Frontal Sinus Cholesteatoma Presenting with Intracranial and Orbital Complications: Diagnosis and Treatment

Case Report

Débora Cristina Gaspar Gonçalves[1], Tiago Soares Santos[1,2], Vera Claúdia Miranda Silva[1], Hugo Narcy Amaral Amorim Costa[1], Carlos Magalhães Ferreira Carvalho[1]

[1]Clinic of Otorhinolaryngology, Centro Hospitalar de Entre o Douro e Vouga, Santa Maria da Feira, Porto, Portugal
[2]Department of Otorhinolaryngology, Unit for Multidisciplinary Research in Biomedicine, "Abel Salazar" School of Medicine, Biomedical Sciences - University of Porto, Porto, Portugal

Abstract

Frontal sinus keratoma or cholesteatoma is a rare disease of paranasal sinuses and presents as a slow-growing mass that becomes symptomatic as it grows to the surrounding structures. Intracranial complications are not a common presentation and are potentially life-threatening. Frequently the final diagnosis is only made intraoperatively because several other frontal sinus tumors behave likewise. Definitive treatment requires complete removal of the keratoma, and a combined endoscopic and external frontal sinus approach is a good treatment option. In this report, we presented a 68-year-old female with frontal sinus cholesteatoma with diagnostic and therapeutic features of this pathology with the review of the literature.

Keywords: Fontal sinus, cholesteatoma, complication, orbit, surgery, case report

ORCID IDs of the authors:

D.C.G.G. 0000-0003-4272-2215;
T.S.S. 0000-0003-2706-7531;
V.C.M.S. 0000-0002-8930-9383;
H.N.A.A.C. 0000-0001-6064-7461;
C.M.F.C. 0000-0002-0112-422X.

Cite this article as: *Gaspar Gonçalves DC, Santos TS, Miranda Silva VC, Amorim Costa HNA, Ferreira Carvalho CM. Frontal Sinus Cholesteatoma Presenting with Intracranial and Orbital Complications: Diagnosis and Treatment. Turk Arch Otorhinolaryngol 2022; 60(4): 234-237.*

Corresponding Author:
Débora Cristina Gaspar Gonçalves;
debora.goncalves@chedv.min-saude.pt

Received Date: 20.07.2022
Accepted Date: 13.12.2022

DOI: 10.4274/tao.2022.2022-7-7

Introduction

Paranasal sinus keratoma or "cholesteatoma" is a tumour-like mass composed of keratinizing squamous epithelium involved by a sac within an air-filled space. This type of lesion is frequent in the middle ear but rare on paranasal sinuses (1). The term "cholesteatoma" is a misnomer and to avoid confusing and inaccurate classification of paranasal sinuses cholesteatoma Hopp and Montgomery (2) proposed the use of the term "keratoma". Keratomas have been reported at the ethmoidal cells and the maxillary sinus, but the most frequent location at the paranasal sinuses is the frontal sinus (3). Treatment involves complete surgical excision, which sometimes can be challenging considering the size and structures involved by the tumour. We report a case of frontal sinus keratoma presenting with intracranial and orbital complications submitted to combined endoscopic Draf type III procedure and frontal sinus external approach with osteoplastic flap technique.

Case Presentation

A 68-year-old female patient was evaluated with complaints of headache and fever lasting two days. She also presented right eyelid oedema, ptosis, proptosis, chemosis and conjunctival redness. There was no restriction of eye movements, decreased visual acuity, focal neurologic signs, or nasal symptoms. She reported a previous history of acute frontal sinusitis complicated with subdural fronto-parietal empyema 10 years before. At that time, she was treated in a different institution, by a combined frontoparietal craniotomy and endoscopic drainage of frontal sinus.

Computed tomography (CT) scan revealed right frontal sinus filled with heterogenous soft tissue mass. The lesion eroded the posterior wall of the frontal sinus and a small extra-axial collection was present in-between (Figure 1). The orbital roof was also eroded allowing the mass to contact with orbital contents. Cerebrospinal fluid (CSF) analysis revealed hyperleukocytosis with a predominance of polymorphonuclear neutrophils. The diagnosis of frontal mucocele complicated with orbital cellulitis, meningoencephalitis and subdural empyema was established. Empirical antibiotic therapy was initiated with ceftriaxone, metronidazole, and vancomycin. Magnetic resonance imaging (MRI) of the paranasal sinuses showed a heterogeneous expansive mass filling the frontal sinus predominantly hyperintense on T2 weighted fat saturated images and hypointense on T1-weighted images (Figure 2, 3).

We performed an endoscopic sinus surgery, to drain the right frontal sinus mucocele. However, intraoperatively we found a thick white cheese-like mass filling the right frontal sinus (Figure 4). Given the lack of frozen section analysis availability, a definite surgical excision was postponed. The patient completed 12 days of antibiotic therapy, with progressive clinical improvement. Reduction of sinus inflammation and resolution of the empyema was reported on CT-scan three days after surgery. The patient was discharged from hospital with no neurologic or ophthalmologic sequels.

Final diagnosis of frontal sinus keratoma was made after histopathologic examination. We performed a revision surgery through a combined endoscopic Draf type III procedure and frontal sinus external approach with osteoplastic flap technique via a coronal incision (Figure 5). The keratoma was well delimited and not attached to the anterior fossa dura so we were able to perform a complete macroscopic excision with no iatrogenic CSF leak. At 6-month follow-up, no endoscopic signs of recurrence were present (Figure 6), and MRI evaluation did not show signs of cholesteatoma. Written informed consent regarding publishing her data and photographs were provided by the patient.

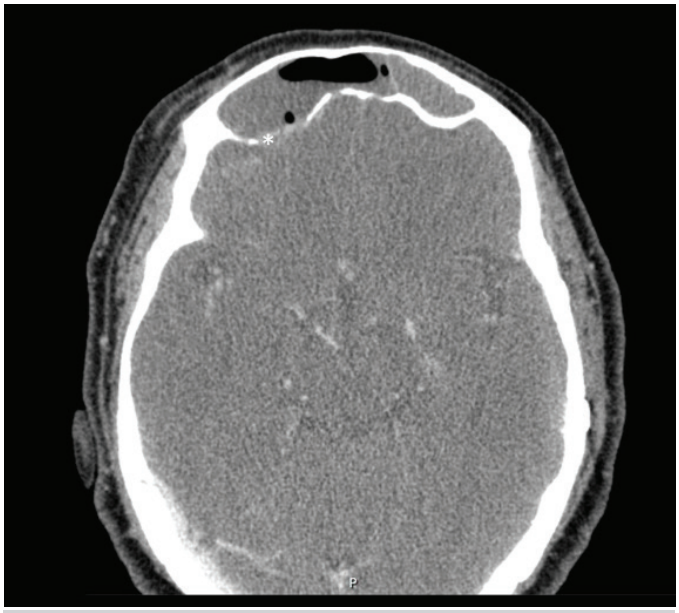

Figure 1. CT-scan at the emergency department (before any therapeutic intervention) a soft tissue opacification of frontal sinus and erosion of the posterior wall of the frontal sinus with associated extradural empyema (*)

CT: Computed tomography

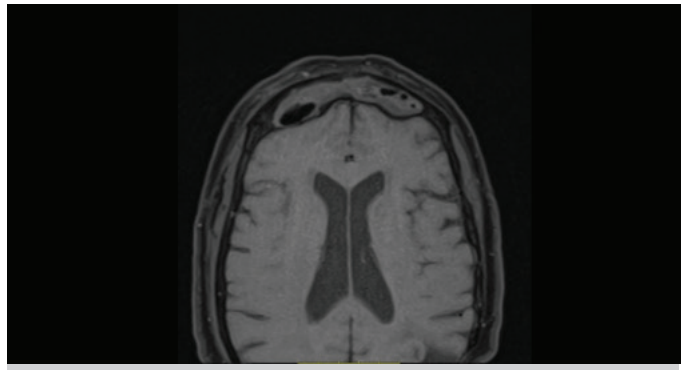

Figure 2. MRI T1 weighted imaging sequence showing a frontal sinus mass with a predominant hypointense sign

MRI: Magnetic resonance imaging

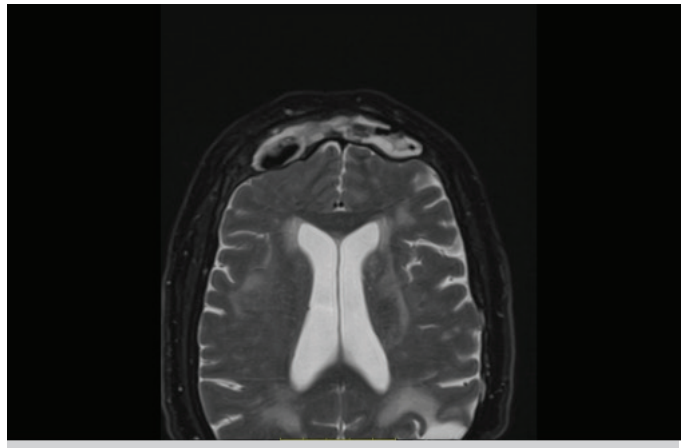

Figure 3. MRI T2 + fat saturation weighted imaging sequence showing a frontal sinus mass with a predominant hyperintense sign

MRI: Magnetic resonance imaging

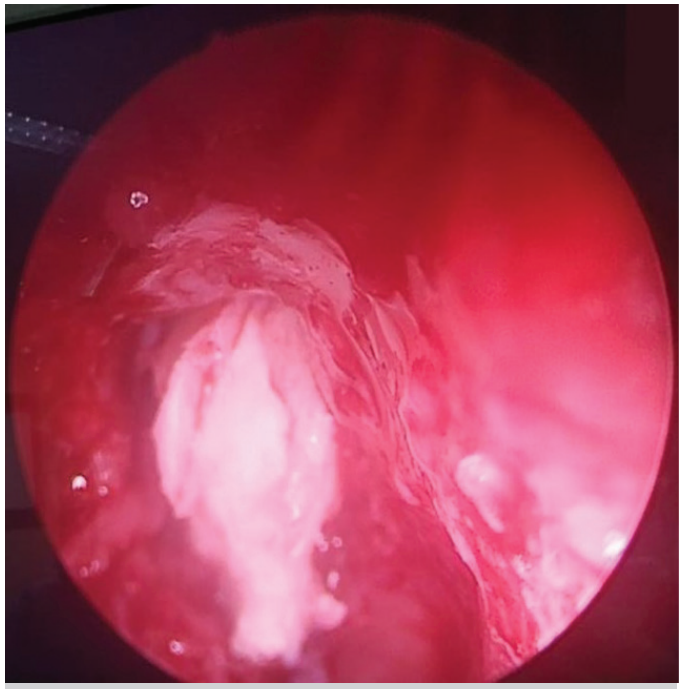

Figure 4. Endoscopic view of the frontal sinus during urgent sinus drainage, reveals a white cheesy mass suggestive of keratoma

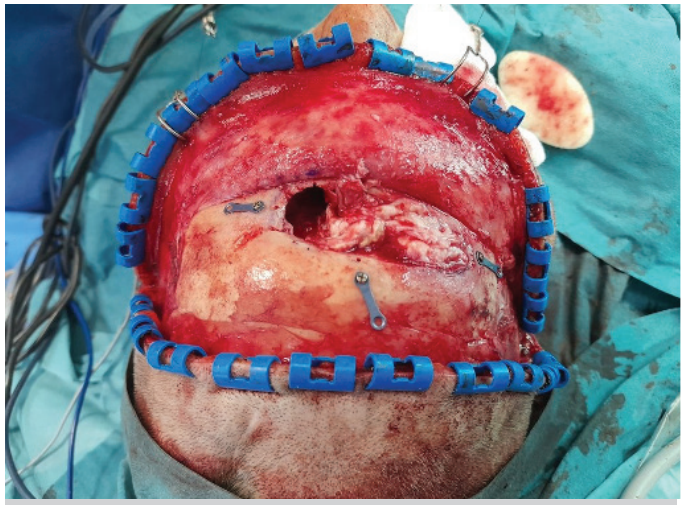

Figure 5. Surgical revision procedure: a combined approach with endoscopic Draf type III procedure and frontal sinus external approach with osteoplastic flap technique

Discussion

Frontal sinus keratomas are rare, a literature review in 2006 revealed 13 cases (1). The etiopathogeneses is not clear (2). Acquired cholesteatomas can be explained by implantation theory (squamous epithelium transport from other locations to the sinuses by surgery or trauma), migration theory (epithelium migrates from the nasal vestibule to the frontal sinus) or squamous metaplasia theory. In our case, the history of previous nasal surgery combined with craniotomy may favour the implantation theory, however we consider more plausible the previous presence of the cholesteatoma that kept undiagnosed until now.

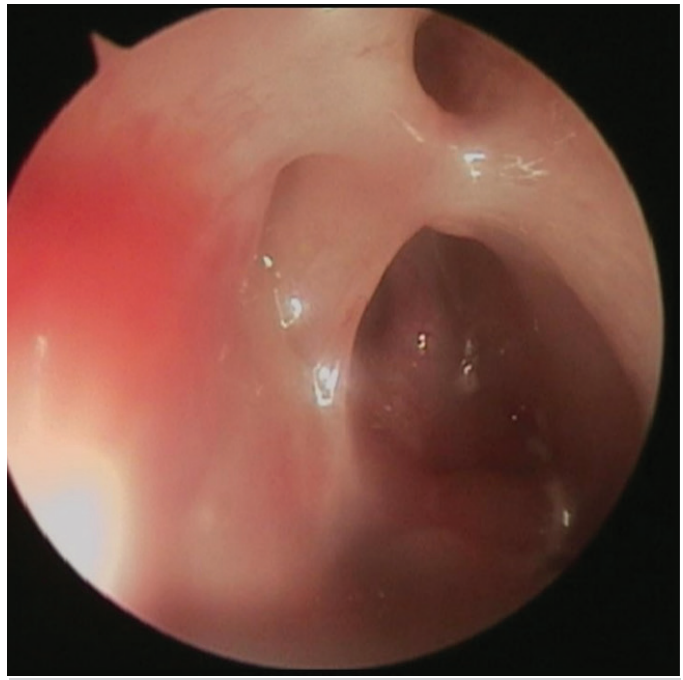

Figure 6. Endoscopic view of a Draf type III procedure, after 6 months, reveals no signs of keratoma

Frontal sinus keratomas are slow growing lesions, and most symptoms are secondary to erosion of the surrounding bone (2, 4). Anterior growing causes frontal headache and forehead deformity, on the other hand, inferior erosion allows growing into the orbit causing diplopia and ocular pain (1). Erosion of the posterior wall may cause intracranial complications like meningitis, empyema, and cerebral abscess (2). Our case is one of the few that presented with intracranial complications (3, 4). Chronic sinusitis symptoms are not typically present because the keratomas tend to lie laterally on the frontal sinus not blocking the frontal sinus duct (2).

Several other lesions of the frontal sinus behave like the keratoma. On MRI, keratomas usually appear hypointense on T1 and hyperintense on T2 imaging sequences, like the mucoceles (depending on the degree of fluid content) (3). However, keratomas lack enhancement with contrast and are hyperintense in diffusion-weighted imaging compared with CSF and brain parenchyma (5). However, frequently the keratoma is only diagnosed intraoperatively or after histopathologic examination (1).

The treatment is surgical excision. Osteoplastic frontal sinus obliteration procedure is frequently described in the literature (2). However, given the progressive improvements of endoscopic paranasal sinus surgery, re-establishing the draining route, allows a patent frontal sinus and may represent a good alternative, but evidence on this subject is sparse (4). Nevertheless, endoscopic approach is frequently insufficient for removal of the entire lesion, especially in a well pneumatized frontal sinus, so a combined osteoplastic frontal sinus approach should be considered. Once the frontal

sinus is cleared from the keratoma, obliteration may be performed, usually with abdominal fat (4). When it's possible to create a good tract of flow, frontal sinus obliteration is not required and leaving an open cavity will allow better control of recurrence (6). Clinical examination with nasal endoscopy and MRI are appropriate for follow-up (3).

Conclusion

Frontal sinus keratoma is a slow growing tumour that can reach significant proportions before any symptoms are reported and present with intracranial complications that are fortunately uncommon.

The combined endoscopic and external approach to the paranasal sinuses is a good therapeutic option, restoration of the frontal sinus recess offers an alternative to sinus obliteration.

Informed Consent: Written informed consent regarding publishing her data and photographs were provided by the patient.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: T.S.S., H.N.A.A.C., Concept: D.C.G.G., T.S.S., H.N.A.A.C., C.M.F.C., Design: D.C.G.G., T.S.S., C.M.F.C., Data Collection and/or Processing: D.C.G.G., V.C.M.S., Analysis and/or Interpretation: D.C.G.G., T.S.S., V.C.M.S., H.N.A.A.C., C.M.F.C., Literature Search: D.C.G.G., V.C.M.S., Writing: D.C.G.G., T.S.S.

Conflict of Interest: There is no conflict of interest to disclose.

Financial Disclosure: The authors declared that this study has received no financial support.

Main Points

- Frontal sinus keratoma may present with intracranial complications.
- Diagnosis of frontal sinus keratoma is challenging because other diseases like mucoceles can mimic this entity.
- Treatment with a combined endoscopic and external frontal sinus approach is a good treatment option.

References

1. Hansen S, Sørensen CH, Stage J, Mouritzen A, Cayé-Thomasen P. Massive cholesteatoma of the frontal sinus: Case report and review of the literature. Auris Nasus Larynx 2007; 34: 387-92. [Crossref]
2. Hopp ML, Montgomery WW. Primary and secondary keratomas of the frontal sinus. Laryngoscope 1984; 94: 628-32. [Crossref]
3. Hammami B, Mnejja M, Chakroun A, Achour I, Chakroun A, Charfeddine I, et al. Cholesteatoma of the frontal sinus. Eur Ann Otorhinolaryngol Head Neck Dis 2010; 127: 213-6. [Crossref]
4. Kurien R, Thomas L, Varghese L, Nair BR. Frontal sinus cholesteatoma: a masquerading diagnosis. BMJ Case Rep 2019; 12: e231495. [Crossref]
5. Schwartz KM, Lane JI, Bolster BD, Neff BA. The utility of diffusion-weighted imaging for cholesteatoma evaluation. AJNR Am J Neuroradiol 2011; 32: 430-6. [Crossref]
6. Nicolai P, Caltelnuovo P. Benign Tumors of the Sinonasal Tract. Flint PW, Haughey BH, Lund VJ, Niparko JK, Robbins KT, Thomas JR, Lesperance MM, editors. Cummings Otolaryngology - Head and Neck Surgery. Sixth ed. Philadelphia: Elvesier Saunders; 2015. p. 740-51. [Crossref]