Sphenoidal mucocoele

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In view of the fact that the incidence of sphenoidal mucocoele is rare and most of the cases have been misdiagnosed and many craniotomies have been performed wrongly, we consider this case of sphenoidal mucocoele worth reporting. So far, perhaps one dozen cases have been diagnosed correctly pre-operatively. Linthicum *et al.* (1946) says that very rarely has it been preoperatively diagnosed. Whenever a patient presents himself with headache, particularly in the occipital region, hyposmia or cacosmia, nasal symptoms associated with eye symptoms one should keep in mind the possibility of sphenoidal sinus pathology.

Looking into the literature we find that about 300 cases of frontal sinus mucocoele have been reported. Ethmoid and maxillary mucocoeles are much less common. Only about 40 cases of sphenoidal mucocoele have been reported to date. To our knowledge no case has been reported in India. Until 1907, no case was reported. Until 1959 only 9 cases were reported, in English literature (Cody and Hallberg, 1959). Holt and Rötth, in 1940 suggested the name 'Orbital Apex and Sphenoidal Fissure Syndrome', to this symptom complex. Tassman in 1947 suggested the term 'Sphenoid Fissure-Optic Canal Syndrome with Complete Ophthalmoplegia' because he considers the syndrome to be incomplete when the optic nerve or other structures are spared. In 1958 Smith introduced the phrase 'Orbital Apex Syndrome' for the clinical manifestations of a lesion which affects the nerves and vessels passing through the Superior orbital fissure and optic foramen, and recognized the importance of sphenoid disease in causing the characteristics of this syndrome. Cody and Hallberg (1959) suggested the phrase 'Orbital Inlet Syndrome' in view of the fact that it would describe adequately the various combinations of signs and symptoms.

There is considerable controversy as to its aetiology. Defective outlet from the sinus was thought to be the cause (Neffson, 1967). Primarily it develops in the sphenoid and erodes the posterior ethmoidal cells, or in the presence of an anatomic variant, develops in an intermediate cell and spreads centrifugally. The majority of authors favour extension of local inflammation from a neighbouring part which causes partial, intermittent or complete osteal block leading to raised intrasinus pressure, followed by pressure necrosis of the bony walls with subsequent extension through the paths of least resistance, viz. extradural spaces, superior orbital fissure and optic canal. According to Dabney (1921), it is 'An accumulation and retention of mucous secretion within the accessory nasal sinus, generally associated with a distension of one or more of its walls'. He says that cystic dilatation of one or more mucous glands and cystic degeneration of the mucosa, or polyps in the sinus can give rise to mucocoele. Occlusion of the osteum by chronic inflammatory changes of the membrane or by tumour, congenital narrowing of the osteum and absence of communication between the sinus and nose have been suggested as possible causes (Thomson and Negus, 1955). This blockage is unlikely to be a basic cause. Tamari and O'Neil (1949),

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feel, on the findings of Turner and later Dabney who found that in a significant number of cases the osteum was patent and adequate for normal drainage, that it is due to submucosal injury due to trauma, haemorrhage or infection with an accumulation of exudate beneath the mucosa and as the exudate increases, the elevated mucosa herniates towards the sinus cavity. Howarth (1946), supports this view. In many cases the ostea have been found blocked but no mucocoele was present. Killian and others (quoted by Neffson, 1967), suggested injury as a cause, though Turner presented 10 cases of frontal and maxillary sinus mucocoeles in which there was no history of trauma. Nevertheless trauma appears to be one of the important factors. Schüller discussed the possible development of a hypophyseal cyst from the inner or under half of the sella turcica itself. There is no adequate proof of this concept. Linthicum *et al.* (1946), concludes that the onset of these cases is often so gradual, and the course so slow, that it is difficult to assign a definite cause in an individual case, much less accept any single aetiologic factor which is common to all of them. It appears that a combination of different factors is responsible for this condition.

Case report

Name: P.K.B. Age: 40 Sex: Male.

The patient presented himself with a two-year history of intermittent bifrontal headache spreading towards the temples, throbbing in nature, associated with nausea at the height of headache but no vomiting. He used to get throbbing pain behind the eyes, more particularly the right one. Headache was worse early in the morning and at about 1 p.m. For ten days prior to admission to this hospital headache became more or less continuous and so intense as to interfere with sleep. To start with, the headache used to be present for about an hour or two and be relieved with aspirin. He had a feeling of tension in the posterior neck muscles. At times he experienced offensive smell in the nose, both sides. There was no history of diminution or loss of the sensation of smell. No engorgment of vessels was noticed in the head region during the headache and no visual disturbances were present. There was neither any history of fits, fever, diplopia and head injury in the past, nor any suggestive family history of vasomotor disturbances.

Past History

He had been suffering from recurrent sinusitis for the past 10 to 15 years and had had several antrum punctures done irregularly on both sides which produced pus on several occasions. He had an attack of acute thyroiditis two years back and he reported that since that attack he has been experiencing the above symptoms. He attributes his present symptoms to the attack of thyroiditis.

On Examination

He was a healthy looking adult. He appeared very apprehensive. There was no anaemia, jaundice or oedema. General examination did not show any abnormality. No lymph nodes were palpable in the neck or anywhere else in the body. He was myopic (–9 diopters both eyes). Neurological examination

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revealed questionable fullness of the cups on fundoscopy. The right lateral rectus showed questionable paresis but no diplopia on looking to either side. Pupils were normal. The other cranial nerves were normal. Motor and sensory systems did not show any abnormality. No positive cerebellar signs were present. Reflexes (superficial and deep) were normal and equal on both sides. Perimetry-normal. Head and spine were normal. Respiratory, cardiovascular and gastro-intestinal systems were normal. The patient was admitted under the service of the neurosurgeons.

X-rays

X-rays of the spine (cervical) revealed no pathology. Right lateral stereo of the skull showed 'marked ballooning of the pituitary fossa. There is erosion of the dorsum sellae and there is encroachment of the floor of the pituitary fossa. The sphenoid air sinus is seen only in its anterior part. The left anterior clinoid process is less prominent than the right one. Possibly there is a linear intrasellar calcification. The posterior clinoid processes seem to be lifted up.'

Impression: X-ray skull is consistent with intrasellar space occupying lesion.

Right carotid angiogram 'Shows slight widening of the carotid siphon. The anterior and the middle cerebral groups of vessels seem to be normal. Posterior communicating and posterior cerebral arteries are normal. The capillary and venous phases do not show any abnormality.' Anteroposterior film shows: 'The anterior cerebral artery seems to be slightly shifted to the right side in its proximal course. Venous phase does not show any abnormality.' Submentovertical view 'Possible shift of the internal carotid artery to the lateral side.'

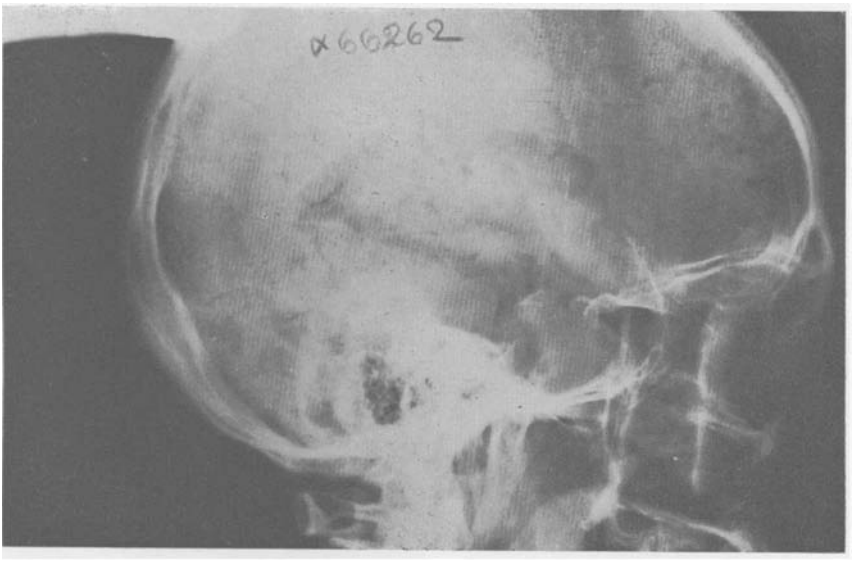


FIG. 1.
Lateral X-ray, pre-operative.

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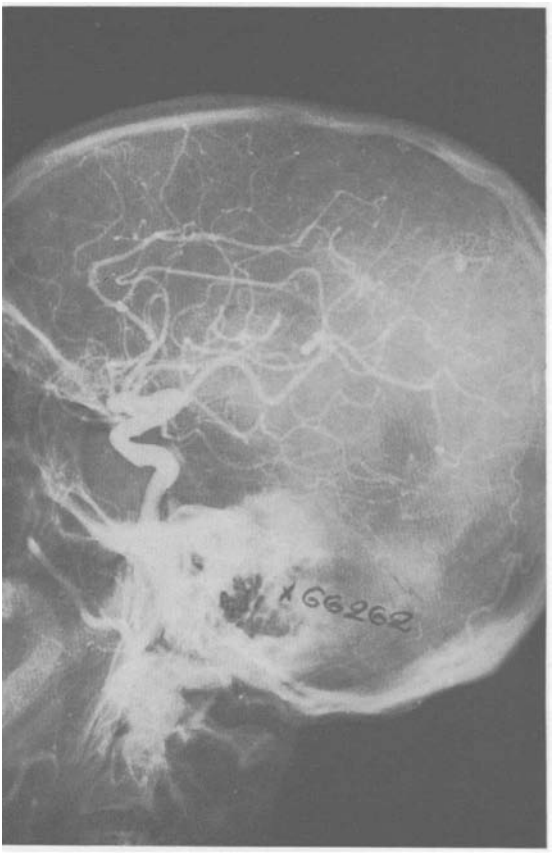


FIG. 2.
Lateral view, carotid angiogram, showing slight elevation and compression of carotid siphon.

Impression: Angiographic films are not very diagnostic of pituitary or parapituitary tumour.

Fractional pneumo-encephalogram shows good visualization of the IVth ventricle, aqueduct, IIIrd ventricle and both lateral ventricles. The interpeduncular cisterns seem to be normal in position. Subarachnoid spaces are also filled which seem to be normal.

Laboratory Investigations:

C.S.F.: Pressure: Normal Total Cell 2/cu. mm.
Protein 30 mg. per cent.
Chloride 720 mgm. per cent.
Sugar 85 mgm. per cent.

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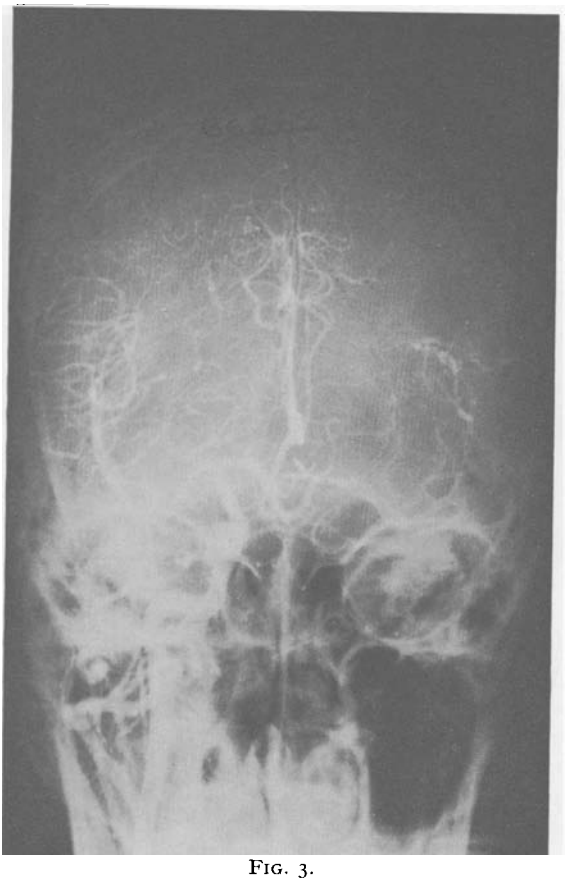


FIG. 3.

Carotid angiogram showing slight elevation of carotid syphon. No other abnormality.

Blood:

V.D.R.L. Negative.
W.B.C. Total 5,600/cu. mm.
Differential:
Poly: 60 per cent.
Eosino: 11 per cent.
Lympho: 28 per cent.
P.C.V. 47 per cent.
E.S.R.: 1st hour 14 mm., 2nd Hour 38 mm.

Urine and Stool

Nothing abnormal detected.
Serum cholesterol: 220 mg. per cent.
Total 17—ketosteroids excretion 10 mg./24 hours.
Total volume of Urine—1,700 ml./24 hours.
Blood Sugar (Random) 90 mg./100 cc.
E.E.G.: Normal.

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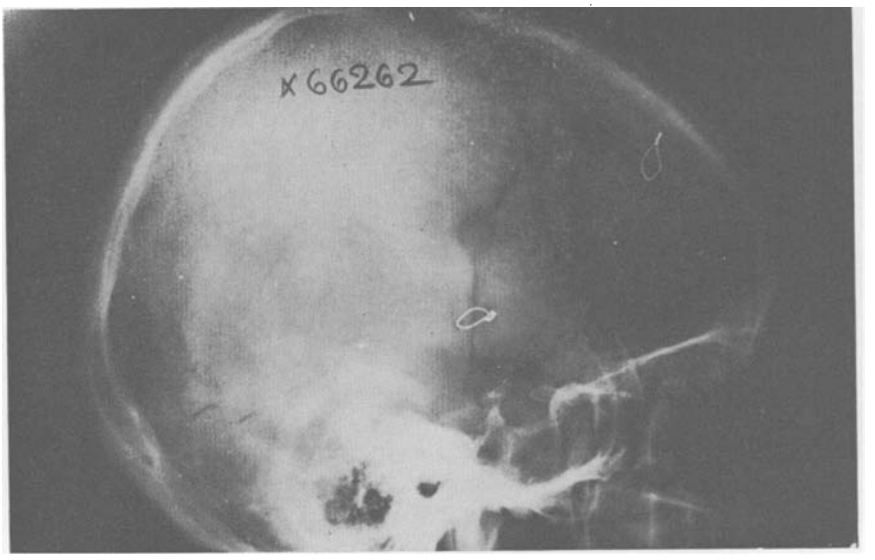


FIG. 4.
Lateral X-ray eight months after operation.

On 18 March 1964 under general anaesthesia, *right frontal craniotomy and exploration of the pituitary fossa were done.* There was no abnormality.

The symptoms were not relieved. The patient was very much disappointed, and was fixed in his mind that he had a cancer in the head and would die inevitably. He was assured that it was very unlikely to be cancer because no cranial nerves were involved in spite of the skiagraphic evidence of so much bone destruction.

The patient was next referred to the E.N.T. department. Preoperatively he was not referred to the department of Otolaryngology in spite of a history of sinusitis and occasional cacosmia.

On Examination

Ears: N.A.D.

Nose: Right turbinals were slightly swollen and congested, but shrinks well.

Nasopharynx: A little pus on the roof. A small bulge just behind the vomer was seen, the mucosa over it was intact. Olfactometry was not done. On review of the X-ray films a provisional diagnosis of sphenoidal mucocoele was made. The impression that the sphenoid sinus is seen only in the anterior part was obviously erroneous. X-ray of the paranasal sinuses was ordered.

X-ray of paranasal sinuses; Waters view:

'Right maxillary sinus is opaque: the right sphenoid sinus appears bigger.'

On 10 April 1964 the right sphenoid was explored through the right external ethmoidectomy approach under hypothermia. There was hardly any bleeding.

Operative Findings

On reaching the anterior wall of the sphenoid no osteum was found. How-

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ever, the wall was very thin, soft and yielding. The anterior wall was removed, and the sinus entered. It was filled with straw coloured thick tenacious material. The sinus was enormous in size and the septum had been destroyed or the opposite sinus completely obliterated. After aspirating the contents, the thickened mucosal lining of the sinus was curetted away and the tissue sent for histopathological examination. No tumour was encountered. After completing the curettage a fenestrated rubber drainage tube was put into the sinus and brought out through the nose. The sinus was then loosely packed with sulphanilamide petroleum jelly gauze which was brought out through the nose. The incision was then closed with interrupted cotton sutures.

Histopathology Report Biopsy No. 2676/64.

Macro: Two flattened pieces each measuring about 1 cm of soft tissue.

Macro: Section shows dense connective tissue containing scattered lymphocytes and plasma cells and a few pigmented cells. A small area is covered by flattened squamous epithelium. There is no evidence of malignancy in this specimen.

Impression: Chronic inflammatory lesion.

Post-operatively there was slight rise of temperature. Headache was as before but no nausea.

Second post-operative day:

Six inches of the packing removed. Drainage tube sucked out. There was mucosanguineous discharge.

Third Post-operative day:

Rest of the packing removed.

Fourth Post-operative day:

Drainage tube sucked, practically no discharge. Patient was feeling much better and cheerful.

Fifth Post-operative day:

Half-inch tube removed, sucked.

Nasopharynx: The postvomerine swelling was absent perhaps due to generalized swelling.

Sixth Post-operative day:

Tube and stitches were removed. Nose, after suction, looked fine.

Seventh Post-operative day:

Headache was much less. Nose: hardly any discharge, no cacosmia was present.

Eighth Post-operative day:

Nose looked fine. There was no discharge at all. He was discharged with a feeling of slight pulling or tension behind the nose and neck (posterior aspect).

Follow-up

The patient came for a check-up after 8 months with no symptoms at all. Check X-ray revealed, 'as compared to the pre-operative X-ray, the appearances are much improved. The base of the sella is reforming. The sphenoid appears to have some amount of air in it.'

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Discussion

Since most of the cases have been wrongly diagnosed pre-operatively, we like to discuss the diagnostic features. If one keeps in mind the possibility of sphenoid mucocoele, it is not impossible to diagnose the cases correctly and in this case we are reporting, one could make a tentative diagnosis if one was conscious about this.

The signs and symptoms are due to the local cause and extension of pathology beyond the confines of the sphenoid sinus. These for the most part however, arise from pressure on the neighbouring structures transmitted to or from the orbital cavity through the superior orbital fissure and the optic foramen at the orbital apex. These structures include (i) Oculomotor nerve, (ii) Trochlear nerve, (iii) Ophthalmic division of trigeminal nerve, (iv) Abducent nerve, (v) Some filaments from the cavernous plexus of the sympathetic nerves, (vi) Orbital branch of the middle meningeal artery, (vii) A recurrent branch from the lachrimal artery to the dura, (viii) Ophthalmic veins, (ix) Optic nerve, (x) Ophthalmic arteries (Cody and Hallberg, 1959; Proetz, 1948).

Usually there is history of headache (in the vertex or frontal, temporal or occipital region), anterior and/or post-nasal discharge, cacosmia or anosmia, nasal obstruction and previous attacks of sinusitis. The headache is often described as pressure headache. Peculiarly the left side is more affected. Eye symptoms include mainly diplopia, blurred vision and even complete loss of vision. There may be pain in and behind the eye. Litwimowicz and Neffson (1957), stressed the occurrence of amaurosis after the onset of headache. There may be proptosis, unilateral exophthalmos (may be pulsatile) (Robertson, 1949; O'Shea, 1932). and ptosis. Fluctuations of all these symptoms are frequent (Cody, 1959). There may be facial pain due to pressure on the Gasserian ganglion. There may be a history of attacks of unconsciousness due to pressure in the region of the IIIrd ventricle (Tamari and O'Neil, 1949; Neffson, 1967). Olfactory disturbances and convulsions also have been reported. The pulsation, sometimes present in the sphenoid region has been found after opening the sinus and was ascribed to the exposure of the dura and close proximity of large vessels and the cavernous sinus. Ophthalmologic examination may reveal exophthalmos; extra-ocular movements may be normal, impaired or absent, depending on the extent of involvement of the oculomotor nerves.

Fundoscopy: Usually normal: but early in the course of the disease optic neuritis or retrobulbar neuritis may be suspected and later on optic atrophy discovered.

X-ray findings: (Robertson, 1949; Simson, 1955; Wigh, 1950).

There should be satisfactory demonstration of the superior orbital fissure, optic foramina, lateral walls of the ethmoid sinuses, floor of the sella turcica, walls of the sphenoid sinuses and walls of the orbits. One may find the following abnormalities:

1. Enlargement and irregularity of the superior orbital fissure.

2. Lateral displacement of lamina papyracea or destruction (loss of marginal continuity which in some projections may be mistaken for a calcified structure, e.g. a vessel or an aneurysm).

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3. Erosion and destruction of the floor of the sella turcica; occasional concomitant changes in the anterior and posterior clinoid processes.

4. Loss of floor of optic foramen. There may be widening and irregularity of the foramen.

5. Lateral wall of sphenoid: enlarged and destroyed:

6. Lateral X-ray shows anterior wall of affected sphenoid sinuses closer to the anterior nasal spine than the other.

7. Tomogram (antero-posterior laminogram) may show destruction of jugum sphenoidale, irregularity and destruction of lamina papyracea and lateral sphenoidal wall and depression of floor of sphenoid sinus. Lateral laminograms should also be taken.

The following diseases may simulate sphenoidal mucocoele:

(i) Congenital anomalies: defective development of sphenoid bone is usually associated with unilateral pulsating exophthalmos (O'Shea, 1932; Robertson, 1949). Here visual disturbances are not necessarily present. X-ray shows enlarged orbit with depression of its floor; absence of shadow corresponding to orbital plate of frontal bone and wings of sphenoid, narrowing of ethmoidal cells medial to orbit and the appearance of the lateral wall of the sphenoid closer to the median line. The optic foramen, foramen ovale and foramen spinosum may be absent. One is able to differentiate the pulsating exophthalmos associated with defective development of sphenoid bone from arteriovenous aneurysm by the following points, viz. absence of distressing symptoms, the presence of slow deliberate pulsations and the absence of bruit. In arteriovenous aneurysm the pulsations are less marked and there is often dilatation of retinal and neighbouring vessels. In orbital meningocoele or encephalocoele, usually the cough impulse is present and the swelling is often compressible and the pneumoencephalogram may show presence of gas extending into the orbit from the cranium through a gap between the cribriform plate and the orbital plate of the frontal bone, and between the orbital plate of frontal bone and lamina papyracea. Since the defect is anterior, the eye is pushed laterally and downwards but is not unduly prominent. Dermoid and epidermoid tumours also can give rise to pulsating exophthalmos. X-ray shows circumscribed dehiscensces with smooth margins and increased density of the surrounding bone.

(ii) Acquired causes: Metastasis and xanthomatosis produce usually irregular erosion of bones seen on skiagraphy or felt. Optic nerve and chiasmatic glioma cause circular enlargement of the optic canal. One has to keep in mind retinoblastoma and neurofibromatosis. Meningioma of the sphenoid ridge, especially in its medial part, which is a common erroneous diagnosis of sphenoidal mucocoele, causes irregular enlargement and thickening of optic canal. Suprasellar, intrasellar and parasellar tumours probably are among the most difficult to diagnose. In pituitary tumour, destructive changes localized to the inferior strut of the optic canal are most significant. Aneurysm of the extradual portion of the carotid siphon often erodes the strut.

Comments

In this case with the history of throbbing and bursting headache, post-nasal discharge, cacosmia, pain behind the eyes assoicated with a long history

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of recurrent attacks of sinusitis and absence of eye field or fundal changes and neurological findings one should have thought about the possibility of sphenoidal sinus involvement. Whenever X-ray shows erosion of the dorsum sellae, Water's view with mouth open and submento-vertical view should be taken to see the sphenoid sinuses. Caldwell's view and views to show superior orbital fissures and optic foramina should also always be included. A lateral view to show the ethmoid cells and the walls of sphenoids is helpful. If there is suspicion on the above investigations, one should washout the sphenoid sinuses and examine the washing for culture and sensitivity and presence of cholesterol crystals. Following washing, X-ray pictures with contrast medium introduced into each sphenoid sinus separately may help in diagnosis. In this case the aetiology appears to be the closure of the osteum due to chronic inflammation.

Summary

A case of right sphenoidal mucocoele, possibly due to osteum blockage, is presented with comments and review of literature.

Acknowledgements

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REFERENCES

ANTHONY, W. P., and WILLIAMS, H. L. (1951) *Archives of Otolaryngology*, **53,** 189.
CODY, D., and HALLBERG, O. (1959) *Archives of Otolaryngology*, **70,** 495.
DABNEY, V. (1921) *New York J. Med.*, **114,** 619.
HOLT, H., and ROTTH, A. (1940) *Archives of Ophthalmology*, **24,** 731.
HOWARTH, W. G. (1946) *Lancet*, **2,** 744.
LINTHICUM, F. H., RAND, C. W., and REEVES, D. L. (1946) *Journal of Neurosurgery*, **3,** 444.
NEFFSON, A. H. (1967) *Archives of Otolaryngology*, **66,** 157.
O'SHEA, H. V. (1932) *Lancet*, **1,** 253.
PROETZ, A. W. (1948) *British Medical Journal*, **2,** 243.
ROBERTSON, E. G. (1949) *American Journal of Roentgenology*, **62,** 44.
SIMON, H. M., and TINGWALD, F. R. (1955) *Radiology*, **64,** 538.
SMITH, A. T. (1958) *Annals of Otology, Rhinology and Laryngology*, **67,** 742.
TASSMAN, I. S. (1947) *Archives of Ophthalmology*, **37,** 294.
TAMARI, N. J., and O'NEIL, J. J. (1949) *Journal of Laryngology and Otology*, **63,** 24.
WILKERSON, W. W. (1945) *Laryngoscope*, **55,** 294.
WIGH, R. (1950) *Radiology*, **54,** 579.

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