A giant intracranial mucocele associated with an orbitoethmoidal osteoma

Case report

YOSHIKAZU NAKAJIMA, M.D., TOSHIKI YOSHIMINE, M.D., MAKOTO OGAWA, M.D., MAYAKO TAKANASHI, M.D., KANA NAKAMUTA, M.D., MOTOHIKO MARUNO, M.D., HIROSHI HASEGAWA, M.D., AND JUNICHIRO YOKOTA, M.D.

Osaka Prefectural Senshu Critical Care Medical Center; Department of Neurosurgery, Osaka University Graduate School of Medicine; Department of Neurology, Izumisano Municipal Hospital; and Department of Neurosurgery, Osaka Kosei Nenkin Hospital, Osaka, Japan

The authors present a rare case of a giant intracranial mucocele associated with an orbitoethmoidal osteoma in a patient suffering from a generalized convulsive disorder. The broad pedicle of the osteoma had penetrated the cribiform plate and extended intracranially to form a nodular mass in the olfactory groove. The intracranial portion of the osteoma was surrounded by a mucocele. Both the cyst wall and multilayered intracystic septations of the mucocele were indented by layers of the osteoma. Although the extracranial portion adhered to the mucosa of the ethmoidal sinus, there were no signs of sinus obstruction. No direct communication other than the osteoma was identified between the mucocele and the ethmoidal mucosa. The large cerebral defect, which the mucocele occupied, communicated directly with the lateral ventricle without any intervening membranous structures. A frontal craniotomy is recommended for exposure of the lesion and plastic repair of the dural defect.

Key Words • mucocele • osteoma • cranial base tumor • intracranial extension

A PARANASAL osteoma that projects into the frontal fossa has been previously reported to cause an intracranial mucocele as well as CSF rhinorrhea and pneumocephalus. We treated a rare case of a giant intracranial mucocele associated with an orbitoethmoidal osteoma in a patient suffering from a generalized convulsive disorder.

Case Report

History and Examination. This 46-year-old man presented with a sudden-onset, generalized tonic convulsion that persisted for 30 minutes, and he was transferred to our hospital for treatment. He had no history of seizure and his family history was unremarkable. He was alert and displayed no focal neurological signs. Electroencephalographic studies revealed a theta burst in the left frontal region. The results of his CSF culture were normal.

Radiological Findings. Plain cranial x-ray films demonstrated a nodular calcified mass in the left olfactory groove (Fig. 1 left). A CT scan revealed a large cyst in the left frontal lobe and a calcified mass on the medial aspect of the cyst (Fig. 1 center and right). Three-dimensional and multiplanar reconstructed CT scans revealed that the calcified mass had penetrated the cribiform plate with a broad pedicle and was protruding into the ethmoidal sinus and the orbital cavity. No enhancement was detected in any part of the calcified mass. A large cystic mass that bulged into the ventral aspect of the left frontal lobe was revealed on MR imaging (Fig. 2). The signal intensity of the cyst contents was slightly higher than that of CSF on T1-weighted MR images and noticeably higher on T2-weighted images. No anatomical structures were recognized between the cyst and the ventricular system. There was a marginal rim around the cyst and weak pericystic edema in the left frontal lobe. There were no signs of inflammation or obstruction in the paranasal sinuses. Digital subtraction angiography revealed shifts in the cortical vessels, indicating large intraaxial masses.

Operation. The patient underwent a left-sided frontal craniotomy for total removal of the mass and skull base reconstruction. The cyst was exposed by uncapping the overlying thin cerebrum (Fig. 3 upper left). The cystic cavity was compartmentalized into multilayered septations that were indented by layers of the nodular calcified mass (Fig. 3 upper right). An arachnoid-like membrane that appeared to be continuous with the ependymal lining covered the surface of the defective cerebrum. A thin layer of dark-brown greasy material overlying the cyst pre-
vented adhesion between it and the surrounding brain. The cyst had occupied the cerebral defect, which communicated directly with the left lateral ventricle without any intervening membranous structures (Fig. 3 lower). The calcified mass and cyst were removed en bloc after drilling the cranial base with a surgical burr. The inferior part of the calcified mass had adhered to the mucosa of the ethmoidal sinus. The bone defect was closed with a flap of the pericranium, which was sutured to the dura, and the dural defect was repaired with a fascial patch taken from the temporal muscle.

Pathological Findings. The cyst was attached to the intracranial portion of the calcified mass. Both the cyst wall and multilayered intracystic septations were indented by layers of the hard calcified mass. No communication besides the calcified mass was identified between the cyst and the ethmoidal mucosa (Fig. 4 left). The cyst contents consisted of a lemon-colored, nonturbid fluid (Fig. 4 right). Biochemical analysis showed that the fluid was made up of 2000 mg/dl protein, 63 mg/dl glucose, 144 mEq/L sodium, 2.4 mEq/L potassium, and 112 mEq/L chlorine. The cell count was 6/mm³. Cytological examination revealed no atypical findings, and results of a bacteriological examination were negative. Histological investigation revealed that the calcified mass was an osteoma (Fig. 5 left). The cyst wall was covered with ciliated columnar epithelium (Fig. 5 right). The intracystic septa were also composed of respiratory epithelium and connective tissues.

Postoperative Course. The patient’s postoperative course was uneventful except for a mild chemical meningitis that lasted for a few weeks. The patient returned to his work and normal lifestyle 1 month later.

Discussion

There have been several reports of paranasal osteoma extending intracranially, and in most cases, the lesion was
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diagnosed on the basis of clinical symptoms associated with intraorbital extension or intracranial complications such as CSF rhinorrhea, pneumocephalus, and intracranial infection. In our present case, a giant mucocele was found in the left frontal region and was associated with a paranasal osteoma in a patient suffering from a late-onset generalized convulsive disorder. There was no evidence of CSF leakage.

In our patient, the intracranial surface of the osteoma was surrounded by the mucocele, and the opposite surface was attached to the mucosa of the ethmoidal sinus. This pathological anatomy is consistent with those previously reported. More precisely, our case was similar to one reported by Campbell, et al. According to their report, a mucocele was attached to the intracranial portion of a paranasal osteoma at the upper margin of the cone-shaped dural collar, and dural perforation into the lateral ventricle resulted in ventricular pneumocephalus. The difference between their case and ours was that in ours the dura was attached to the osteoma around the penetration site of the skull base, and no dural perforation was recognized.

In our case, layers of epithelial membranes (the cyst wall and intracystic septa) were indented by layers of the osteoma, and the extracranial portion of the osteoma adhered to the ethmoidal mucosa. There was no anatomical continuity other than that created by the osteoma between the mucocele and the ethmoidal sinus. This unusual pathological anatomy indicates that the growing orbitoethmoidal osteoma had isolated the developing mucocele from the ethmoidal sinus involving the mucosa.

We presume that the mucocele in our patient had been developing very slowly because it caused a large cerebral defect without any serious symptoms before the generalized convulsion. The cerebral defect was in communication with the lateral ventricle without any intervening membranous structure such as the ependyma or glial membranous tissues. Longstanding direct compression and other influences of a mucocele may disrupt the ependymal linings. During the past 75 years, ventricular perforation associated with frontal paranasal osteoma has been documented in only two cases. Cushing described four cases of orbitoethmoidal osteoma associated with intracranial complications, one of them associated with nasal rhinorrhea and ventricular pneumatocele. The other case was that described earlier by Campbell, et al. In both reports, there were no descriptions or conjectures as to why the ventricular perforation occurred. In our case, the presence of an arachnoid-like membrane covering the cerebral defect and the greasy material over the mucocele were similar to findings previously reported. These structures prevented adhesion between the mucocele and the surrounding brain.

Fig. 3. Upper: Photographs of operative views. Left: A partial incision of the thick grayish cyst (C) wall (arrow) showing layered septations and serous cyst fluid. Right: A nodular calcified mass (Ca) on the frontal base is covered by the cyst wall. Lower: Photograph of operative view and accompanying illustration. The cerebral defect communicates directly with the left lateral ventricle (V) without any intervening membranous structures. Asterisk denotes the choroid plexus. B = brain (or surface of defective cerebrum); R = retractor.
Although the extracranial portion of the osteoma bulged into the ethmoidal sinus, there were no signs of sinus obstruction. Although a mucocele, a mucoid-filled encapsulated mass, usually occurs secondary to obstruction of the paranasal sinuses, there have been several other explanations of the origin of a frontal paranasal osteoma and mucocele. A small ectopic sinus not visualized by our radiological method or by direct observation during surgery may be a cause of mucocele. Alternatively, there could once have been an intracranial connection of the frontal sinus that had disappeared early in our patient’s life.

Pluridirectional tomography and CT scanning are con-
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cidered the most useful diagnostic modalities for demonstrating an intracranially extended, avascular mass associated with the paranasal sinuses. \textsuperscript{5,6,8,11} Differential diagnoses include various types of bone tumors, epidermoid tumor, calcified meningioma, rare gliomas developing extraaxially, parasite infections (cysticercosis), and posttraumatic porencephaly.

Despite the usually chronic nature of disease involving a mucocele, in our present case, perifocal edema on MR imaging and electroencephalographic abnormalities associated with the convulsive episode seemed to be caused by a mucocele. A frontal osteoplastic craniotomy is recommended for exposure of the lesion and plastic repair of the dural perforation.\textsuperscript{10} Care should be taken not to induce chemical meningitis, which could be caused by the greasy material overlying the surface of the mucocele as well as by the cyst contents themselves.

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References


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Address reprint requests to: Yoshikazu Nakajima, M.D., Ph.D., Department of Neurosurgery, Osaka University Graduate School of Medicine, 2-2 Yamadaoka, Suita, Osaka 565-0871, Japan. email: ynakaj@nsurg.med.osaka-u.ac.jp.