Frontal Sinus Osteoma with Osteoblastoma-like Histology and Associated Intracranial Pneumatocele

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Abstract Osteomas of the cranial sinuses are rare, benign bony tumors that can be complicated by the formation of an intracranial pneumatocele. If not treated promptly, a pneumatocele can lead to abscess formation, meningitis, or ventriculitis. In the present case, an intracerebral pneumatocele was formed when an 18 cm³ osteoma breached the posterior wall of the frontal sinus creating a one-way valve through which air could enter the intracranial cavity. The patient presented after forceful sneezing with non-specific symptoms of headache, nausea, and vomiting. CT demonstrated a frontal collection of loculated air with mass effect within the left cerebral hemisphere. A partly mineralized mass occupied the left superior nasal ethmoid sinus and left frontal sinus. Of interest pathologically in this case, the tumor had a substantial osteoblastoma-like component. Surgical repair involved frontal craniotomy to remove the osteoma and debride frontal sinus mucosa, plugging the frontal nasal ducts and sinus with fat and bone wax, and dural restoration using an underwater closed drainage system to vent intracranial air and stabilize the patient.

Keywords Frontal sinus osteoma · Osteoblastoma-like areas · Pneumatocele · Headache

Introduction

Osteomas are relatively rare, slow-growing, osteogenic tumors [1]. Osteomas are the most commonly encountered neoplasm of the frontal sinus and the frontal bone accounts for 80% of all paranasal sinus osteomas [2]. A survey of 1,500 coronal sinonasal CT studies gives an estimate of the incidence of asymptomatic sinus osteomas: 46 patients had paranasal sinus osteomas [3]. So, the majority of paranasal osteomas are discovered serendipitously. On rare occasions, large osteomas of the frontal or ethmoid region have displaced the ocular globe forward resulting in proptosis [4]. The formation of an intracranial tension pneumatocele secondary to a frontal sinus osteoma has been infrequently publicized [2, 5] since the seminal paper of Harvey Cushing [6].

As in the present case, patients with intracerebral pneumatocele often present with nonspecific symptoms: headache, nausea and vomiting, and even cerebral spinal fluid rhinorrhea [5]. The treatment of choice for pneumocephalus complicating frontal sinus osteoma is surgical [7]. If not treated promptly, a pneumatocele can lead to abscess formation acquiring infection from the sinodural fistula, meningitis, or ventriculitis and eventually death [8].

Case Report

A 30-year-old previously healthy young male presented to the emergency department with a sudden onset of severe bifrontal headache. The ictus was precipitated by a forceful and violent sneeze while driving his automobile. Intense pain was accompanied by nausea and vomiting such that he pulled off the road and his wife drove him to the nearest hospital. He had a longstanding history of nasal and post-

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nasal mucoid sinus drainage which was attributed to his occupation and place of residence. His work included tending to large herds of dairy cattle in the dusty Central Valley of California. There was no history of cerebral spinal fluid (CSF) leak or meningitis. He had no previous sinus surgery but did suffer from an uncharacterized frontal skull injury as a toddler.

Physical examination revealed no focal neurological deficits or mental status alteration. The pupils were 3 mm, symmetrical, and briskly reactive. There was no photophobia but fundoscopic examination did reveal bilateral blurring of the optic disc margins. There was no meningeismus or CSF leak with forward head flexion (negative reservoir sign). In the hospital, he clutched his forehead and leaned forward, indicating a severe bifrontal headache. He was afebrile and the vital signs were normal.

CT scan of the head revealed a 5.2 cm (anteroposterior) $\times$ 4.9 cm (craniocaudal) $\times$ 4.1 cm (transverse) collection of loculated air in the left frontal lobe intraparenchymally (Fig. 1b, c) causing a significant mass effect with a 0.8 cm midline shift to the right. There was effacement of the cortical sulci throughout the brain suggesting diffuse cerebral edema. The ventricles were small,

but within normal limits for the patient's age. The CT bone window settings showed a 3.5 $\times$ 1.4 $\times$ 3.5 cm lobulated heterogeneous soft tissue mass of bony density emanating from the nasoethmoid sinus superiorly, essentially filling the left frontal sinus cavity. Areas of central increased density interpreted as inspissated mucus extended into the anterior left ethmoid air cells filling the left frontal sinus and abutting the cortex of the posterior wall of the sinus. No definite cortical defect was evident in the ethmoid sinus; however, a defect was demonstrated in the cortex of the posterior wall of the frontal sinus, to which the intracranial air cavity tapered to a point consistent with origin at this site (Fig. 1b, arrow). The other paranasal sinuses and mastoid air cells were clear. Pneumocephaly did not extend to either the basal cisterns or subarachnoid space. No mass lesions with increased attenuation were seen, excluding acute intracerebral hemorrhage. Neurosurgical consultation was obtained and the patient admitted to the ICU for stabilization prior to urgent craniofacial surgery.

The entire left half of the sinus was filled with an ossific mass. The tumor was dissected off the posterior wall of the frontal sinus where a 0.4 cm defect in the bone was identified. This small portal penetrated the dura exposing

Fig. 1 **a** Preoperative lateral plain film radiograph indicates an anterior location of the air accumulation that abuts the posterior surface of the frontal bone. **b, c** Preoperative CTs display a cyclone-shaped pneumatocele displacing substantial amounts of frontal lobe.

A short blunt projection of the air accumulation (*arrow*) indicates the point of air entry. **d** Postoperative sagittal CT indicates that by postoperative day three, the pneumatocele is less than half its former size

Fig. 2 Transverse CT, frontal sinus **a** a preoperative axial CT scan presents a vague granular/trabecular density pattern correlating with the fibro-osseous and osteoblastoma-like areas that predominated in the pathologic specimen (Fig. 3). The comma-shaped fleck of density in the left anterior portion of the bean-shaped frontal mass

corresponds to the limited amount of dense compact, concentrically laminated osteoma in it. **b** Postoperative CT study; the osteoma is gone. The defect in the posterior table of the frontal bone has been surgically sealed

cerebral cortex. Its base of attachment was on the posterior aspect of the nasofrontal duct system. In summary, the pneumatocele was evacuated, the underlying skull base was debrided down to normal bone, a bone graft from the intersinus septum was used for calvarial reconstruction, and an onlay collagen dural graft (DuraGen®) was used to cover the dural defect. The galea was reapproximated and skin closed in a standard fashion. The patient was returned to the ICU with stable vital signs and no neurological deficit.

A post-operative CT scan confirmed total tumor resection (Figs. 1d, 2b). The tension pneumocephaly showed gradual resolution with serial scans over 10 days.

One irregular $3.1 \times 1.3 \times 0.5$ cm fragment and a $4.2 \times 2.9 \times 1.5$ cm aggregate of multiple pieces of bony tissue were submitted for histological analysis.

Histopathology

After 48-h formalin fixation and decalcification, tumor sections displayed conventional osteoma with concentric laminations of dense compact bone best appreciated with polarized light (Fig. 3a). However, this constituted a minority of the specimen, which otherwise presented a gradation of solidity. The vast majority (90%) of the lesion showed the fibro-osseous histology depicted in Fig. 3b. The remaining 10% was worrisome due to its histologic resemblance to osteoblastoma (Fig. 3c). Gradations between woven and lamellar bone suggested the replacement of the one (immature osteoblastoma-like bone) by the other (lamellar bone) over time.

Discussion

It is decried that there is no clear pathological or clinical classification for fibro-osseous lesions involving the

paranasal sinuses that embraces the variety of lesions that exhibit such diverse pathological and clinical behavior [9]. Fibrous dysplasia, ossifying fibroma, and osteoma are three distinct entities that lie along a continuum containing the least to the most bony content [10]. The diagnosis of fibrous dysplasia and ossifying fibroma is made by correlation between clinical, radiologic, and pathologic findings. Craniofacial bone is the second most common site of involvement by fibrous dysplasia (the skull base is the first) and should be differentiated from osteoma and ossifying fibroma, but it is rarely found in the paranasal sinus. Radiographically, fibrous dysplasia shows “ground-glass” bone appearance on CT scans with bone window [11]. Histopathologically, it presents woven-type bone embedded in a cellular fibrous stroma without osteoblastic rimming. The present biopsy had uncommon woven bone networks of “C” and “O” shape pattern resembling fibrous dysplasia (Fig. 3b). A pseudo-Paget mosaicism (the result of chronic remodeling activity) was brought out under polarized light; lamellar rather than woven bone excludes fibrous dysplasia and absent is the rim of plump osteoblasts seen in ossifying fibroma.

Ossifying fibroma is a benign, usually slow-growing tumor. Most of these affect the mandible. Their behavior in the mid-face and paranasal sinuses is more aggressive than their mandibular counterparts [12]. However, in ossifying fibroma, fibrous tissue predominates with a minor component of mature (lamellar) bone rimmed by active osteoblasts and psammoma-type mineralizations scattered throughout [13], quite unlike the solid laminated bone in the present case (Fig. 3a).

An osteoblastoma is a benign bone tumor that most often occurs in the vertebral column and the long bones of the extremities. Paranasal sinus location of osteoblastoma is very rare [14]. In such reports, the histopathologic description actually sounds more like osteoma with osteoblastoma-like areas: proliferating osteoblasts along with

Fig. 3 **a** Dense, compact bone of a classic frontal sinus osteoma has concentric laminations brought out by polarized light ($\times 200$, H&E), **b** the majority of the mass was fibro-osseous, resembling fibrous dysplasia but lacking the woven bone structure and prominent peripheral osteoblasts ($\times 100$; polarized light, H&E), **c** 10% of the mass had histopathology identical to osteoblastoma; plump atypical osteoblasts redundantly encircle their irregular bone product ($\times 400$, H&E)

small trabeculae of woven bone and rich vascular fibrous stroma [11]. Sinus osteomas may have osteoblastoma-like areas and distinguishing them from true osteoblastoma can

be challenging. Some believe osteomas with osteoblastoma-like features behave more aggressively. Seventeen of 45 surgically excised craniofacial sino-orbital osteomas (38%) [15] had osteoblastoma-like areas. These manifested more frequent orbital involvement (41% vs. 13%), and so, visual changes were more frequent. Osteomas with osteoblastoma-like features were more often incompletely excised (25% vs. 14%). However, clinical recurrence was less common (8% vs. 27%). It was concluded that osteoblastoma-like features are common in sino-orbital osteomas and represent active remodeling within an osteoma rather than defining a distinct clinicopathologic entity. Osteoblastoma-like features did not correlate with more adverse clinical features or worse outcome. Distinguishing this from true osteoblastoma requires careful histologic evaluation and radiographic correlation.

Osteosarcomas can resemble osteoblastoma [16]. However, even low-grade malignant lesions do not mature into the osteoma pattern as seen in the present specimen (Fig. 3c).

Extension of osteoma through the posterior table of the frontal sinus can result in life-threatening complications [17]. A “succession splash” is considered diagnostic of pneumocephalus, but is not always present. Radiologic imaging is the most reliable means of diagnosing frontal sinus osteoma associated with intracranial pneumatocele. The intracranial air usually depresses cerebral hemispheres extradurally. Rappaport and Attia have stated that “posteriorly located frontal sinus osteomas [should] be considered for early excision because of the thin-walled bone in that location and the potential for serious intracranial complication should that layer be breached” [2].

In a review of 295 cases of pneumocephalus, 75% occurred after craniofacial surgery in trauma, while 13% were caused by tumors, the majority of which were frontal or ethmoidal sinus osteomas [18]. Two theories explain the formation of intracranial pneumatoceles. Both presume a breach in the dura or deeper layers to allow ingress of air. The first theory is appropriately termed the “inverted soda pop bottle phenomenon”, whereby leakage of cerebrospinal fluid through a dural or subarachnoid defect exerts a siphon effect to draw air up into the relatively low-pressure intracranial space. The second theory relies on the breached dura to act as a simple flap valve whenever extracranial pressure exceeds intracranial pressure, as in the act of blowing the nose [19]. Thus, intuitively, air flows intracranially and becomes trapped, setting up a pressurized cavity that exerts a mass effect and causes neurologic symptoms.

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