

CLINICAL CASE - TEST YOURSELF

Neuro/Head and Neck Radiology

Acute decrease of visual acuity and exophthalmos in a male patient

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PART A

A 73-year-old male patient presented to the emergency department for acute worsening of visual acuity and exophthalmos of the left eye. The patient reported that a piece of wood had fallen close to his left eye about a week prior to

admission. Ophthalmologic physical examination showed proptosis and papilloedema in the left eye. Visual acuity was 3/10 in the left eye and 7/10 in the right eye. A computed tomography (CT) scan was performed (**Figs. 1-3**).



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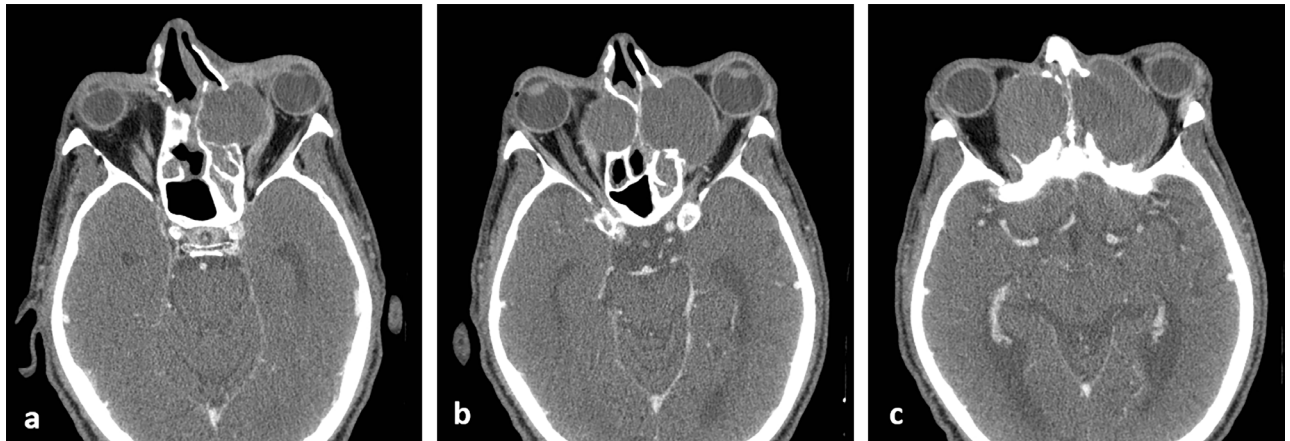


Fig. 1. Axial contrast-enhanced CT images of the sinuses and the orbit.

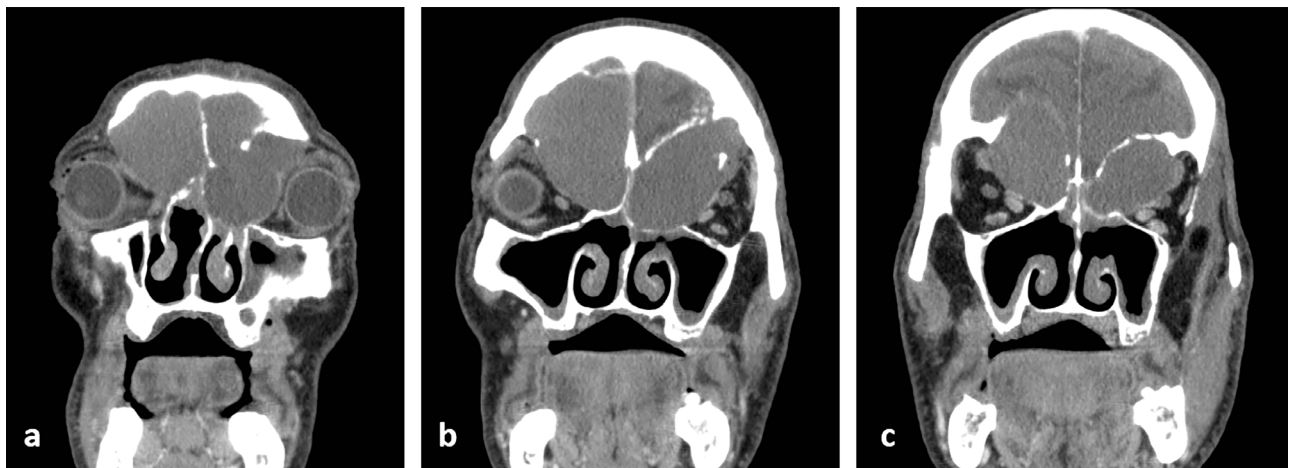


Fig. 2. Coronal CT image reconstruction of the sinuses and the orbit.

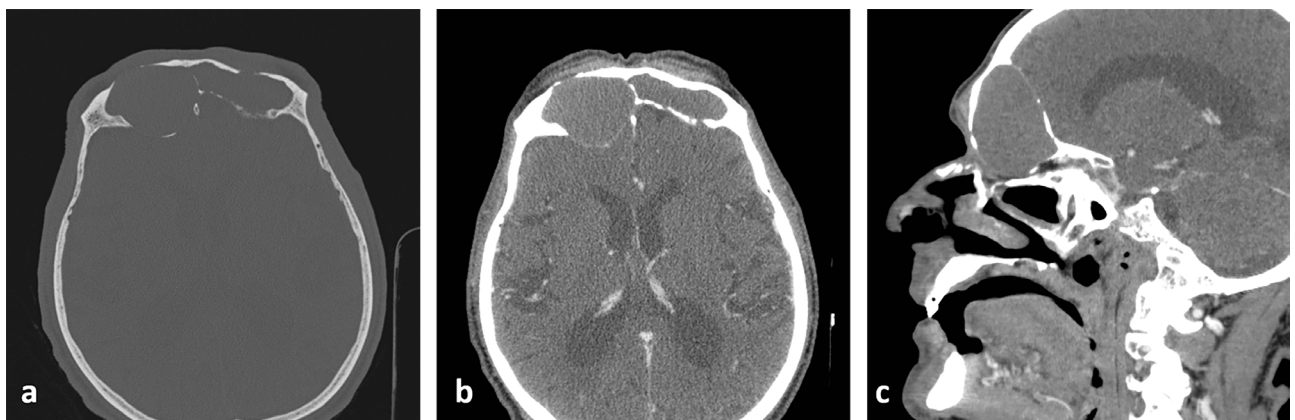


Fig. 3. a, b. Axial CT images of the frontal sinuses (bone window a, soft tissue window b) and c. sagittal CT image reconstruction of the right frontal sinus.

PART B

Diagnosis: Bilateral fronto-ethmoido-orbital mucocoele.

Paranasal sinus mucocoeles are benign, slow-growing expansile masses that develop after obstruction of the sinus ostium [1]. The accumulation of secretions within a mucoperiosteal lined cavity completely fills the associated paranasal sinus, unlike retention cysts which only partially occupy the cavity [2]. There is only a slight male gender predilection according to various studies and the highest incidence occurs in the third and fourth decades of life [1, 3]. Mucocoeles expand in a path of less resistance, inducing erosion and remodelling of the surrounding bone. New bone formation or thickening can also occur [2-4]. The most commonly affected sinus is the frontal, followed by the ethmoid (70-90% of cases), probably because of their complex and variable drainage. Maxillary (10%) and sphenoid sinuses (1%) are not usually involved [1, 3]. Mucocoeles can be primary or secondary. The most common causes of primary lesions are chronic inflammation (infection, allergy, mucociliary dysfunction), mucus drainage blockage, secretory duct obstruction, mucus gland cystic dilatation and cystic degeneration of polyps. Secondary mucocoeles are caused by trauma or surgery [1, 2, 5]. There is a strong association between the presence of nasal polyps and the mucocoele formation, probably due to the role of inflammation [6]. If a mucocoele is infected, it becomes a mucopyocoele [4].

The manifestation of paranasal sinus mucocoele is determined by the location and the vital structures involved and can be classified as rhinologic, ophthalmologic and neurologic symptoms and signs, with the ophthalmologic symptoms being the most common [1]. When there is intraorbital extension, the mass effect upon the orbit leads to exophthalmos, diplopia, paresis of extraocular muscles, epiphora and periorbital swelling, with or without pain. On the other hand, when there is direct optic nerve compression, the clinical manifestations include visual compromise or even unilateral blindness, impaired papillary reflex and optic atrophy [1, 3, 5, 7]. This is

the result of erosion of the paper-thin lamina papyracea, which forms the posteromedial wall of the orbit [5, 8]. Infection and elevation of intraocular pressure constitute additional possible mechanisms causing optic neuropathy [5]. The third, fourth, fifth and sixth cranial nerves and the pituitary gland may also be involved [3]. In the case of larger lesions, the primary site cannot be determined. The expanding mucocoele causes extensive osteolysis of the bone walls and destruction of anatomical structures and can obstruct the adjacent sinuses' drainage ostia [2]. Serious complications of mucocoeles include epidural abscess, meningitis, subdural empyema, brain abscess and cranial nerve palsies [4].

The diagnosis of sinus mucocoele is confirmed by CT or magnetic resonance imaging (MRI). CT can better delineate bone details and is preferable for head and neck evaluation. MRI has proven to be superior in terms of tissue contrast [4, 8]. On CT, a mucocoele appears homogeneous and isodense to the cerebral parenchyma, with sharp margins [2, 3, 9]. The adjacent osseous structures are remodelled with areas of thickening and erosion. There may be osteolysis of the osseous wall, without signs of infiltration of adjacent anatomical structures. However, in areas of lower resistance, such as the anterior cranial fossa, there may be herniation [2, 4]. After contrast media administration, there is a thin rim of enhancement, which corresponds to the mucoperiosteal lining of the cavity. If the rim is thick or the mass is hyperdense in the pre-contrast images, a mucopyocoele must be suspected [1, 2, 9]. A distinguishing feature between mucocoeles and neoplasms is the absence of internal enhancement, exclusively seen in mucocoeles [2, 3, 9]. MRI is used to identify the sinonasal tumours which may be the aetiological factor in some cases of large mucocoeles [2]. Mucocoeles show variable signal intensity on both T1- and T2-weighted images, depending on the fluid and protein content, viscosity and degree of dehydration [1, 4]. Water-rich content results in low signal intensity on T1W and high signal intensity on T2W MR images. When the protein content increases, there is hyper-

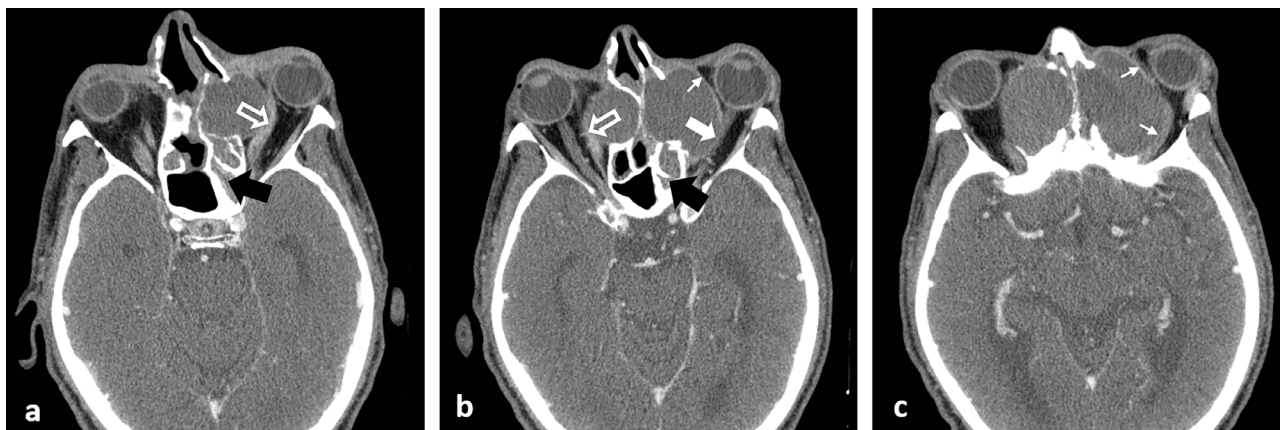


Fig. 1. Axial CT images (a-c) of the sinuses and the orbit showing expansile, soft tissue lesions, with rim enhancement (thin white arrows in b, c), into the ethmoid sinuses (black arrows in a, b) and the orbit bilaterally. The mass compresses the left optic nerve (thick white arrow in b) and the medial rectus muscles (open white arrows in a, b) and causes outward displacement of the globe.

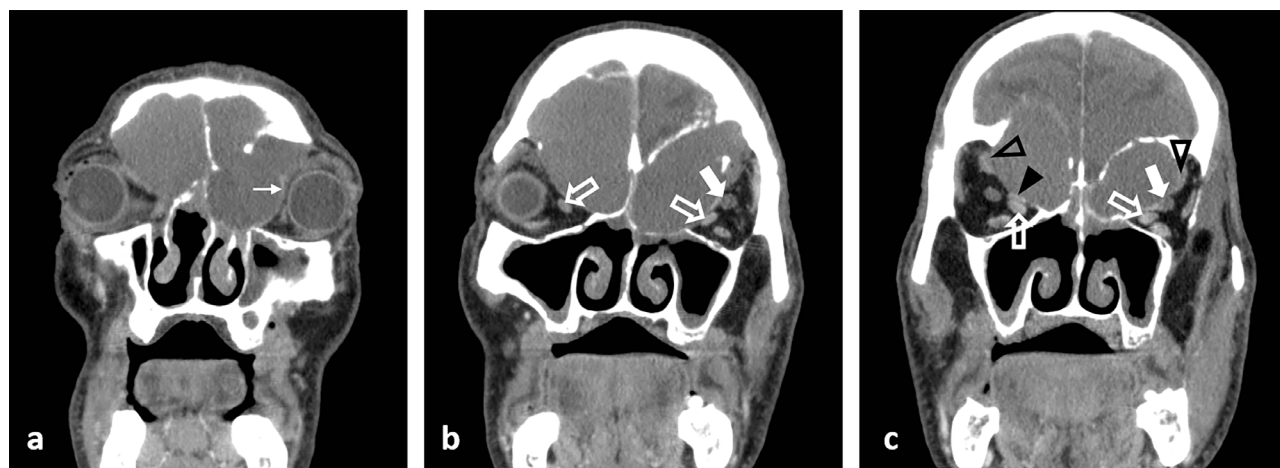


Fig. 2. Coronal CT reconstructions of the sinuses and the orbit showing an expansile, homogeneous mass with rim enhancement (thin arrow in a), filling the frontal sinuses and eroding the roof and the superomedial wall of the orbit. The mass compresses the left optic nerve (thick white arrows in b, c), the superior rectus (open black arrowhead in c), medial rectus (open white arrows in b, c) and the superior oblique (black arrowhead in c) muscles and causes downward and outward displacement of the globe.

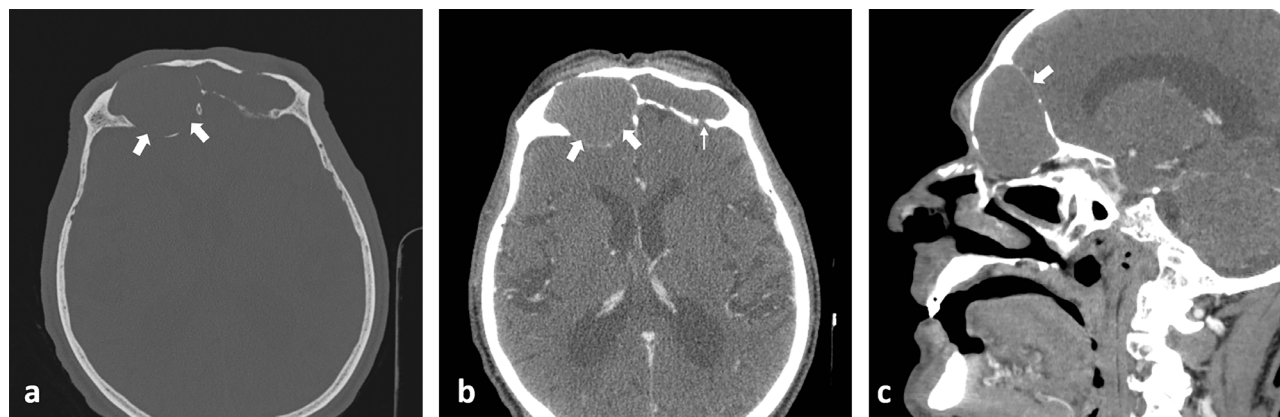


Fig. 3. a, b. Axial CT images of the frontal sinuses and c. sagittal CT reconstruction of the right frontal sinus showing thinning and erosion of the posterior wall of the right frontal sinus and the thin enhancing periosteum (thick arrows in a-c), separating the mucocoele from the anterior cranial fossa. Areas of thickening and erosion of the posterior wall of the left frontal sinus are also noted (thin arrow in b).

intensity in both sequences [2, 3]. When the high protein content is even higher and coexists with a high degree of dehydration (such as in *Aspergillus* or other invasive fungal infections), the signal intensity appears low on T1W and T2W MR sequences, and hence the sinus can be mistaken to be aerated [10]. CT in these cases is useful by demonstrating an opacified hyperdense sinus.

The differential diagnosis of mucocoeles, depending on their location, includes paranasal sinus neoplasms, inverted papilloma, Rathke's cleft cyst, dermoid cyst, hypophyseal adenoma, cranio-pharyngioma, optic glioma, schwannoma, chondroma-chondromyxoma and lymphoma [1, 2, 9].

Surgical excision is the treatment of choice. In the past, mucocoeles were removed with external approaches or an osteoplastic flap, however recently, most patients are treated with endoscopic techniques, allowing lower morbidity and better cosmetic result [1, 3, 6]. The treatment is based on marsupialisation and enlarging the normal drainage pathways. Detection of recurrence requires a long follow-up with CT or MRI [1, 3].

In our case, CT revealed an expansile, homogeneous mass with rim enhancement, filling the frontal sinuses and expanding into the ethmoid sinuses and the orbit bilaterally (**Fig. 1**). The mass eroded the roof and superomedial wall of the orbit and caused gross downward and outward displacement of the globe. It also compressed the medial rectus, superior rectus and superior oblique muscles, as well as the left optic nerve (**Fig. 2**). The posterior wall of the right frontal sinus was partially eroded. Consequently, the thin enhancing periosteum was the only remaining structure to separate the mucocoele from the intracranial cavity (**Fig. 3**). The typical imaging features established the diagnosis of an immense mucocoele.

The mucocoele was evacuated by functional endoscopic sinus surgery under systemic antibiotic prophylaxis. The post-operative ophthalmologic examination revealed progression of the visual acuity to 5/10 in the left eye and stability in the right eye. **R**

Conflict of interest

The authors declared no conflicts of interest.



KEY WORDS

Mucocoele; Frontal sinus; Ethmoid sinus; Computed Tomography; Exophthalmos; Optic nerve

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