas aeruginosa* [13]. Bilaterality occurs in one-in-six to one-in-12 cases [1, 14], and synchronous bilateral cases have been reported [33].

Otoscopic findings include a localised area of epithelial erosion, an intact tympanic membrane, and a normal middle ear [9]. There is frequent osseous sequestration of the underlying EAC, and the defect is filled with keratin debris [9]. The majority of idiopathic EACCs occur along

the inferior EAC [18], and most patients present with a Naim et al. Stage II EACC [17].

Epidemiological risk factors for the development of EACC include smoking, diabetes mellitus, and repeated micro-trauma (e.g. from cotton buds or hearing aids) [18, 19]. Smoking and diabetes are thought to result in micro-angiopathic changes in the EAC that potentially impair epithelial migration [18].

### Radiographic features

Computed tomography (CT) of the temporal bones has become accepted as the gold standard for staging and preoperative planning in EACC [36]. Previous reports have shown that EACC is often more extensive than what is suggested by the clinical findings [37]. CT allows accurate evaluation of the extent of local bone erosion and examination for the involvement of adjacent landmark structures, namely the middle ear, tegmen tympani, mastoid, skull base, and fallopian canal. The involvement of these structures may determine management [13, 38].

Heilbrun et al. sought to define the CT features of EACC, as well as to determine which of these features impact its clinical management most prominently [13]. The authors concluded that the cardinal features of EACC are a soft tissue attenuating mass, most commonly involving the inferior-posterior portion of the EAC, with an erosion of the sub-adjacent bone [7, 37]. Bone fragments are often visible within the soft tissue mass as small, hyperattenuating flecks. The bone erosion adjacent to the soft tissue mass may be smooth, similar to a middle ear cholesteatoma, or irregular due to periostitis and osseous necrosis [13, 39].

Shin et al. classified EACC according to CT findings, with suggested management strategies for each stage of the disease (Table 2) [36].

### Management

The current management of EACC is highly variable between centres [38]. Holt suggested dividing the treatment options for EACC into conservative and surgical, based on the pathological extent of the disease [23]. Treatment aims are disease eradication, preservation of normal structure and function, and the restoration of normal epithelial migration and thus a self-cleaning EAC [38].

Conservative therapy, with frequent debridement of the keratin debris and sequestered bone, is favoured in early-stage disease [18, 40]. Topical treatment with antibiotic and corticosteroid-impregnated gauze [41] or drops [1] may aid in expediting disease control. For refractory or more advanced cases (Naim et al. Stages II–IV) [17], surgical intervention is necessary [1, 7, 14, 38].

**Table 2** Shin et al. classification of EACC [36]

Stage	CT findings	Treatment
Stage I	Limited to the EAC	Local care or canaloplasty
Stage II	Extension into the middle ear	Canaloplasty + tympanoplasty
Stage III	Extension into the mastoid air cell system	Canaloplasty + mastoidectomy $\pm$ tympanoplasty $\pm$ reconstruction of the EAC wall
Stage IV	Extension beyond the temporal bone	Removal of cholesteatoma via various approaches

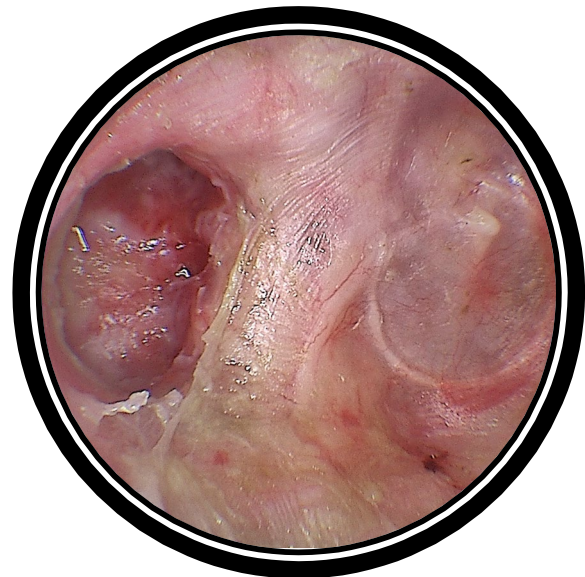
EACC external auditory canal cholesteatoma, CT computed tomography, EAC external auditory canal

If the lesion does not extend into the mastoid air cell system, either a per-meatal or postauricular approach may be used. The postauricular approach is favoured for more adequate cholesteatoma extirpation, necrotic bone debridement, canaloplasty, and fascia/perichondrium/cartilage grafting to cover the canal defect [8, 17, 37, 42]. Garin et al. also suggested considering a split-thickness skin graft to cover the defect [37]. Tos recommended an inferiorly based subcutis-muscle-periosteum flap, harvested along the posterior edge of the postauricular incision, to obliterate the defect [41]. Jahnke and Lieberum suggested that the decision to place a graft must be based on the surgeon's satisfaction that no epithelial fragments remain obscured in the osseous defect, lest these provoke recidivism [43].

### Case presentation

A 66-year-old man with no relevant family history, no concurrent or intercurrent illnesses, having undergone no relevant surgery, and visited the Department of Otorhinolaryngology regarding right-sided otorrhoea and aural pruritis 9 years ago. He was lost to follow-up after micro-debridement of what was clinically described at the time as KO. He now presented, 9 years later, with the same symptoms. Oto-microscopy of the right ear revealed irregular keratinous desquamation. Aural toiletting uncovered a granulomatous excavation of the posterior EAC containing keratin and sequestered osseous fragments, against the backdrop of a normal tympanic membrane. Oto-endoscopy demonstrated extension posteriorly into the mastoid segment of the tympanic bone (Fig. 1).

Contralateral otoscopy disclosed a hyperkeratotic lesion of the inferior EAC. Computed tomography (CT) evinced soft tissue attenuation containing hyper-attenuating flecks within an excavated right-sided temporal bone. This excavation extended into the mastoid air cell system, making it a Stage III EACC as classified by Shin et al. [36]. The contralateral ear enclosed early tympanic plate erosion, making it a Stage I EACC according to the classification of Shin et al. [36]. The CT scan thus bespoke bilateral EACC [13], consistent with the suggestion by



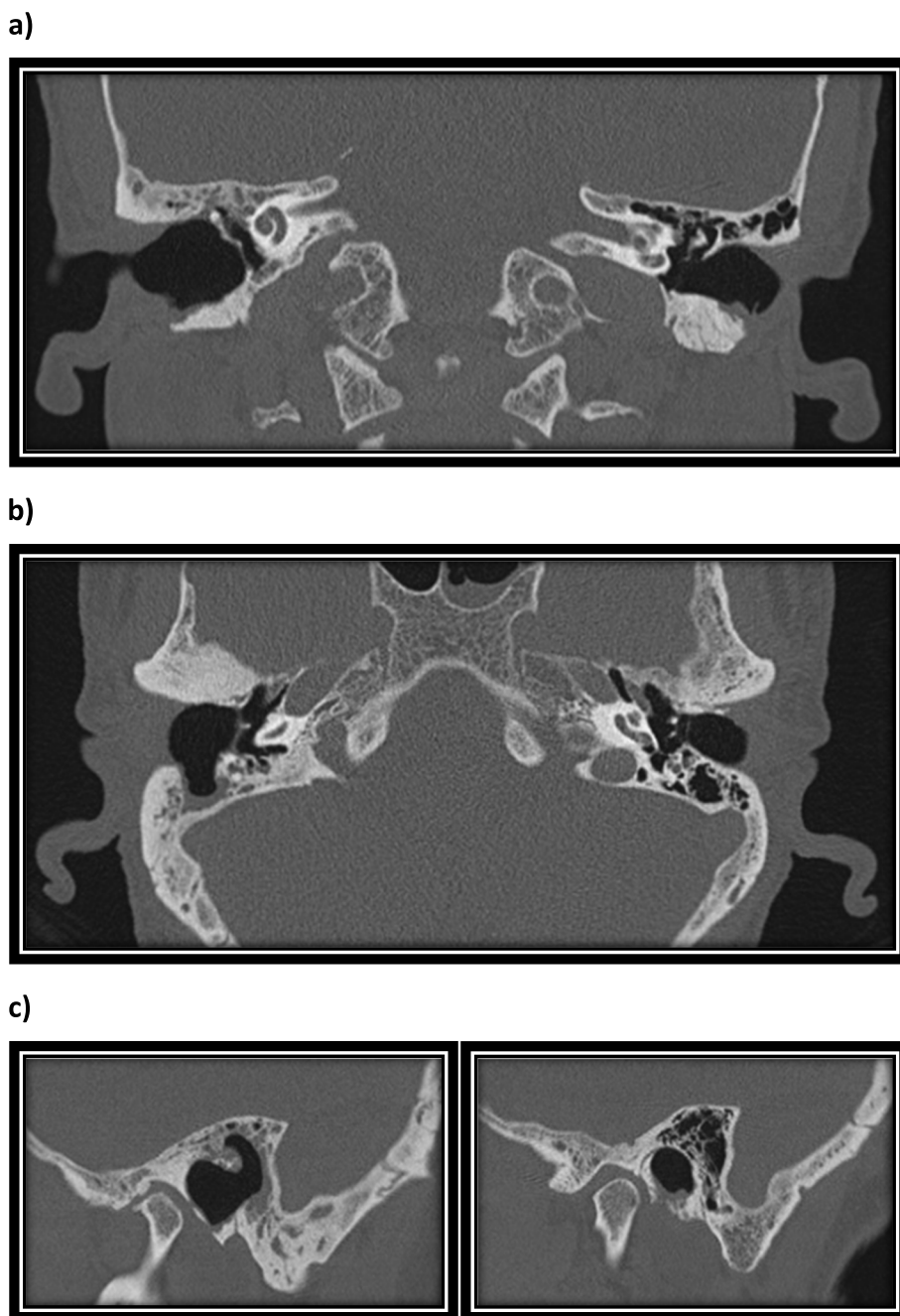
**Fig. 1** Oto-endoscopic image of the right external auditory canal cholesteatoma

Holt (1992) that most of these lesions are idiopathic and therefore discovered incidentally (Fig. 2) [23].

Pure-tone audiometry showed an asymmetrical, bilateral, and pantonal sensorineural hearing loss. The right ear presented a moderate and the left ear a severe loss. This demonstrated poor correlation with disease severity between the respective ears. As such, it was decided that the hearing loss was most likely presbycusis, rather than being directly related to EACC.

A canaloplasty and canal wall-up mastoidectomy of the right ear were performed, as advocated by Shin, Shim, and Lee (2010) [36]. A retro-auricular incision was used for the surgical approach, which allowed harvesting of temporalis muscle fascia as well as cartilage of the auricular concha via the same incision. The tympanic membrane, being uninvolved by the disease process, was not operated upon. All squamous epithelium was removed from the EAC and mastoid air cell system, and the defect created by the disease was drilled smooth using a diamond burr, being careful not to spuriously open the mastoid segment of the fallopian





**a) Coronal, b) Axial and c) Parasagittal Computed Tomography Images**

**Fig. 2** Computed tomographic images of synchronous bilateral external auditory canal cholesteatomas

canal of the facial nerve. The resultant cavity, which now connected the EAC and the mastoid air cell system, was irrigated with normal saline so as to remove all remaining squamous epithelial cells. The posterior wall of the EAC was then grafted with cartilage and fascia, the harvesting of which has been described above,

in accordance with the recommendations of a variety of authors (Fig. 3) [8, 29, 37].

In anticipation of disease control, the left ear was managed conservatively with regular debridement and topical antibiotics, congruent with the suggestions of Tos (1997) [41], as well as those of a recent



**Fig. 3** Intraoperative oto-microscopic image of the right external auditory canal cholesteatoma

meta-analysis by Dubach, Mantokoudis, and Caversaccio (2010) [18].

Six months post-surgery, however, the fascial graft underwent necrotic breakdown. The resultant defect was small and was allowed to heal by primary intention. The contralateral lesion gradually progressed into a Naim et al. (2005) Stage III EACC (Fig. 4) [29].

**Discussion**

This case illustrates the diagnostic challenge presented by EACC and is unusual regarding its synchronous bilateralism [1, 14, 33].

The hallmark of EACC is focal osteo-necrotic epithelial invasion, in comparison to KO which features circumferential, geometrically lamellar keratinous accumulation within a uniformly expanded EAC. Differentiation between these diseases influences both management and outcome. The significance of early diagnostic differentiation is emphasised, considering EACC’s

indolent yet erosive nature which allows unperceived extension beyond the EAC [38]. Expansive visualisation, as is afforded by endoscopy, would thus facilitate timeous diagnosis.

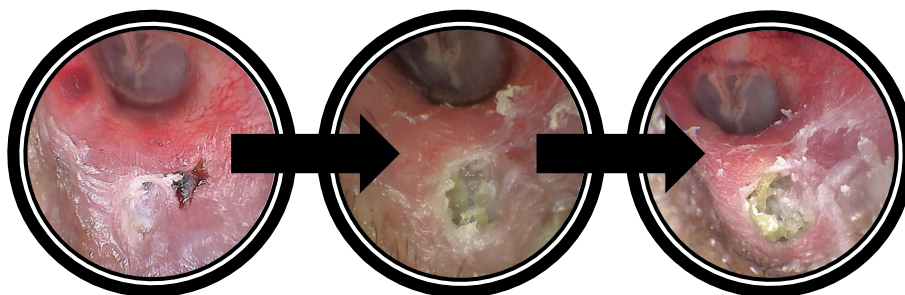
Computed tomography is the accepted gold standard for staging and surgical planning regarding EACC [36]. Current management, however, remains variable between centres [38]. We believe that oto-endoscopy may aid with decision-making in this regard.

We speculate that the treatment-refractory nature of the lesions in this report may be a feature of their position within the EAC. Makino and Amatsu (1986) demonstrated that the epithelial migratory rate, normally highest along the inferior EAC (142 µm/day), diminishes most markedly in this region with age [26]. Migratory curtailment, predisposing to recalcitrant lesions, could result from ischemia to an already tenuously perfused area. Such conditions would prompt secondary hypoxemic angiogenesis, altering keratinocyte proliferation in favour of EACC formation, as suggested by Adamczyk, Sudhoff and Jahnke (2003) [9], and Naim et al. (2004; 2005) [28, 29] in their research into the biomolecular behaviour of EACC.

Another possible explanation for surgical failure is that of incomplete disease extirpation. Jahnke and Lieberum (1995) suggested that the decision to graft should be based on the surgeon’s satisfaction that there remain no occult epithelial fragments, lest these provoke recidivism [43].

**Conclusion**

This case illustrates aberrations in the clinical presentation and treatment response of EACC. Attention is drawn to the significance of accurate diagnosis with regard to appropriate disease management. Garin, Degols, and Delos (1997) demonstrated that EACC is often more extensive than what is suggested clinically [37]. We, therefore, emphasise the value of combining visualisation techniques towards an enhanced understanding of disease idiosyncrasies.



**Fig. 4** Oto-endoscopic images of the left external auditory canal cholesteatoma demonstrating its evolution over time

**Abbreviations**

EACC	External auditory canal cholesteatoma
EAC	External auditory canal
KO	Keratinosis obturans
HGF/SF	Hepatocyte growth factor/scatter factor
VEGF	Vascular endothelial growth factor
TGF- $\alpha$	Transforming growth factor- $\alpha$
EGFR	Epidermal growth factor-receptor
MIB1	MIB1 gene encoded E3 ubiquitin-protein ligase MIB1 enzyme
CT	Computed tomography

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**Authors' contributions**

GJK was responsible for researching and writing the case report, while CF oversaw said research and writing. All authors read and approved the final manuscript.

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**Availability of data and materials**

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**Declarations****Ethics approval and consent to participate**

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**Consent for publication**

Written consent for publication was obtained from the patient in question.

**Competing interests**

The authors declare that they have no competing interests.

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