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Nasal sinuses cholesteatoma: case series and review of the English literature

Ahmed Nabil Abdelhamid Ahmed[1] · **Mohamed M. Elsharnouby**[1] · **Marwa Mohamed Elbegermy**[1]

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Abstract

Background Nasal sinus cholesteatomas are uncommon slow-growing lesions that are frequently misdiagnosed preoperatively. They can develop due to embryologic remnants or iatrogenic factors (surgical trauma or nasal sinus trauma). In addition, they can cause bone destruction resulting in intracranial or intraorbital complications as well as malignant change if neglected. Complete surgical removal is a must with strict postoperative follow-up.

Materials and methods Three cases of nasal sinus cholesteatoma are reported. The first case was found inside the ethmoidal sinus, the second in the frontal sinus, and the third was found inside a concha bullosa. In all three cases, a wide endoscopic surgical excision was performed. Due to the lateral extension of the lesion, frontal sinus trephine was also used in the case of frontal sinus nasal cholesteatoma. In addition, a review of the English literature for the reported cases of nasal sinus cholesteatomas was conducted.

Results There were no reported recurrence or residual during strict postoperative follow-up for 2 years (by endoscopic examination and diffusion-weighted MRI with delayed postcontrast T1 images). A review of the English literature revealed 42 cases of nasal sinuses cholesteatomas (including the present three cases) (17 in the frontal sinus, 15 in the maxillary sinus, 5 in the ethmoid sinus, 3 in the sphenoid sinus, and 2 in a concha bullosa).

Conclusions Although nasal sinus cholesteatomas are uncommon, they must be considered in the differential diagnosis of slow-growing nasal sinuses lesions. Preoperative CT scan and diffusion-weighted MRI are essential for proper diagnosis and to exclude other similar lesions, such as nasal sinus mucoceles, cholesterol granuloma, or neoplastic lesions. Wide complete surgical excision is necessary to avoid recurrence and facilitate postoperative follow-up. As with ear cholesteatoma, strict postoperative follow-up is required to detect recurrence or residual early and is performed by endoscopic examination, diffusion-weighted MRI, and delayed post-gadolinium T1 images.

Keywords Sinus cholesteatoma · Sinus epidermoid · Sinus keratoma · Sinus mucocele · Nasal sinus neoplasms · Diffusion-weighted imaging

Introduction

Cholesteatoma is a common middle ear pathology found in many parts of the temporal bone; however, it is rarely reported in the nasal sinuses [1]. Only a few dozens of cases have been documented in the English literature in all nasal sinuses (mainly the frontal sinus) [2, 3]. Like temporal bone cholesteatoma, nasal sinus cholesteatoma can be congenital or acquired [4–8]. Complete wide local excision is used to treat sinus cholesteatoma, along with clinical and radiological follow-up to detect recurrence or residual [9–14]. In addition to facial disfigurement, nasal sinus cholesteatoma can result in intracranial or intraorbital complications due to bone erosion [15]. Besides, malignant change has been reported in some cases [16–18]. In this study, we presented three cases of nasal sinus cholesteatoma inside the frontal sinus, ethmoid sinus, and a concha bullosa. Furthermore, we reviewed the English literature on cholesteatoma of the nasal sinuses.

✉ Ahmed Nabil Abdelhamid Ahmed
ahnabilent@gmail.com

[1] Department of Otorhinolaryngology, Faculty of Medicine, Ain Shams University, Cairo 11331, Egypt

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Case 1: ethmoidal sinus cholesteatoma

A 17-year-old female patient presented to the outpatient clinic with an accidentally discovered swelling protruding at the medial canthus of the right orbit for 1 month. She also had a headache between the eyes and allergic rhinosinusitis symptoms (morning itching, sneezing, and nasal discharge for the last 4 years). There was no history of previous nasal surgery or head trauma. A defect on the right medial orbital wall and posterior part of the right nasal bone was discovered during the examination due to a medial palpebral cystic swelling. Nasal endoscopy revealed a deviated nasal septum to the left with the right concha bullosa.

Radiologic findings

The CT scan revealed a right ethmoidal expansile soft tissue lesion that eroded the lamina papyracea, the right nasal bone, right lacrimal bone, and the right frontal process of maxillary bone with a right concha bullosa (Fig. 1). Due to her claustrophobia, the patient refused to undergo a preoperative MRI. The lesion was misdiagnosed radiologically as an ethmoid sinus mucocele.

Operative details

- After right conchoplasty, endoscopic opening of the ethmoidal cystic swelling revealed keratin layers filling the cyst and lining the inner endothelium, which were eradicated.
- The cyst wall was removed completely.
- The lamina papyracea was found to be dehiscent, being compressed by the cyst wall, and the cyst was found to expand inside the frontal sinus recess (Fig. 2).
- Cholesteatoma was confirmed after postoperative histopathological examination which revealed a cyst lined by stratified squamous epithelium with a fibrous tissue wall filled with keratin and a subepithelial tissue showing a mixed inflammatory cellular infiltrate.

Case 2: frontal sinus cholesteatoma

A 35-year-old female patient presented with a right frontal headache (of gradual onset and progressive course) that began 3 years ago. She had a history of previous external right frontal sinus surgery in another hospital (for mucocele evacuation 23 months before being presented to our clinic), with no available medical records. A previous right medial infra-brow incision scar was discovered during the clinical

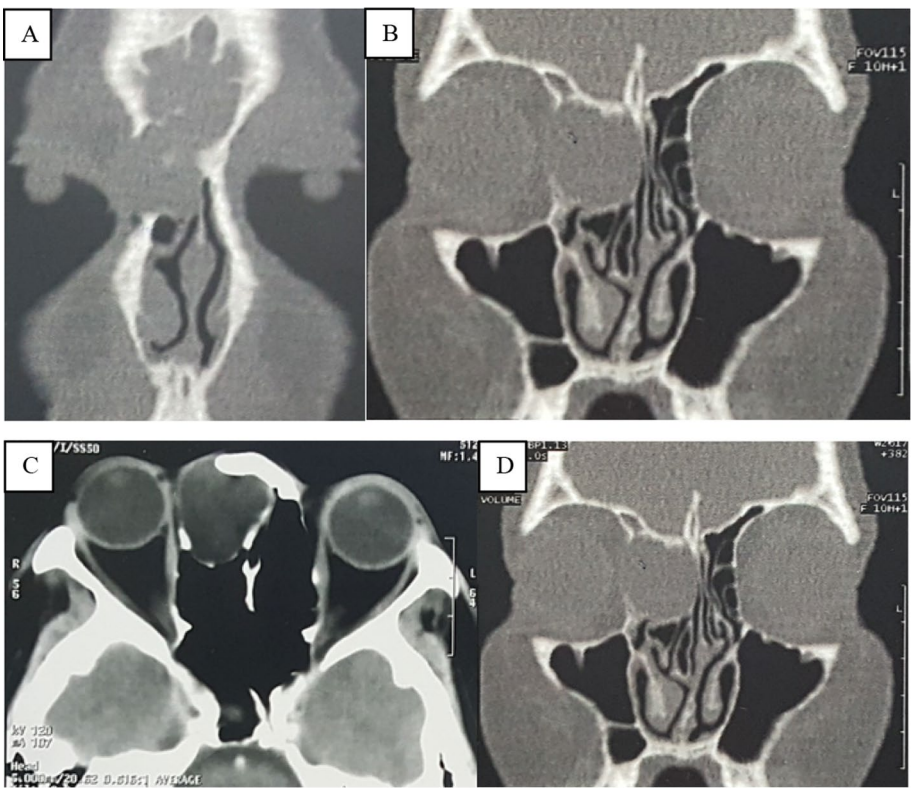

Fig. 1 CT scan coronal (**A**, **B**, **D**) and axial (**C**) cuts, showing a right ethmoid sinus soft tissue expansile swelling with erosion of the right frontal process of maxillary bone (**A**), lamina papyracea (**B**), and right nasal bone (**C**). Right concha bullosa with pneumatized uncinate process (**D**)

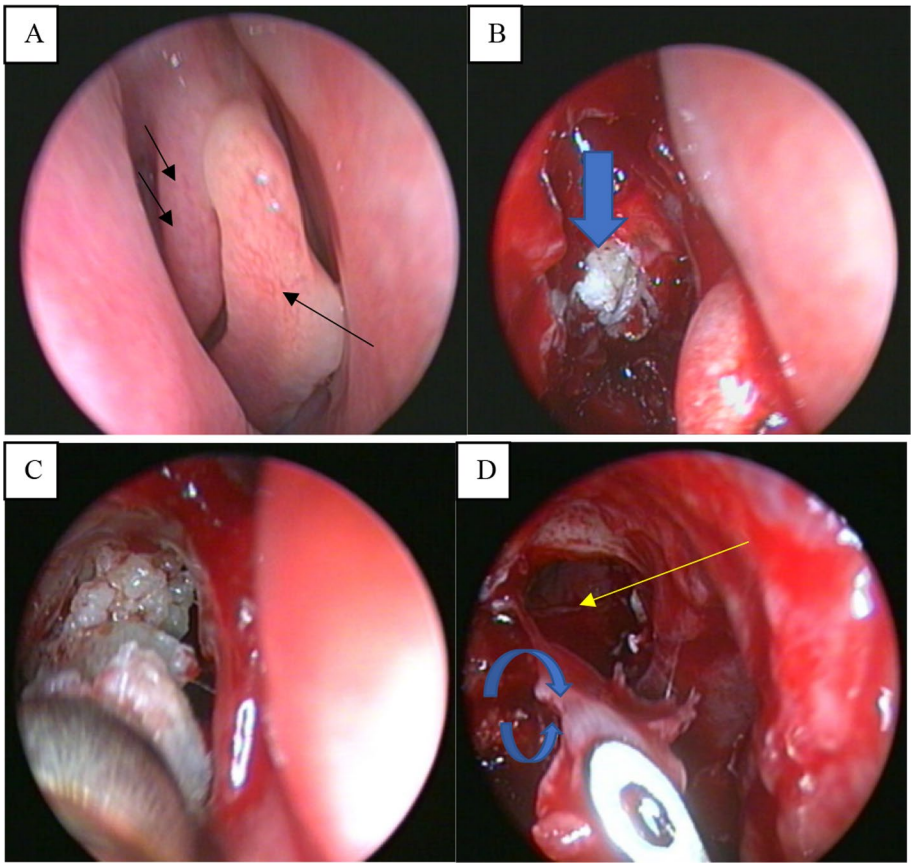

Fig. 2 Intraoperative endoscopic view, showing right concha bullosa (one black arrow) and pneumatized uncinate process (two black arrows) (**A**). The anterior wall of the cyst was opened after right conchoplasty with keratin layers seen inside (thick blue arrow) (**B**). Complete removal of the keratin layers by suction (**C**). Complete removal of the cyst wall keratinized epithelium and underlying thin ethmoidal bone (two curved blue arrows). Roof of the ethmoid sinus is seen (yellow arrow) (**D**)

examination. Endoscopic examination of the nasal cavity was irrelevant. No history of sinusitis or visual problems.

Radiologic findings

A CT scan revealed a soft tissue filling the right frontal sinus with bony fragments inside, erosion of the right orbital roof, and intact anterior and posterior tables of the frontal sinus (apart from a small erosion at the medial part of the posterior table) (Fig. 3). MRI showed a soft tissue filling the right frontal sinus with the erosion of the frontal sinus floor (hypointense in T1 and hyperintense in T2, while T1 with contrast demonstrated heterogenous opacity with enhancement in some areas due to inflammation) (Fig. 4). Preoperative differential diagnosis was sinus mucocele, pyocele due to associated inflammation in the MRI with contrast, and sinus cholesteatoma.

Operative details

- Endoscopic frontal sinusotomy revealed keratin layers filling the right frontal sinus. Due to the lateral extension of the cholesteatoma sac in the frontal sinus recess, a frontal sinus trephine was performed (at the site of previous infra-brow scar), revealing keratin layers filling the entire right frontal sinus. Frontal sinus trephine was the portal for angled endoscopes and instruments**.** The cholesteatoma sac contents and wall were removed, while the frontal sinus ostium was cleaned, being extremely cautious at the right orbital roof, which was found to be dehiscent. During removal of the keratinized mucosal sac wall, a minor defect in the medial aspect of the frontal sinus posterior table (about 3 mm in diameter) was found with CSF leakage. The defect was repaired with an underlay dumbbell fat graft (from the ear lobule) and an overlay septal perichondrium fixed with an absorbable gelatin sponge (GELFOAM®) (Fig. 5). A silastic stent (removed after 6 weeks) was inserted after Draf IIb to prevent stenosis of the frontal sinus ostium during postsurgical follow-up. The frontal sinus recess was found clean of any keratin debris.
- The pathology report confirmed the diagnosis of cholesteatoma with pieces of bone trabeculae.

Since medical records of previous nasal surgical intervention were unavailable, we were not sure whether this frontal sinus cholesteatoma is iatrogenic due to implantation of skin during previous surgery or congenital.

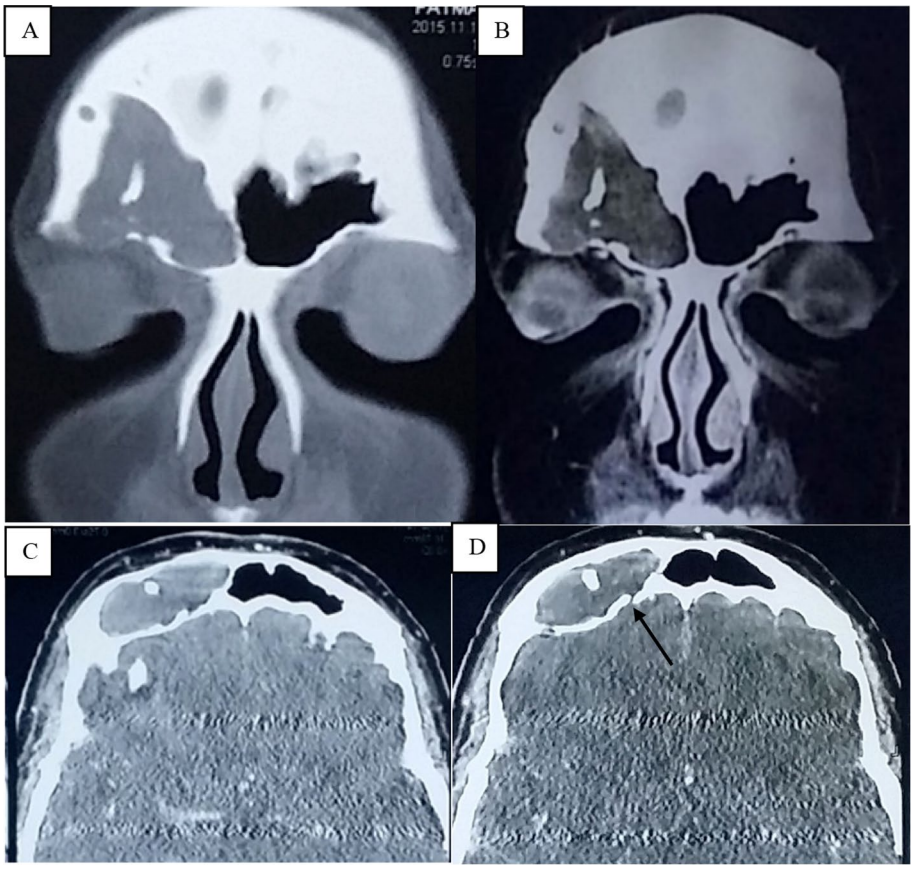

Fig. 3 CT scan, bone (**A**) and soft tissue (**B**–**D**) windows, showing a soft tissue filling the right frontal sinus with bone fragment inside and dehiscence of the roof of the right orbit. A small erosion (black arrow) at the posterior table of the right frontal sinus is seen (**D**)

Case 3: cholesteatoma of a concha bullosa

A 24-year-old female presented with unilateral left nasal obstruction of 4-year duration and left external swelling (below the medial canthus of the left eye) of 1-year duration. On examination, a bony defect was felt over the left nasal bone and the left frontal process of maxilla. Endoscopically, there was a boggy swelling of the left middle turbinate with intact mucosa. No history of previous head trauma or nasal surgery.

Radiologic findings

CT scan revealed a homogenous soft tissue filling the left concha bullosa (surrounded by a thin bony shell). The soft tissue swelling reached the roof of the ethmoid sinus, left frontal recess, and left frontal sinus (with dehiscent lamina papyracea, lacrimal bone, and the left frontal process of the maxillary bone), with no erosion of the skull base (Fig. 6). The MRI revealed a soft tissue cystic swelling (hyperintense in T2, hypointense in T1 and T1 with contrast) with the erosion of the lamina papyracea (Fig. 7). Preoperatively, it was misdiagnosed as a mucocele.

Operative details

- Incision of the anterior end of the cyst revealed keratin debris filling the cyst (Fig. 8). The cyst's medial and Lateral walls were removed, paying particular attention to the dehiscent lamina papyracea, nasolacrimal duct, and sac—no erosion of the ethmoid roof. Keratin was removed with complete removal of the cyst lining. Erosion of the frontal process of the maxilla, lacrimal bone, and nasal bone was found (Fig. 9).
- Histopathologic examination showed fragments of keratinized squamous cell epithelium lining a cyst wall with submucosal nonspecific mononuclear inflammatory cell infiltrate (epidermoid cyst), with no malignancy.
- The patient asked for the cosmetic reconstruction of the nasal bone defect, which was delayed for 2 years to exclude recurrence.

Materials and methods

We did review of the English literature for published cases of nasal sinuses cholesteatoma.

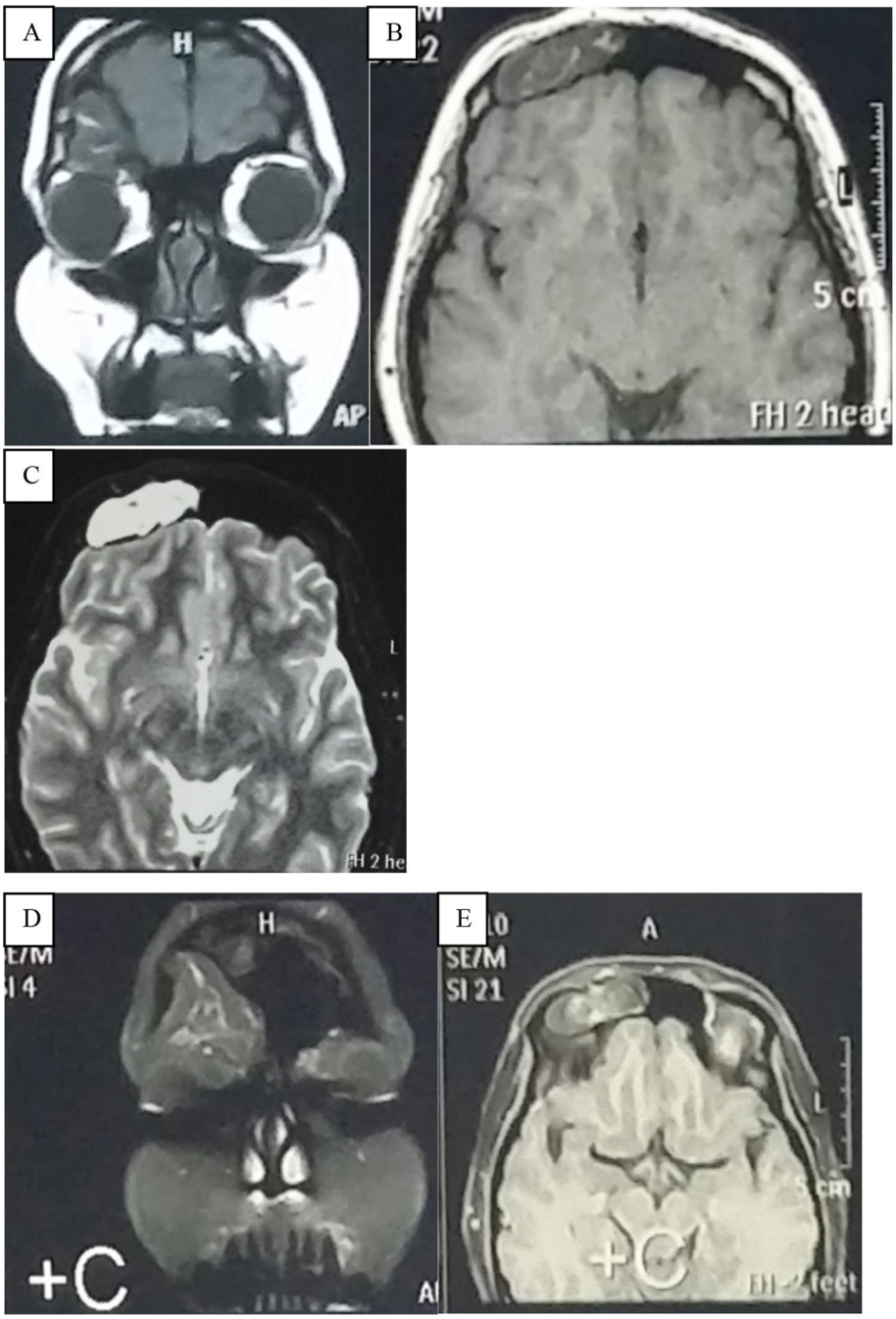

Fig. 4 MRI T1 without contrast coronal (**A**) and axial (**B**) cuts showing a hypointense mass in the right frontal sinus. MRI T2 axial cut (**C**) showing a hyperintense lesion in the right frontal sinus. MRI T1 with gadolinium axial (**D**) and coronal (**E**) cuts showing heterogenous opacity filling the right frontal sinus

Inclusion criteria

Our search was confined to articles

- Published in the midline database: PubMed (https://www.ncbi.nlm.nih.gov/pubmed/).
- In English language.
- Cholesteatomas' primary site was the nasal sinuses.
- Keywords used were cholesteatoma, epidermoid, and keratoma of the (frontal sinus, maxillary sinus, ethmoid sinus, sphenoid sinus, and concha bullosa).

Exclusion criteria

- Articles not in the English language or an undetermined language.
- Cholesteatomas that extend to nasal sinuses from nearby structures such as petrous apex cholesteatoma extending to the sphenoid sinus; intra-diploic cholesteatoma rupturing into the frontal sinus; orbital cholesteatomas involving the frontal or the maxillary sinus, and pterygopalatine fossa cholesteatoma extending to the maxillary sinus [14, 19].

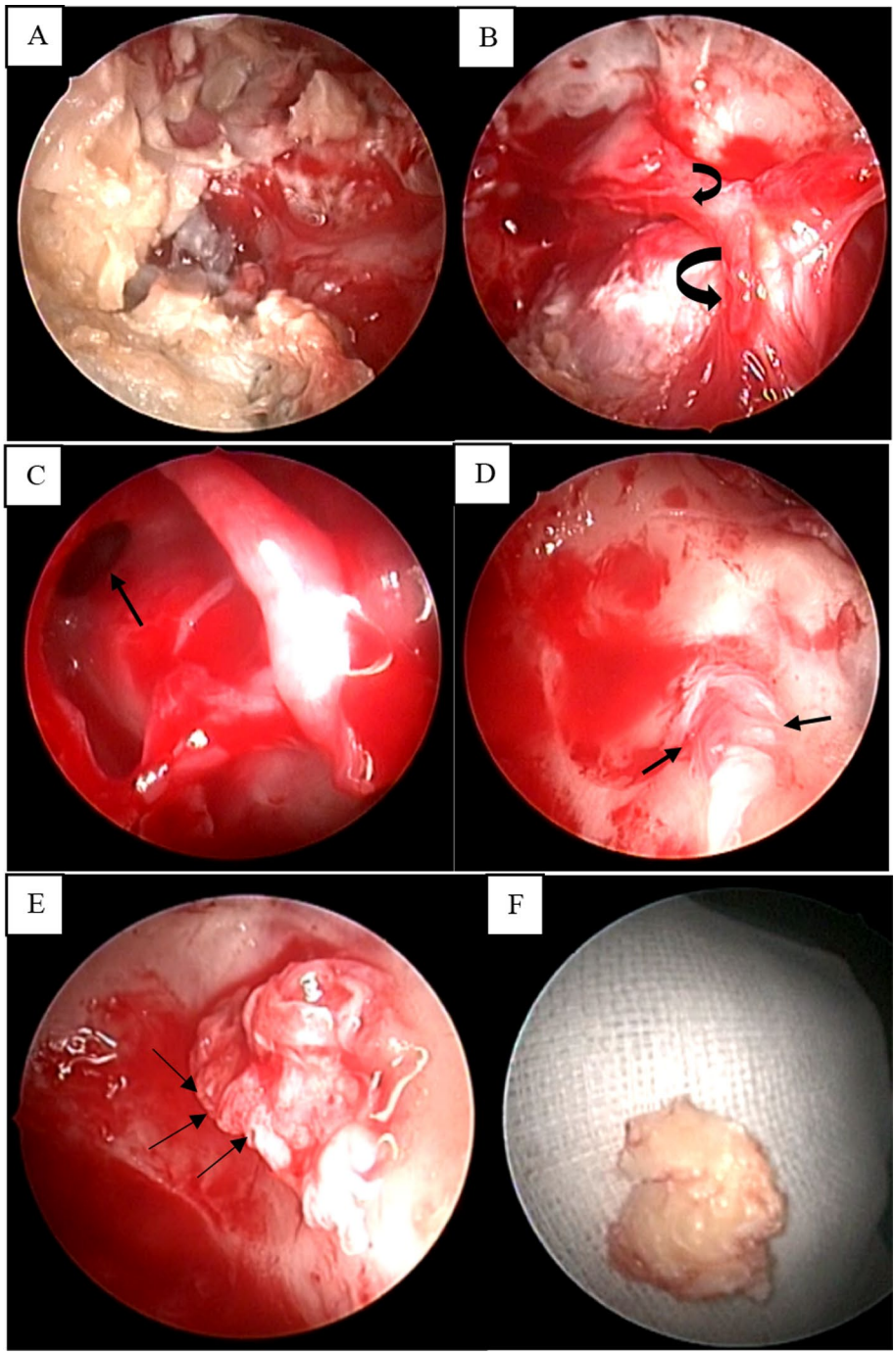

Fig. 5 Intraoperative endoscopic view from the frontal sinus trephine showing cholesteatoma sac with keratin debris filling the whole right frontal sinus (**A**). Endoscopic removal of the wall of cholesteatoma sac keratinized epithelium (two curved arrows) (**B**) and identification of the frontal sinus ostium (one arrow) (**C**). CSF leak from a defect (two straight arrows) in the posterior wall of the frontal sinus (**D**). Underlay dumbbell fat graft and overlay septal perichondrium (three arrows) repairing the defect found in the posterior table of the frontal sinus (**E**). Keratin layers sent for histopathology (**F**)

Results

There are 49 articles about frontal sinus cholesteatoma. After excluding articles not written in English and lesions originating from outside the frontal sinus, we had 14 articles (17 cases, including the present case). We collected data on (age, gender, history of previous head trauma or sinus surgery, and malignant change) (Table 1). Regarding ethmoid sinus cholesteatoma, the search yielded 16 articles. After applying exclusion criteria, we had four articles (5 cases, including the present case).

With respect to the maxillary sinus cholesteatoma, the search yielded 55 articles. After applying exclusion criteria, we get 15 articles (15 cases with a case of maxillary and ethmoidal sinus cholesteatoma after cleft palate repair) [33]. While the sphenoid sinus cholesteatoma was reported in three articles (3 cases), and concha bullosa cholesteatoma was reported in two articles (two cases only including the present case).

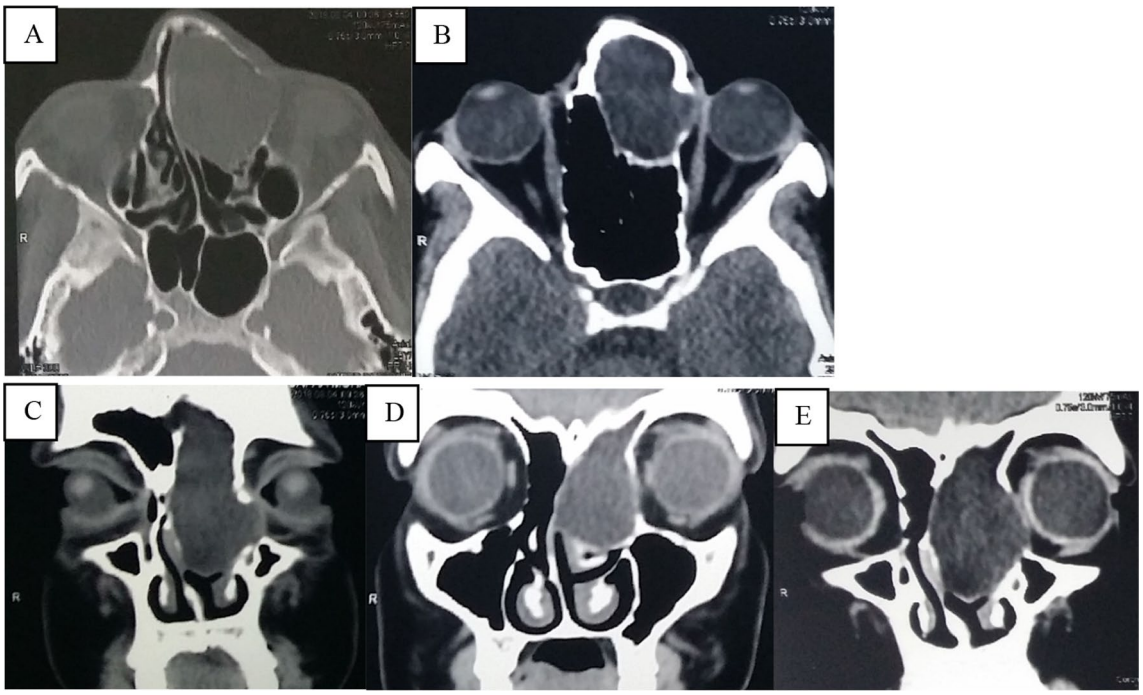

Fig. 6 CT scan (soft tissue and bone windows) axial (**A**, **B**) and coronal (**C**–**E**) cuts showing left expansile soft tissue swelling occupying the left concha bullosa with extension to the left frontal recess and left frontal sinus, but no erosion of the skull base. Nasal septum is seen pushed to the right

Therefore, a total number of 42 cases (including the present three cases) were reported; 24 males (57%) and 18 females (43%); consequently, the lesion was more common in males. The youngest patient was one and half years and suffered from congenital maxillary sinus cholesteatoma [31]. Meanwhile, the oldest patient was 82 years with sphenoid sinus cholesteatoma, with a mean age of 42.5 years [40]. Four cases developed intracranial complications secondary to frontal and sphenoidal sinus cholesteatoma [11, 15, 39, 40]. In addition, three cases of frontal sinus cholesteatoma developed malignant tumours [16–18].

Discussion

Cholesteatoma is a common middle ear pathology but rarely encountered in the paranasal sinuses. The first frontal sinus cholesteatoma was first reported in the English literature by Spencer in 1930 [1]. In the English literature, only a few dozen of cases were reported. It primarily affects the frontal sinus and, to a lesser extent, the ethmoid and, finally, the maxillary, sphenoid sinuses, and concha bullosa. Hence, it has been reported in all nasal sinuses [2, 8]. Paranasal cholesteatomas presentation varies according to the lesion's anatomical location and growth pattern [3].

There are four theories explaining the pathogenesis of cholesteatoma of the paranasal sinuses. The first is the congenital theory (the most accepted theory), suggesting that the cholesteatoma's origin is due to remnants of ectodermal epithelial cells during face formation in the third to the fifth week of gestation [4], while acquired cholesteatoma can be explained by three reasons. The first one is the implantation theory due to implanted epithelial cells after surgery or trauma [5, 6]. Second, it may be due to the migration of the squamous epithelium to a non-squamous epithelium-lined area. However, this migration should come from the nasal vestibule with a connecting tract of the epidermis, which has not been reported before [7, 8]. Third, epithelial metaplasia after long-term irritation as in chronic sinusitis, although many authors reject this theory as metaplasia due to chronic rhinosinusitis results in the non-keratinizing squamous epithelium and not keratinizing squamous epithelium [3, 5, 9].

Due to nonspecific clinical presentation and imaging findings, preoperative clinical diagnosis is typically difficult. They are usually slow-growing lesions that can remain asymptomatic for a long duration with gradual keratin accumulation and destruction of the surrounding tissues and bone. Frontal or ethmoid sinus cholesteatoma may cause painless frontal or orbital swelling, headache, intraorbital complications (proptosis, diplopia, blurred vision, eyelid edema, decreased visual acuity) and intracranial complications (meningitis, cerebrospinal fluid leakage, brain abscess) [7, 24].

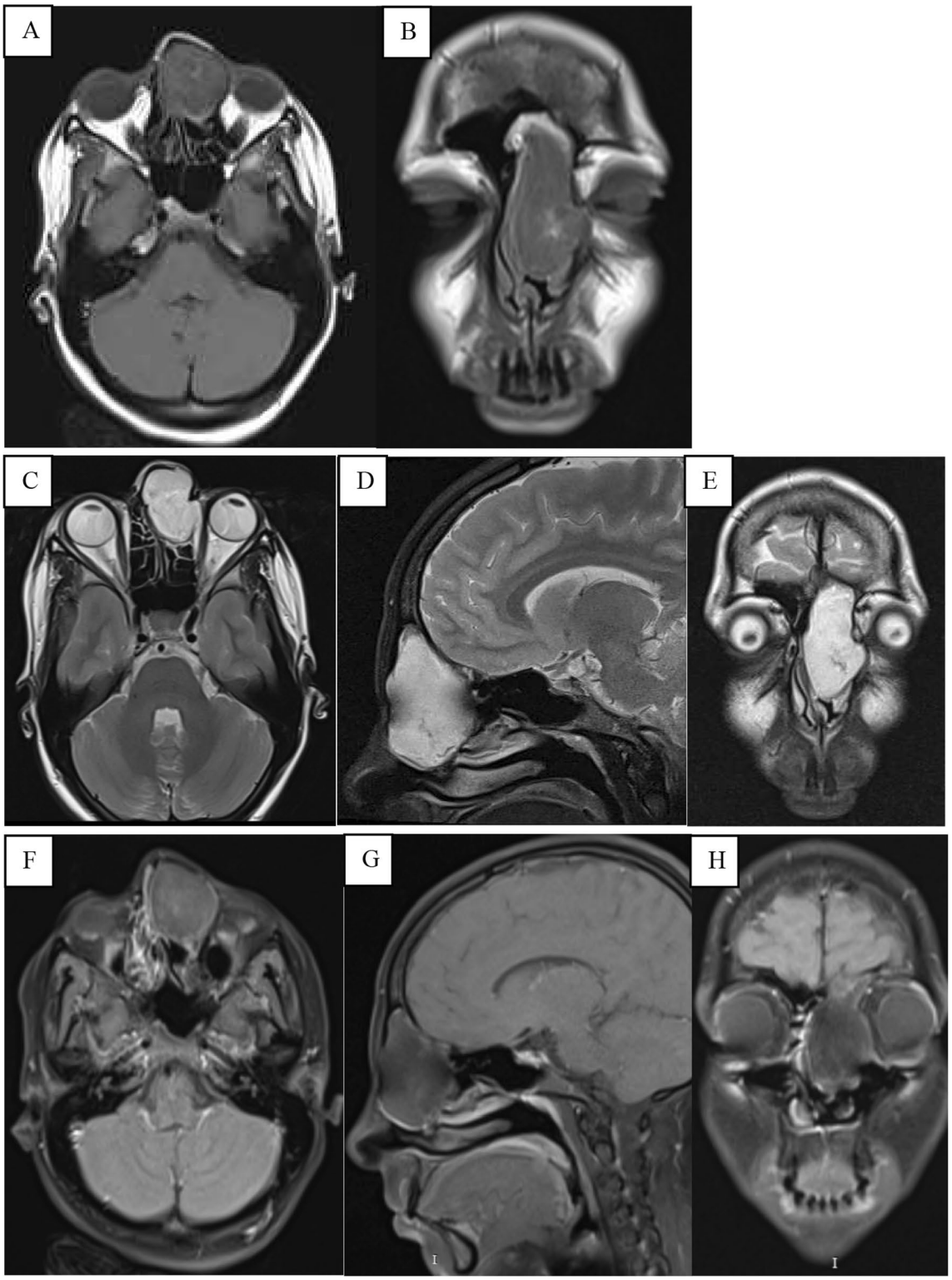

Fig. 7 MRI axial (**A**, **C**, **F**), coronal (**E**, **H**) and sagittal (**D**, **G**) cuts, showing a soft tissue cystic swelling inside a left concha bullosa. It is hypointense in T1 (**A**, **B**) and hyperintense in T2 (**C**–**E**). T1 with gadolinium (**F**–**H**) showed no enhancement

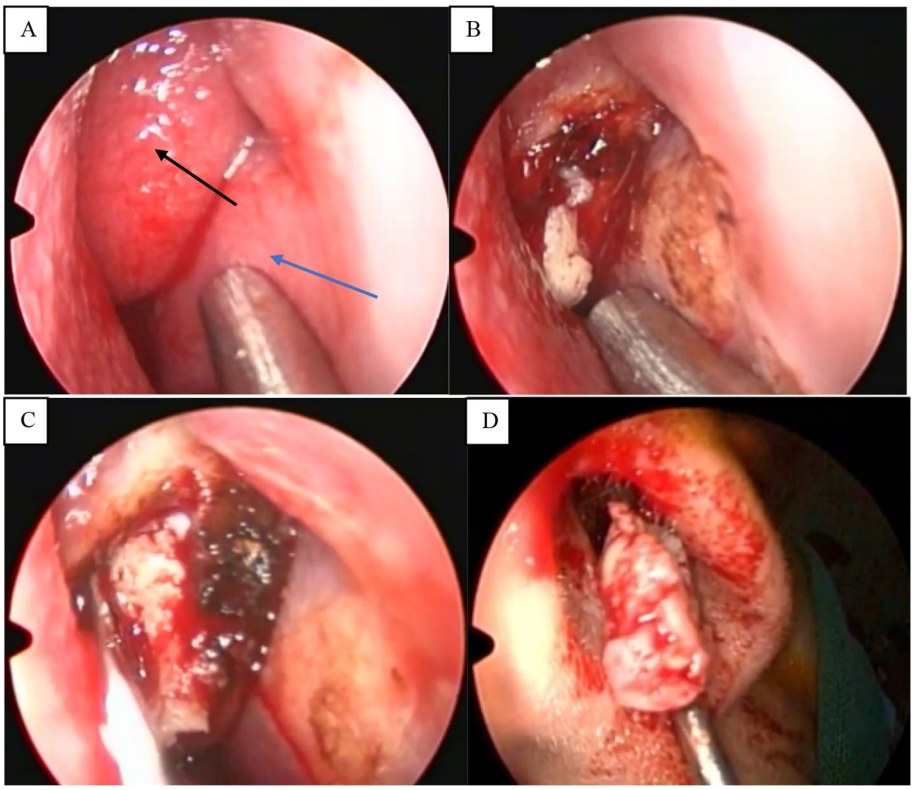

Fig. 8 Intraoperative endoscopic view showing a left concha bullosa expansile swelling (black arrow) (**A**) with suction tip pointing to the left inferior turbinate (blue arrow). Incision of the anterior wall of the lesion (**B**) with keratin layers coming out from it (**C**, **D**)

The differential diagnosis for nasal cholesteatoma includes mucoceles (mainly), pyoceles, cholesterol granulomas, sebaceous cysts, dermoid cysts, and neoplasms (benign and malignant) of the nasal sinuses due to associated bone destruction. Paranasal mucoceles are the commonest painless lesions presenting with similar radiologic findings to nasal cholesteatomas [3, 15]. Nasal cholesteatomas are frequently misdiagnosed preoperatively as paranasal sinus mucoceles, but unlike nasal cholesteatomas, mucoceles are usually preceded by sinusitis [7, 10, 17].

Preoperative radiologic diagnosis includes a CT scan and MRI of the nose and paranasal sinuses. The CT scan can show a non-enhancing smooth expansile soft tissue lesion with an erosion of the surrounding bone without soft tissue invasion. Diffusion-weighted imaging (DWI) MRI can differentiate nasal sinus cholesteatoma from nasal mucoceles (cholesteatoma is non-enhanceable on T1 with contrast and gives a high signal in DWI, denoting restricted diffusion; meanwhile, nasal mucoceles have a low signal in DWI). Furthermore, MRI can differentiate sinusitis from sinus neoplasms [3].

Complete wide surgical resection is the treatment of choice in nasal cholesteatoma to prevent recurrence or residual formation and for easier postoperative follow-up [8]. Some surgeons recommend second-look surgery in case of suspected residual keratin [15].

In this study, we reported three cases of nasal sinus cholesteatoma inside the ethmoid sinus, frontal sinus, and concha bullosa. Like most nasal sinus cholesteatomas, the three cases were misdiagnosed preoperatively. The diagnosis was suspected intraoperatively when layered keratin debris was found and was confirmed postoperatively by histopathologic examination. Since there was no history of trauma or previous sinus surgery in the first and third cases, the etiology was suspected to be congenital. Meanwhile, the etiology of the second case was most probably acquired cholesteatoma from previous frontal sinus surgery. The cholesteatoma matrix was not separable from the sinus mucosa in the three mentioned cases with mucosal layer keratinization; hence sinus mucosal lining was removed with the cholesteatoma matrix. In the frontal sinus cholesteatoma case, we inserted a Silastic stent for 6 weeks to prevent frontal sinus ostium stenosis. Postoperative follow-up included endoscopic examination (1, 3, 6, 12, and 24 months postoperatively) with a postoperative CT scan after 12 months and diffusion-weighted MRI every 12 months for 2 years. MRI was more accurate and specific than CT in detecting residual or recurrent cholesteatoma. Delayed post gadolinium T1 images and diffusion-weighted images can differentiate cholesteatoma from inflammatory or scar tissue. Unlike inflammatory tissues, cholesteatoma does not show enhancement on delayed post-contrast T1 images [11, 12]. Furthermore, postoperative

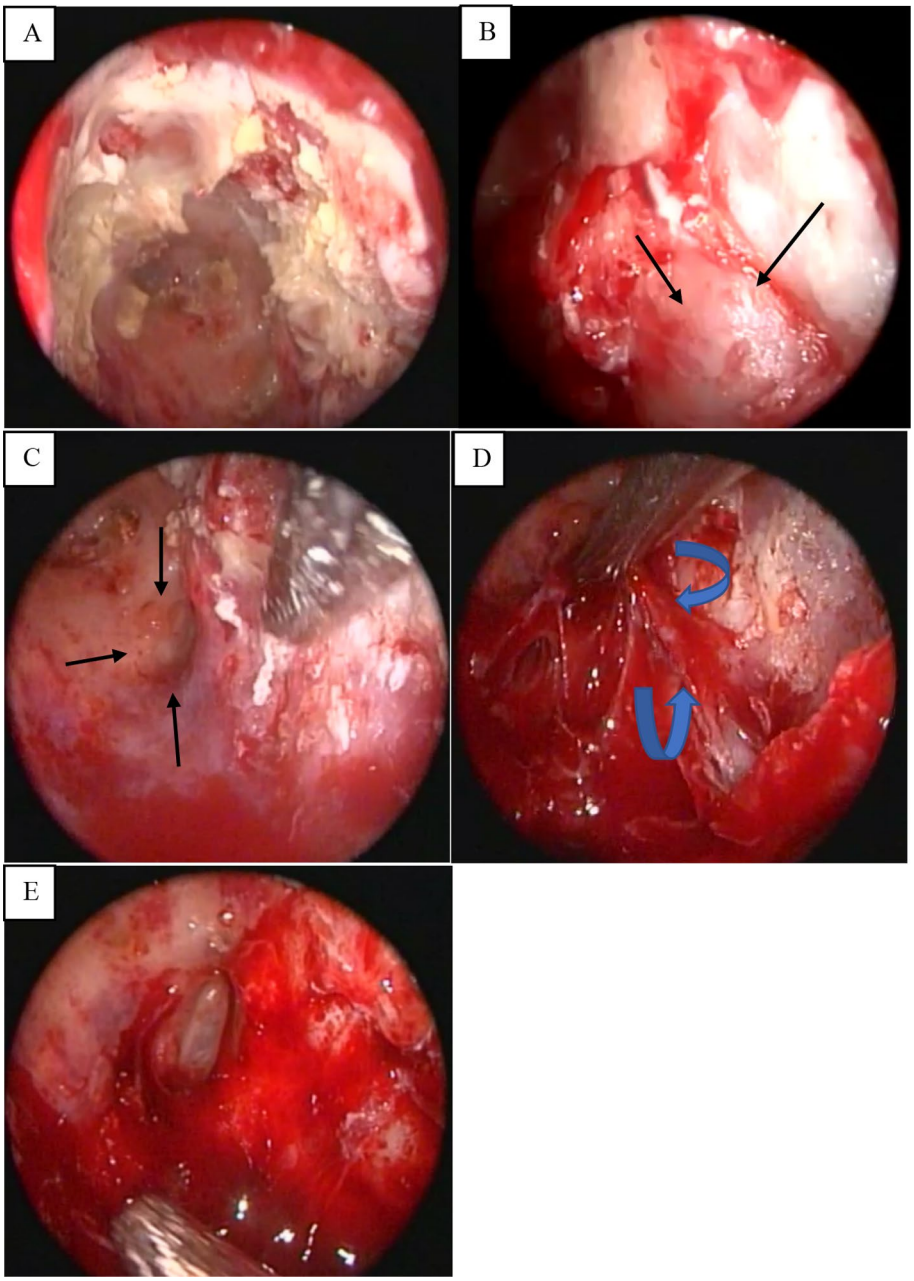

Fig. 9 Intraoperative endoscopic view showing Keratin layers filling the concha bullosa cystic swelling (**A**). Lamina papyracea defect (two black arrows) evident by orbital palpation (**B**). No erosion of the skull base (ethmoidal roof) (three black arrows) (**C**). Complete removal of the cyst wall keratinized epithelium (two curved blue arrows) (**D**). Cavity after suction of keratin and complete excision of the cyst wall (**E**)

follow-up for 2 years revealed no recurrence with no need for second-look surgery.

Frontal sinus cholesteatoma management has evolved from aggressive external surgical approaches such as osteoplastic flaps with frontal sinus obliteration or frontal sinus trephine (eyebrow incision) to endoscopic management with image guidance navigation to have access to its usual lateral extension into the frontal sinus recess [11, 14, 20, 22]. In some cases, frontal sinus cholesteatoma may erode the posterior table with subsequent intracranial abscess or meningitis, which can be the first presentation. In this case, the bicoronal approach and osteoplastic flaps with abscess evacuation are to be performed with possible frontal sinus obliteration in multiple staged surgeries. If the dura is involved, the affected part must be removed and repaired [3, 11, 21]. Lai et al. reported a case of bilateral frontal sinus cholesteatoma that was managed by Draf III only (without an external approach) and showed no recurrence over a 2-year postoperative follow-up [20]. In our case, Draf IIb was done with frontal sinus trephine with no recorded recurrence, because the lesion was strictly unilateral. In primary cases, however, Draf III would be ideal if surgery is only conducted by endoscopy, particularly if the mucosa is not keratinized, to facilitate postoperative monitoring and keep the sinus outflow patent for mucociliary clearance. At the time of our surgery, navigation was not available, and

Table 1 Showing the reported cases of nasal sinus cholesteatomas in the English literature including the present three cases (*SCC* squamous cell carcinoma)

Reported cases ($N=42$)	Site	Gender	Age in years	History of cranial trauma or nasal/sinus surgery	Malignant change or intracranial complication
1. Spencer [1]	**Frontal sinus**	Male	40	Cranial trauma	
2. Osborn et al. [18]		Female	62	Sinus surgery	**S.C.C**
3. Calcaterra et al. [10]		Male	44	None	
4. Holt et al. [6]		Female	24		
5. Maniglia et al. [16]		Male	67		**S.C.C**
6. Campanella et al. [7] (Three cases)		Male	65		
		Male	46		
		Male	36	Cranial trauma	
7. Newman et al. [17]		Male	70	None	**Verrucous carcinoma**
8. Hopp et al. [5]		Male	23		
9. Hansen et al. 2007 [3]		Female	80		
10. Hammami et al. [11]		Male	25		**Meningo-encephalitis**
11. Lai et al. [20]		Male	53		
12. Zoia et al. [15]		Male	32		**Frontal lobe abscess**
13. Kurien et al. [21]		Female	27		
14. Tejani et al. [22]		Male	45	Cranial trauma	
15. Present case		Female	35	Sinus surgery	
1. Campanella et al. [7]	**Ethmoid sinus**	Male	48	Nasal surgery	
2. Cingi et al. [23]		Female	16	None	
3. Barnett et al. [24]		Male	76	None	
4. Chandra et al. [14] (supraorbital cell cholesteatoma)		Female	22	Transsphenoidal hypophysectomy	
5. Present case		Female	17	None	
1. Pogorel et al. [25]	**Maxillary sinus**	Male	36	None	
2. Mills et al. [26]		Female	65	None	
3. Baxter et al. [27]		Male	20	Nasal surgery	
4. Das [9]		Female	55	None	
5. Paaske et al. [28]		Male	76	Cranial trauma	
6. Sadoff et al. [29]		Female	55	None	
7. Storper et al. [30]		Male	12		
8. Vaz et al. [31] (Congenital)		Female	1.5		
9. Palacios et al. [32]		Male	57		
10. Viswanatha et al. [2]		Female	18		
11. Viswanatha et al. 2011 [33] (maxillary & ethmoid sinuses)		Male	10	Cleft lip & palate repair	
12. Buric et al. [34]		Female	37	None	
13. Sozansky et al. [35]		Male	72	None	
14. Jin et al. [36]		Female	36	Sinus surgery	
15. Vakalapudi et al. [37]		Male	36	None	
1. Ohta et al. [38]	**Sphenoid sinus**	Male	36	None	
2. Sani et al. [39]		Male	25		**Pituitary apoplexy**
3. Kanjanawasee et al. [40]		Female	82		**Skull base osteomyelitis**
1. Cukurova et al. [13]	**Concha bullosa**	Female	81	None	
2. Present case		Female	24		

we felt safe performing the combined approach, especially when there was a previous external approach scar. Although Zoia et al. recommended a second look surgery if the frontal sinus mucosa was found keratinized, Tejani et al. dealed with residual keratin debris in the lateral frontal recess (after unilateral endoscopic frontal sinusotomy) by postoperative

in-office endoscopic debridment [15, 22]. In case of recurrence, Zoia et al. recommended frontal sinus obliteration, but obliteration is not preferred if the sinus drainage is normal as it hinders postoperative radiologic follow-up [15]. We think that a combined external approach and Draf III can be considered if recurrence occurs. As malignant change has been reported in a few cases of frontal sinus cholesteatoma, proper diagnosis and strict postoperative follow-up are critical [16–18].

Ethmoid sinus cholesteatoma management has progressed from external (lateral rhinotomy) to totally endoscopic approaches with excellent postoperative results [14, 23, 24, 33].

Maxillary sinus cholesteatoma usually presents as a slowly expansile lesion that can lead to nasal obstruction, recurrent sinusitis not responding to medical treatment, teeth loosening, check or hard palate swelling, and orbital complications due to associated bone destruction. Its management evolved from an external Caldwell–Luc approach to endoscopic excision with excellent postoperative results [27, 33]. Although the cholesteatoma wall must be totally removed to avoid recurrence, **Vaz** et al. treated an 18-month-old patient with congenital maxillary sinus cholesteatoma using endoscopic exteriorisation through inferior meatal antrostomy, reserving the Caldwell–Luc procedure for recurrence to maintain normal facial development [31]. Furthermore, Sozansky et al. showed that marsupialization only with wide middle meatal antrostomy could be effective with no recurrence after 13 years of follow-up [35]. However, complete surgical removal of the wall is still the method recommended to avoid erosion of the surrounding structures and recurrence. A CT scan will help determine the extent of the cholesteatoma and aid in selecting the most appropriate approach prior to surgery. The Caldwell–Luc procedure is preferred for complete removal in extensive cases. In addition, endoscopic monitoring is crucial following surgery, such as inverted papilloma [36]. In contrast to frontal sinus cholesteatoma, where late recurrence was reported, there were no maxillary sinus cholesteatoma recurrence cases despite leaving a residual in some cases [2, 15, 31]. Mucocele, pyocele, odontogenic keratocysts, and neoplastic (benign and malignant) lesions are potential differential diagnoses for maxillary sinus slowly growing expansile lesions [30].

Sphenoid sinus cholesteatoma usually presents with complications due to destruction of the cavernous sinus and optic chiasma (abducent nerve injury, decreased visual acuity or diplopia, ptosis due to third cranial nerve injury) or intracranial extension causing (pituitary apoplexy, severe headache, meningitis, hydrocephalus, and skull base osteomyelitis) [39, 40]. The endoscopic transsphenoidal approach is preferred for completely removing sphenoid cholesteatoma, but external approaches (craniotomy) may be required in severe cases [40].

Cholesteatoma inside a concha bullosa was reported once only in the English literature by Cukurova et al. [13]. Although the same author published another case in 2010, revising the computed tomography and histopathology pictures revealed that it was the same case published in 2009 [41]. Consequently, the case presented here is the second reported case for concha bullosa cholesteatoma in the English literature.

If nasal sinus cholesteatoma is associated with frontal or nasal bone destruction, cosmetic reconstruction must be delayed for 1 or 2 years to exclude recurrence or residual [6, 21, 23].

A review of the English literature demonstrated that there are 42 cases of nasal sinuses cholesteatomas (including the present three cases) (17 in the frontal sinus, 15 in the maxillary sinus, 5 in the ethmoid sinus, 3 in the sphenoid sinus, and 2 in a concha bullosa). The frontal sinus was the most affected, followed by the maxillary and the ethmoid sinuses. The least affected sinus was the sphenoid sinus (Table 1). It is noteworthy that previous reviews mentioned that the frontal sinus was the most common site for nasal sinus cholesteatomas, followed by the ethmoid and then the maxillary sinus [14]. Eleven patients only (26.2%) had a previous history of cranial (sinus) trauma and nasal (or sinus) surgery "implantation theory." Consequently, the most common cause of nasal sinus cholesteatoma is congenital ectodermal remnants during early embryologic development "congenital theory" [4].

Notably, malignant change had been reported only in frontal sinus cholesteatoma (Squamous cell carcinoma and verrucous carcinoma) but not in other nasal sinuses cholesteatomas [16–18]. In addition, paranasal cholesteatomas are usually unilateral, with few reported bilateral cases [8, 20, 42].

Conclusions

Nasal sinus cholesteatomas are rare lesions but must be considered in the differential diagnosis of slowly growing lesions of the nasal sinuses. Preoperative CT scan and diffusion-weighted MRI significantly contribute to proper diagnosis and exclude other similar lesions, such as nasal sinus mucoceles, cholesterol granuloma, or neoplastic lesions. Wide complete surgical excision is necessary to avoid recurrence and facilitate postoperative follow-up. Like ear cholesteatoma, strict postoperative follow-up is mandatory to diagnose recurrence or residual early and is done by endoscopic examination along with diffusion-weighted MRI and delayed post-gadolinium T1 images.

Learning points

- Nasal sinus cholesteatoma can be misdiagnosed as mucocele, so careful preoperative radiologic assessment (including MRI diffusion-weighted imaging) is essential for slowly growing expansile nasal and sinus lesions.
- For nasal sinuses cholesteatoma, wide complete surgical removal and strict follow-up by endoscopic examination and diffusion-weighted MRI is significant to avoid recurrence, malignant change, and intracranial or intraorbital complications.

Declarations

Conflicts of interest No conflicts of interest.

Ethical considerations All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional and the national research committee and with the 1964 Helsinki declaration and its later amendments.

Consent A preoperative informed written consent has been obtained from all patients.

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