

Inverted papilloma of the sphenoid sinus: presentation of three cases*

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SUMMARY

Introduction: *Inverted papilloma deriving originally from the sphenoid sinus is extremely rare. Three such cases are presented aiming to show the particularities and diagnostic clues of this clinical entity.*

Case Reports: *All three patients were male, and presented mainly with nasal symptoms. In all three cases imaging shows the sphenoid sinus was completely occupied by the tumor. Additionally the ethmoids and nasal cavities have been invaded in contiguity to the site of the initial appearance of the tumor. In particular one of the cases was of even more interest as a malignant transformation of the inverted papilloma of the sphenoid sinus was histologically observed. Endoscopic sinus surgery was the therapeutic approach chosen for all three cases, with very good results. Follow up 30 months after the operation has not revealed any signs of recurrence of the disease.*

Conclusion: *Inverted papilloma of the sphenoid sinuses - and even more malignant transformation - although quite rare, may present occasionally. The ENT surgeon should be aware of this condition. Preoperative imaging with CT scan and MRI and the experience of the surgeon will dictate the appropriate surgical approach.*

Key words: sphenoid sinus, inverted papilloma, malignant transformation

INTRODUCTION

Inverted papilloma is an uncommon tumor of the nose and paranasal sinuses. Its name is due to the propensity of the neoplasm to invert, with proliferation of the surface epithelium that lines the nasal cavity and sinuses into the underlying stroma. Despite being a benign tumor it may demonstrate aggressive local invasion, thinning the surrounding bony walls and pushing aside the neighboring structures. Malignant transformation as well as coexistence with carcinoma can occur in a 10% of cases. The usual site of initial appearance is the lateral nasal wall. Initial occurrence of inverted papilloma in the sphenoid sinus is rare. On the other hand isolated sinus disease by itself is uncommon, insidious and a real diagnostic and therapeutic challenge. The series that have been published so far report a very limited number of inverted papillomas. Review of the literature reveals a limited number of inverted papillomas of the sphenoid sinuses.

We present three cases of inverted papilloma deriving from the sphenoid sinus. The particular interest is the difference in clinical symptomatology and the presence of malignancy in one of them. Diagnostic and therapeutic procedures as well as follow up are presented and discussed. These cases are of particular interest, as the sphenoid sinuses present complex anatomy and

difficult surgical approach. More over, as pathology limited only to sphenoid sinuses is quite rare.

CASE REPORTS

Case 1

A 52-year-old male presented to the ENT outpatient clinic complaining of nasal fullness, purulent nasal discharge and postnasal drip over the last 3 years. He also reported several bouts of retrobulbar headache radiating to the occiput (Table 1). Physical examination, anterior rhinoscopy and endoscopy revealed accumulated purulent secretions in both nasal cavities and large polypoid masses occupying and obstructing both posterior nasal choanae and the posterior part of the nasal cavities. Computed tomography (CT scan) (Figure 1) and magnetic resonance imaging (MRI) of the nose and paranasal sinuses revealed a homogeneous opacification of both the sphenoid sinuses and the posterior ethmoid cells. The anterior wall of the sphenoid sinus was eroded with protrusion of the mass to the nasal cavities, particularly the right one. The mass slenderized the sella turcica by compression and pushed the pituitary gland upwards. Due to the suspicious clinical and radiographic appearance biopsy specimens were taken under local anaesthesia from both nasal cavities. Histology showed bilateral IP



Figure 1. Coronal CT view of patient case No 1, showing a large opacifying lesion occupying the entire sphenoid and elevating the sella turcica by compression.



Figure 2. Postoperative coronal CT view of the same patient (case No1), showing complete removal of the mass without any signs of recurrence.



Figure 3. Axial CT view showing the mass of patient case No 2, preoperatively. The mass occupies the whole sphenoid sinus and part of the posterior ethmoids.

without any evidence of malignant transformation. A functional endoscopic approach was decided. Under direct visualization with the use of a 0° and a 30° 4mm rigid endoscope, the mass was meticulously removed piece meal-with the use of a Blakesley-Weil forceps. A curved curette was used to gently remove the mass from the most lateral areas of the sphenoid sinus, which is particularly dangerous due to their proximity to the optic nerve and the internal carotid artery. The mucoperiosteum was partially included in the resection. The vomer was partially excised in order to facilitate access to the sphenoid sinus. The nasal cavities were packed upon completion of the procedure. No pre- and postoperative complications occurred. The final pathology report confirmed the diagnosis of inverting papilloma and was negative for malignancy.

Follow up one year postoperatively with clinical examination CT scan (Figure 2) and MRI did not reveal any signs of recurrence. On further follow up four years postoperatively the patient is still free of disease.

Case 2

A 42-year-old male presented with a six-month history of nasal obstruction with purulent nasal discharge and postnasal drip. He reported a retrobulbar headache and sexual impotence during the last two months (Table 1). The latter symptom alarmed him and visited an endocrinologist who requested a CT scan and MRI and subsequently referred him to the Otorhinolaryngology Department.

The anterior rhinoscopy and endoscopy revealed a polypoid mass in the posterior part of both nasal cavities, occupying part of the left nasal cavity. The CT scan of the paranasal sinuses showed a lesion of soft tissue density filling completely the sphenoid sinus (Figure 3) with slenderization of its walls without apparent signs of erosion of the sella turcica. The mass was extending to the posterior ethmoid cells with signs of erosion of the small intracellular lamellae, and to the nasal cavities, particularly to the left one. Bulging of the lateral ethmoid wall on the left was also observed.

MRI of the pituitary gland and hypothalamus showed invasion of the sphenoid sinus and part of the ethmoid cells by a lesion of moderate signal in both T1 and T2 weighted images, mildly enhancing with contrast. The sphenoid sinus appeared slightly enlarged. The pituitary gland, optic chiasm and cavernous sinus appeared normal.

The patient underwent endoscopic sinus surgery where the mass was excised piece-meal. Frozen sections sent from the sphenoid sinus mass revealed malignancy. The sphenoid, ethmoid and nasal cavity were cleaned from the mass. A part of the posterior septum was excised as well, in order to provide better access to the sphenoid sinus. Due care was taken because of the thinning and erosion of the bony walls and of the malignant nature of the lesion, which had to be completely removed. The procedure was performed very carefully due to the erosion and thinning of the adjacent bony walls and we tried to be very pedantic not to leave residual tumor. The

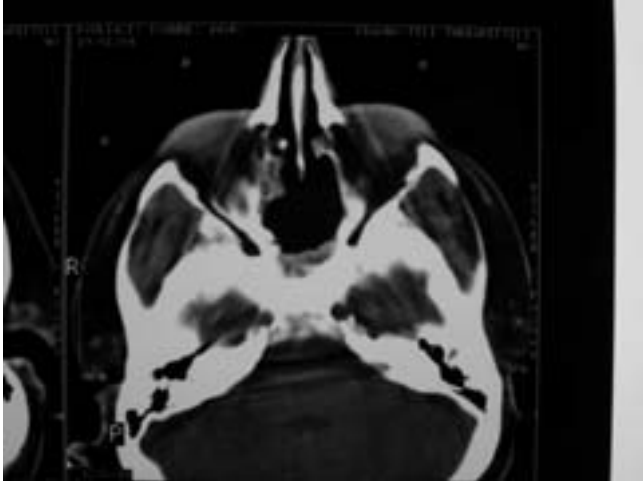


Figure 4. Axial CT view of patient, case No 2, 12 months postoperatively. No signs of recurrence were found.

mucoperiostium was included in the resection. Histology revealed a non-keratinized squamous cell carcinoma with an endophytic and an exophytic growth pattern. Foci of microinvasion of the underlying stroma were found but the bony pieces excised were free from invasion.

The patient was treated additionally with radiotherapy. One month postoperatively a control CT scan was performed revealing a very satisfying postoperative cavity with no signs of residual disease.

Follow-up examinations were performed at intervals of three months, over the next three years without recurrence. A CT scan (Figure 4) 12 months postoperatively did not reveal any alarming findings.

His impotence problem is followed up and treated by a specialist. It is not clear whether it could be attributed to his tumor or not. Thirty-two months after the operation the patient remains recurrence free.

Case 3

A 59-year-old male reported nasal obstruction and hyposmia during the past 12 months (Table 1). He had a history of nasal septoplasty 15 years ago and a nasal polypectomy ten years ago. Clinical examination revealed a polypoid mass that could be seen by endoscopy in the posterior part of both nasal cavities. A large perforation could also be seen at the upper part of the middle third of the nasal septum, possibly a postoperative complication. CT scan (Figure 5) revealed a homogenous mass filling the sphenoid sinus, extending to the nasopharynx and posterior nasal cavities bilaterally. He had a functional endoscopic nasal approach, where a frozen section was sent from the sphenoid sinus mass revealing inverted papilloma. The sphenoid was meticulously cleaned and the polypoid masses and the mucoperiostium were excised. Follow up endoscopy one year later revealed a sphenoid sinus free from recurrence. A CT scan (Figure 6) confirmed a lack of recurrence as well. Thirty months postoperatively the patient remains recurrence free.

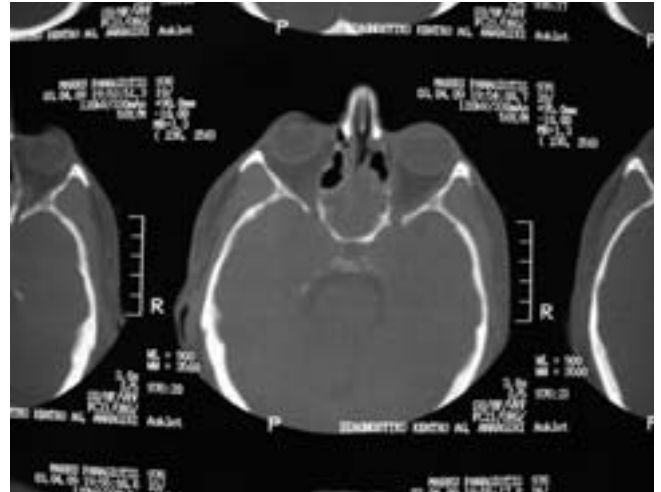


Figure 5. Axial CT view showing the sphenoid sinus of patient case No 3 completely occupied by the opacifying tumor.

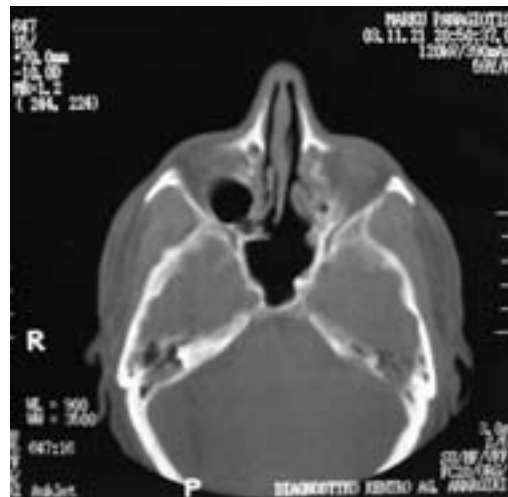


Figure 6. Axial CT view showing the sphenoid sinus of patient case No 3 12 months postoperatively. The sinus appears recurrence free.

DISCUSSION

Isolated sphenoid sinus disease is rare. The sphenoid sinus is more often affected by pathology originating from the neighboring structures, being either inflammatory or neoplasma. Studies⁽¹⁻⁴⁾ have shown that inflammatory lesions mainly sphenoid sinusitis and mucoceles are the most probable diagnosis when dealing with isolated sphenoid sinus disease.

The presence of Inverted Papilloma in the sphenoid sinus is therefore a rare finding of a clinical entity, which is by itself unusual. Sethi et al.⁽⁴⁾ reported 2 cases of IP in a series of 21 patients with isolated sphenoid lesions. Hadar et al.⁽¹⁾ reported 1 case of IP in a series of 21 patients with isolated sphenoid lesions, and Wang et al.⁽⁶⁾ reported 4 cases of sphenoid inverted papilloma in a series of 122 patients. The very few publications, which are focused on IP deriving from the sphenoid sinus^(4,7-10), do not exceed 2 or 3 cases each.

Our series of three cases is of a particular interest because one

of the cases presented malignant transformation. The main clinical symptoms were nasal obstruction and headache. Concerning presenting symptoms, cases limited to the sphenoid sinuses usually present with subtle symptoms such as retro-orbital headache or a sense of discomfort^(1,2,5). Symptoms such as nasal obstruction are present in cases of papillomas occupying the nasal cavity, arising either from the maxillary sinuses or from the sphenoids⁽⁹⁾.

A question arose concerning the original site of the inverted papillomas. Both the CT and the MRI revealed tumors occupying the nasal cavity, and the sphenoid sinus almost fully, with unification of both compartments of the sinus. This image of advanced sphenoid disease allows us to speculate the sphenoid sinus as the site of initial presentation rather than of contiguous expansion.

The patient with malignant transformation did not differ from the other two in any respect apart from the sexual impotence that he mentioned. The sella turcica and pituitary gland were unaffected by the tumor according to imaging and intraoperative findings. However, the proximity of the gland to the sphenoid sinus could allow a possible scenario of toxin-mediated effect on the gland, inhibiting normal hormone production.

Extensive literature search^(2,5) revealed that in cases of isolated sphenoid sinus disease, due to the anatomical structures and the limited access available, preoperative investigation with both CT and MRI and occasionally MRA is necessary. According to Savy⁽¹¹⁾ the main advantage of MRI is in defining the extent of the tumour and in differentiating it from adjacent inflammatory tissue, but it cannot differentiate IP from sinus malignancy.

Nachtigal et al.⁽¹²⁾ reported that in his series of IP with malignant transformation (8 patients), all of them exhibited maxillary antrum involvement at the time of diagnosis but the incidence did not differ statistically from the benign IP patients. However, ethmoid and sphenoid involvement was significantly more frequent in IP with malignant transformation patients at the time of diagnosis, compared with benign IP patients. Our patient did not exhibit any involvement of the maxillary sinus, but the tumor filled completely the sphenoid sinus and part of the ethmoids. Histology showed the inverted type of malignant papilloma, which according to Nachtigal is the most frequent papilloma subtype in case of malignancy (comparing with the fungiform and exophytic subtypes).

A controversy exists if endoscopic or external open approach is the most appropriate. It is our opinion that in particular concerning the sphenoid sinuses either approach provides limited access. Therefore, other factors should determine the decision: the surgeon's skills and knowledge, the available instrumentation and experience. Sham et al.⁽¹³⁾ summarized the gradually increasing and sometimes controversial experience of all surgeons who tried to apply endoscopic methods of resection, in the conclusion that the endoscopic approach is inadvisable for removal of tumors in endoscopically inaccessible areas of the maxillary, frontal or sphenoid sinuses as well as for those

extending to the orbit or intracranial structures. Lund et al.⁽¹⁴⁾ claim that it is the extent of disease that primarily determines the choice of surgical approach, with previous treatment, individual patient factors and surgical expertise as secondary determinants. The use of 30° and 45° endoscopes in addition to 0° makes the procedure easier and safer, allowing inspection of more remote areas of the surgical field.

In our cases endoscopic was the surgical approach chosen. This was done by a highly experienced surgeon with great care and effort to be as radical as possible not only in the case of malignancy but in benign cases as well, due to the well documented tendency of the tumor to recur (from 7% after radical extranasal excision to 59% after non endoscopic intranasal excision)⁽¹³⁾. Others report equal recurrence rate of inverted papilloma comparing endoscopic to extranasal open procedures⁽¹⁵⁾. The patients were informed and consented to the possibility of an alteration of the procedure to an external approach, if endoscopy did not secure a satisfying extraction of the tumor.

All three patients were treated satisfactorily endoscopically and remain free from disease after intervals that range from 30 months to four years. According to Nachtigal⁽¹²⁾ most authors believe that recurrence mainly presents within two years after the original excision. He recommends a follow-up at regular intervals for at least 5 years after surgical intervention. Indeed several authors agree with this length of follow-up (Pasquini⁽¹⁶⁾ suggested a mean follow-up of 54 months, Tomenzoli⁽¹⁷⁾ 55 months, Lawson⁽¹⁸⁾ 5.2 years). Therefore, follow up should cover this period of possible recurrence. In our cases follow up was four years, 32 months and 30 months for cases 1, 2 and 3, respectively. Patients 2 and 3 are still being followed up.

Follow-up should be done with endoscopy primarily, but CT scan and MRI should be used as well. In all series of sphenoid sinus inverted papillomas reported, some authors^(1,8,9) use mainly CT scan in addition to endoscopy and others⁽⁷⁾ use both CT and MRI. In a review regarding radiological follow-up of IP, Petit⁽¹⁹⁾ reports that MRI supplies the deficiencies of endoscopy in case of extensions to the frontal sinus or the lateral recess of the antrum especially if mucosal hyperplasia or sinusitis is associated. However for cost-effectiveness reasons CT is more frequently used for follow-up and it is up to the surgeon's judgement to use MRI whenever he finds it necessary.

CONCLUSION

Inverted papilloma of the sphenoid sinus is rare. However, the insidious progress of the tumor and the erosive behavior of it, together with a small but non-negligible possibility of malignancy, make the diagnosis of this entity very important. The particularities of the sinus topography that require special skills in its surgical approach mandate a thorough and careful consideration and the reference of patients to specialized surgeons. Functional endoscopic sinus surgery is an option becoming more popular as experience grows, but it should

always be chosen after logical and justified consideration and a meticulous endoscopic and imaging preoperative control.

Despite the satisfactory early results, recurrence should always be kept in mind and follow-up should continue beyond the first two years.

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