

Giant Intradural Mucocele in a Patient with Adult Onset Seizures

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Key Words

Mucocele · Mucopyocele · Epilepsia

Abstract

A rare case of mucopyocele in a patient who presented with epileptic seizures is reported. The computed tomography scan (CT) and the magnetic resonance (MR) imaging revealed an intradural extension of a giant fronto-ethmoidal mucopyocele, eroding the cribriform plate and compressing both frontal lobes. The lesion was removed by craniotomy with elimination of the mass effect and reconstruction of the anterior skull base. An intracranial-intradural mucopyocele is an extremely rare cause of generalized convulsion as a presenting symptom, with only 6 cases reported in the literature. The total removal of the lesion associated with anterior fossa reconstruction is the treatment of choice.

Introduction

Mucocele is a chronic, expanding, mucosa-lined lesion of the paranasal sinus characterized by mucous retention that can be infected, becoming a mucopyocele [1]. Although benign, mucoceles have a tendency to expand by eroding the surrounding bony walls, which displaces and destroys structures by pressure and bone resorption [2]. They originate from obstruction of the sinus ostium by congenital anomalies, infection, inflammation, allergy, polypoidiasis, trauma (including surgery), functional endoscopic sinus surgery (FESS) and benign or malignant tumors [1]. The frontal sinus is most commonly involved and the sphenoid sinus most rarely. Their intracranial and/or intradural extension is uncommon. We present a patient with recent onset seizures due to a giant intradural mucopyocele. The patient was treated with frontal craniotomy, total surgical expansion of the lesion and repair of the dural and bony defect.

Case Report

A 50-year-old female patient initially admitted to the Otolaryngology Department with clinical and radiological signs of frontal sinusitis (i.e. fever and frontal headaches) was transferred to the Neurosurgery Department following seizures. She was put on antibiotics and treated conservatively. Further radiological investigations were required following the onset of generalized tonic-clonic epileptic seizures. Neurological examination was unremarkable. Fundoscopic examination detected no papilledema. Intranasal examination by fiberoptic scope disclosed no evidence of paranasal infection. Skull radiography showed opacification of the right frontal sinus. The brain CT scan showed two homogeneous isodense masses, one in the right frontal sinus, which eroded the posterior wall of the frontal sinus and formed a large extension into the anterior cranial fossa, and a second which arose from the ethmoid sinuses and through the cribriform plate extending into the frontal cranial fossa with serious compression at the frontal lobes. Brain magnetic resonance (MR) imaging revealed two large round lesions in the frontal cranial fossa. On T1-weighted images, the first lesion had high signal intensity and was in direct contact with the wall of the right frontal sinus, while the second lesion had intermediate signal intensity and arose from the ethmoid sinuses, projecting upwards to the cranial fossa. In T2-weighted images, the lesions appeared with intermediate signal intensity. Sagittal MR imaging showed that the lesions were intradural and compressing both frontal lobes of the brain. Pre-operative EEG disclosed focal theta activity and epileptiform discharges over the frontal leads bilaterally. The patient's past medical history included a tonsillectomy in childhood, a car accident with cranial trauma 33 years ago, and a frontal sinusitis 16 years ago. She did not suffer from headaches, while no behavioral changes were reported by her relatives. She was allergic to penicillin.

The patient was operated upon and bifrontal craniotomy was performed. The two lesions were completely removed by suction. The mucous membranes of the frontal sinus were curetted and the sinus was packed with muscle. The first lesion contained pus and the other one mucous secretion. The dura mater over the frontal base and the cribriform plate bone were eroded. The dural defect was repaired with pericranium. Cultures of the pus were negative. Histopathology showed that mucosa was partially lined by respiratory epithelium, partially showed ulcer with chronic inflammatory cells, pus and reactive foaming histocytes, compatible with mucopyocele. Postoperatively, the patient was asymptomatic without seizures and with no medication. She was discharged after 14 days without neurological deficits. A repeat EEG on day 14 disclosed a significant reduction of theta activity and elimination of the epileptiform discharges.

Discussion

Mucoceles are benign, slowly growing epithelial cysts. They arise from the gradual accumulation of mucus material, causing progressive enlargement of the sinus cavity. They may grow sufficiently to compress orbital or intracranial structures [10]. They can be infected, becoming mucopyoceles. This occurs more often in the frontal (65%) and ethmoid sinuses (25%), rarely in maxillary sinuses (10%), while it is extremely rare in the sphenoid sinus [4]. Intracranial and especially intradural extension of mucoceles and mucopyoceles is rare.

They may present with multiple symptoms, such as proptosis with or without pain, periorbital swelling, exophthalmos, diplopia and decrease of visual acuity, acute onset of blindness, visual field abnormalities, dizziness, nausea, vomiting, anosmia, nasal obstruction, intermittent epistaxis. Very rarely, transient hemiparesis due to stenosis of a cerebral artery, meningitis, meningoencephalitis pneumocephalus, brain abscess, CSF fistulas and seizures may occur. In the literature, 6 cases with mucopyocele and recent onset seizures are reported.

CT has proven to be an excellent diagnostic tool and is essential in surgical planning. There are three major CT criteria in the diagnosis of mucocele: homogeneous isodensity mass, clearly defined margin and patchy osteolysis around the mass [11]. MR imaging may provide additional information by the examination of the orbit and may be the preferred imaging technique if other soft tissue tumors causing proptosis cannot be

excluded [5]. The signal intensity varies greatly due to protein content [6, 7]. Mucocele presents with intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images, reflecting increased water content [8]. The mucopyocele, due to increased protein content in the presence of macrophages which contain iron, may exhibit paramagnetic behavior with high signal intensity on T1-weighted images and low intensity or vacuum signal on T2-weighted images and STIR [9]. MR imaging is useful in detailing the relationship between intracranial, intraorbital structures and mucocele. Radiological differential diagnosis includes dermoid cysts, parasite infections (histiocytosis), fungal infection, extra-axial gliomas and post traumatic porencephaly. However, the final diagnosis relies on the histological examination of the lesion.

In our case, in agreement with those reported in the literature, late-onset generalized epileptic seizures were caused by a mucopyocele and were completely cured after surgical removal of the lesion. No further anti-epileptic treatment was required.

We conclude that mucopyocele should be included in the differential diagnosis of epileptic seizures, since it constitutes a treatable condition.

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