

An Unusual Presentation of a Giant Frontal Mucocoele Manifesting with Frontal Lobe Syndrome

Dear Editor,

Mucocoeles develop from paranasal sinuses. They are epithelium-lined, mucus containing lesions and present as slowly enlarging masses. The frontal sinus is most commonly involved, whereas sphenoid, ethmoid and maxillary mucocoeles are rare.¹ Accumulation of mucus secretion, generally, is caused by paranasal sinus inflammation, fibrosis, trauma, previous surgery, anatomical abnormality, or mass lesion such as osteoma. Mucocoeles can spread both intraorbitally and intracranially.^{2,3}

Frontal lobe syndrome (FLS) was first defined in 1868 by Harlow.⁴ FLS is a pattern of signs and symptoms associated with the damage of the frontal lobe.⁵ Typically, FLS includes general impairment of planning functions, boastfulness, lack of inhibition, hypomanic episodes, impulsiveness, anti-social behaviour, depression, apathy and perseveration.⁴ Manifestations of a frontal lobe syndrome depend on many factors: patient's baseline intelligence, education, site of lesions, age, sex and function of non-frontal brain regions.⁴ Causes of FLS include mental retardation, cerebrovascular disease, head trauma, brain tumours compressing the frontal lobe parenchyma, brain infections, neurodegenerative diseases and normal pressure hydrocephalus.^{4,5}

A 68-year-old male complained of a worsening headache for 3 to 4 weeks prior to admission. He had diplopia for 1 to 2 months. Proptosis, periorbital swelling and exophthalmus were other findings. There was no history of surgery or trauma. His relatives stated that he had personality and behavioural alterations recently. The patient complained of memory disturbances including inability to recall long-known and major details such as his own address, perception difficulties and impaired long and short-time memory.

Neurological examination revealed unilateral papilledema in fundoscopy. Ocular movements were limited at the right side. Memory evaluation tests were performed (Wechsler Memory Scale-Revised test). Verbal and visual memory scores were low. These findings supported that his behaviour alterations were due to memory dysfunction.

Thyroid function tests, white blood cell count, C-reactive protein and B-12 levels were normal and serology for syphilis, tests for HIV or connective tissue disorders were negative. Laboratory examinations did not suggest infection or another systemic disease like thyrotoxicosis.

Radiological imaging using computed tomography (CT)

and magnetic resonance imaging (MRI) were performed. CT showed a low density homogeneous mass in the right frontal sinus. In soft tissue window scans (Figs. 1 a-b), it was seen that the mass had eroded the posterior wall of the frontal sinus and it had a large extension into the anterior cranial fossa. The posterior margin of the lesion was partially surrounded by bony fragments. The lesion had severely compressed the right frontal lobe and orbital components.

On MRI, T1-weighted axial and coronal scans showed that the internal signal of the lesion was hyperintense compared to the brain parenchyma. It had compressed the brain tissue and extended towards the anterior cranial fossa. Bulbus oculi was displaced anteriorly and laterally. At T2-weighted axial scans (Fig. 2), the lesion was found extending intracranially and it was hyperintense compared to the brain. The mucocoele's dimension was 50 x 60 x 70 mm. It had caused subfalcian herniation and had compressed the lateral ventricle. Based on the clinical and radiological findings, the most likely diagnosis was frontal mucocoele. Diagnosis of frontal mucocoele was confirmed at surgery.

Frontal lobe syndrome is a disorder affecting the prefrontal areas of the frontal lobe.⁴ The prefrontal lobe comprises the vast area of the frontal lobe anterior to the motor cortex and includes the undersurface of the frontal lobe or the orbital region.^{4,5} The frontal lobe syndrome is said to be present when an individual who is previously capable of judgement and sustained application and organisation of his life becomes aimless and improvident, and may lose tact, sensitivity and self-control.⁴ Additionally, the individual affected by pathology in the prefrontal cortex may demonstrate impulsiveness and a failure to appreciate the consequences of his or her reckless behaviour.⁴ Frontal lobe syndrome can be caused by head trauma or may be the consequence of a brain tumour, a cerebrovascular accident, infection or a degenerative cortical disease such as Pick's disease.^{4,5} This syndrome represents an organic explanation for psychologically-based symptoms a patient may demonstrate.

Clinical presentation of the mucocoeles varies from asymptomatic to incapacitating headache and visual disturbance.^{1,3} Proptosis (83%) and diplopia (45%) are the most common complaints.³ On physical examination, periorbital tenderness, swelling, chemosis, decreased visual acuity and extraocular movement can be determined.³ In the literature, there was an unusual giant mucocoele,

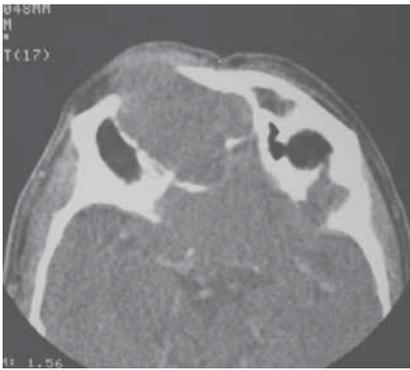


Fig. 1a.

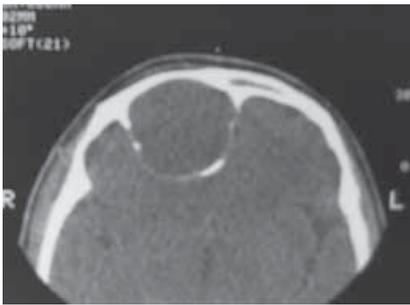


Fig. 1b.

Figs. 1a-b. CT (soft tissue window-series from 2 different level) shows extension of the frontal sinus mass inferiorly into the orbit as well as superolaterally, eroding the frontal bone. Mass heavily compressing the brain parenchyma, orbital components and causing exophthalmus.

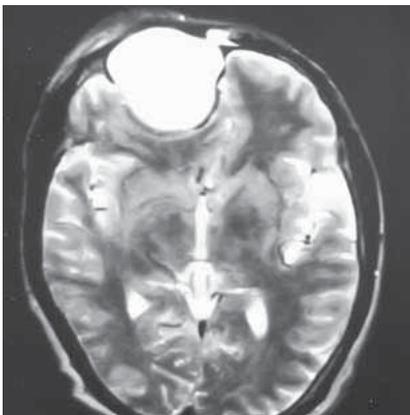


Fig. 2. T2 weighted axial MRI demonstrating that the mass is hyperintense compared to the brain, extending into the orbit and cranium, strongly compressing the right frontal lobe and causing exophthalmus.

manifesting as a subcutaneous forehead mass.¹ In our case, a giant mucocoele was present at the anterior cranial fossa, compressing the frontal lobe. Mildly increased intracranial pressure was determined. Headache, recently worsening exophthalmus and behavioural disturbance were the other clinical findings. We could not find a similar case in the literature similar to ours in which the frontal sinus mucocoele caused behaviour, memory and emotional alterations.

There are 3 criteria for CT diagnosis of a mucocoele: homogeneous isodense mass, clearly defined margin, and patchy osteolysis around the mass.¹⁻³ Erosion of the sinus wall with marginal sclerosis is also an indicative finding.³ Typically, mucocoeles tend to be fairly bright on T1W images compared to the brain, and iso-hyperintense on T2W images.³ It is pathognomonic MRI finding for mucocoeles.³ Neoplastic processes tend to be isointense relative to the brain on both T1 and T2W images.³ Hyperintensity on T1W images suggests proteinous or haemorrhagic content of a lesion.¹⁻³ This may lead to mis-diagnosis. One of the other pitfalls of MRI in the diagnosis of mucocoeles is that if it contains inspissated proteinaceous content, it could become almost of void signal on T1W and T2W images, like that of air.³ This would make it difficult to detect on MRI alone. On the CT, however, the inspissated content would be of high density, making diagnosis straightforward.³ CT and MRI are complementary in complicated cases.¹

Dermoid cysts, histiocytosis, fungal and tuberculosis infections, frontorbital cholesterol granuloma and other uncommon neoplasms must be considered in the differential diagnosis.^{1,3} Because of higher hyperintensity from other processes on T1W images, the differentiation is easy on MRI.³

We presented a rare cause of FLS with giant frontal mucocoele. To our knowledge, this has not been reported before in the literature.

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