



The many faces of fungal disease of the paranasal sinuses: CT and MRI findings

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ABSTRACT

The prevalence of fungal rhinosinusitis has increased worldwide over the last two decades. Fungal rhinosinusitis includes a wide variety of infections, from relatively innocent to rapidly fatal processes. Fungal infection may be one of the most challenging forms of sinonasal pathology to manage, especially the invasive forms, which have high mortality rates. Therefore, it is essential to correctly diagnose and classify fungal disease of paranasal sinuses in order to accurately predict prognosis and implement effective therapy. This essay describes the different manifestations of fungal sinusitis on computed tomography and magnetic resonance imaging to optimize differentiation, and includes correlation with the pathologic classifications.

Fungal disease of the paranasal sinuses (FDPNS) is a relatively uncommon entity. The disease is broadly categorized into either invasive or noninvasive forms, and subdivided into five distinct entities. Invasive fungal sinusitis can be categorized as acute or chronic invasive and chronic granulomatous forms, and noninvasive fungal sinusitis includes allergic fungal sinusitis and fungus ball.

Preoperative suspicion of FDPNS is often helpful in prompting the surgeon to obtain appropriate tissue during surgery and alerting the pathologist prior to histopathologic processing for proper identification of fungi. Cross-sectional imaging features along with clinical presentation are critical for early diagnosis, prognosis, and appropriate treatment. In this essay, we discuss the categories of FDPNS and their imaging features.

Clinical and imaging findings

Acute invasive fungal sinusitis

Acute invasive FDPNS is defined as a fulminant fungal infection less than four weeks in duration, usually in an immunocompromised setting (1). The infection results from a rapid spread of fungi from the paranasal sinuses to the adjacent orbits and central nervous system, followed by vessel wall invasion, resulting in vascular thrombosis and resultant arterial and/or venous ischemia (2–6). Spread from the paranasal sinuses can occur through direct invasion or hematogenous spread. Acute invasive fungal sinusitis is common in immunocompromised and diabetic patients, although it has also been rarely reported in immunocompetent individuals (1, 2). A high index of suspicion and early diagnosis are critical, especially in individuals who are immunocompromised.

Noncontrast computed tomography (CT) scan of the paranasal sinuses reveals hypoattenuating mucosal thickening or intrasinus and/or intranasal soft tissue attenuation, associated with osseous erosion. Early changes cannot be differentiated from a nonspecific sinusitis (Fig. 1). The nasal cavity (middle turbinate) is often the primary site of infection (Figs. 2 and 3). There is a predilection for unilateral involvement of the ethmoid and sphenoid sinuses.

Aggressive osseous destruction is followed by intracranial, cavernous sinus (Fig. 4), and intraorbital extension. The fungi can extend via perivascular channels beyond the paranasal sinuses with intact bony walls. Therefore, bone erosion and mucosal thickening may sometimes be very subtle and appear insignificant.

Obliteration of the normal fat density within the periantral regions suggests soft tissue extension of FDPNS and may be the earliest sign of extrasinus involvement. CT is the tool of choice for the detection of os-

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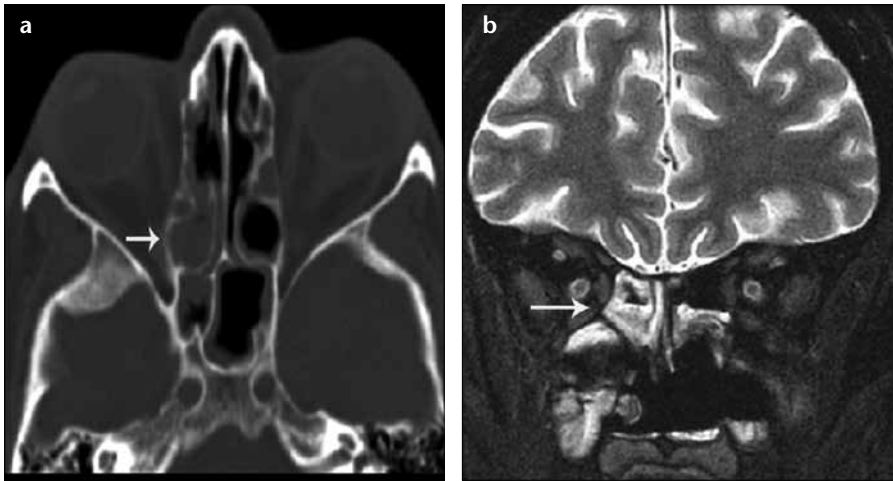


Figure 1. a, b. A 32-year-old male presenting with facial pain in early stages of acute invasive fungal rhinosinusitis. Axial CT image (a) shows nonspecific soft tissue attenuation filling a right ethmoid air cell (arrow). The coronal T2-weighted MR image (b) shows mixed signal intensity filling a right ethmoid air cell (arrow) with areas of signal void, representing fungal infection. The clinical picture progressed, and the patient was found to have acute invasive fungal sinusitis.

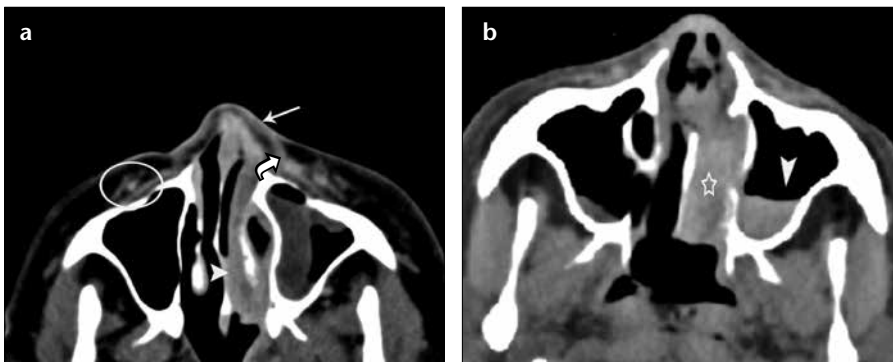


Figure 2. a, b. Acute invasive fungal rhinosinusitis in a 62-year-old female with acute myelogenous leukemia blast crisis presenting with facial and orbital pain. Axial contrast-enhanced CT image (a) shows left nasal and anterior septal soft tissue thickening and enhancement (arrow). There is mild soft tissue thickening involving the left middle turbinate (arrowhead), which is a common location for fungal infection. Note also mild enlargement of the left facial vein (curved arrow) compared to the normal contralateral side (circle). Axial contrast-enhanced CT image (b) shows complete opacification of the left nasal cavity (star), which is a common presentation for acute invasive fungal disease of the paranasal sinuses. Both left nasal cavity and left maxillary sinus contents (arrowhead) have attenuation values higher than simple fluid. There was associated nasal septal perforation.

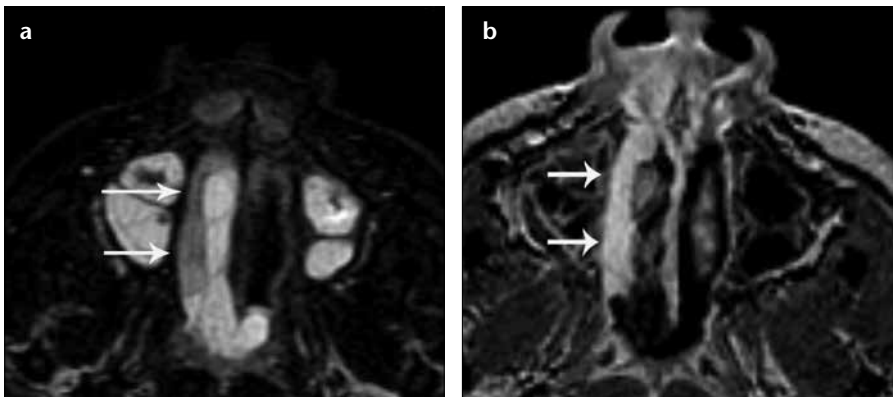


Figure 3. a, b. Fungal rhinitis in a 37-year-old male with nasal discharge. Axial fat saturated T2-weighted MR image (a) shows thickened mucosa along the lateral wall of right nasal cavity with hypointense signal (arrows). Axial postcontrast T1-weighted MR image (b) shows associated prominent mucosal enhancement (arrows).



Figure 4. Acute invasive fungal disease. A 36-year-old male presenting with multiple right cranial nerve palsies. Coronal T2-weighted MR image shows a loss of normal hyperintense signal intensity within Meckel's cave on the right (arrowhead), representing fungal invasion. The normal contralateral side (arrow) is shown for comparison.

seous erosion. Complications such as vascular occlusions with infarcts and cerebritis (Figs. 5 and 6), leptomeningeal involvement, intracranial granulomas (characteristically hypointense on T1- and T2-weighted images) (Fig. 7), and pseudoaneurysms are better depicted on magnetic resonance imaging (MRI) (3, 5, 7).

Chronic invasive FDPNS

The chronic invasive form is a slowly progressive fungal infection with a time course longer than 12 weeks (1, 4). Affected patients are usually immunocompetent, but those receiving corticosteroid therapy or with diabetes mellitus or AIDS are also susceptible. Patients present with symptoms of chronic sinusitis, but focal neurological deficits, facial soft tissue swelling, or ocular symptoms may be the initial presentation. Prominent or slightly hyperdense soft tissue in the paranasal sinus with associated sinus wall erosion is commonly seen on CT. This may have the appearance of an aggressive sinus mass (Figs. 8 and 9). Paranasal sinus walls may show mottled lucencies or irregular bone destruction (5). There is decreased signal intensity on T1-weighted images and markedly decreased signal intensity on T2-weighted images (Figs. 8 and 9) (3). Infiltration of the periantral soft tissues about

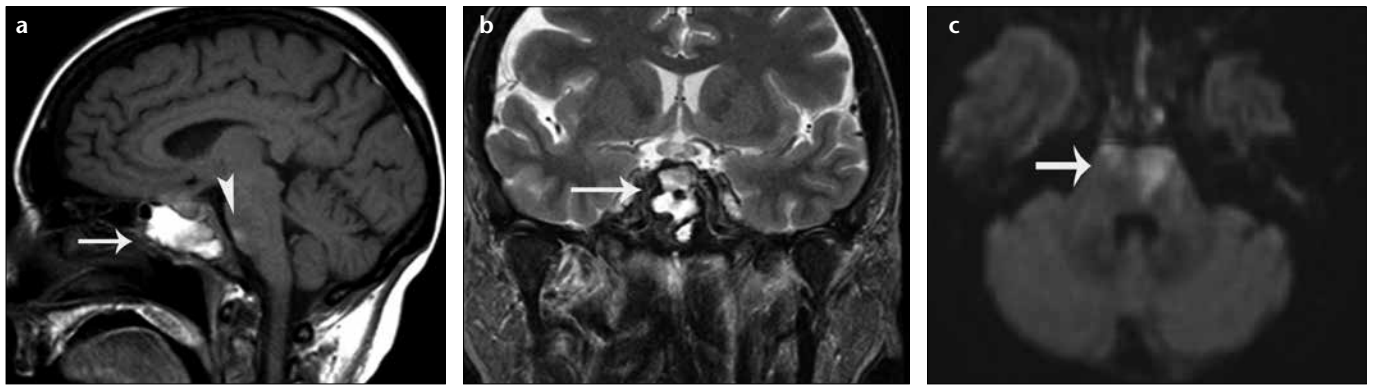


Figure 5. a–c. Acute invasive fungal disease in a 60-year-old female with history of acute lymphoblastic leukemia presenting with fever and altered mental status. Sagittal T1-weighted (a) and coronal T2-weighted (b) images show hyperintense sphenoid sinus contents (arrows). Sagittal T1-weighted image (a) also shows some intrinsic hyperintensity in the anterior pons (arrowhead), which is consistent with hemorrhage. Axial diffusion weighted image (c) shows acute brainstem infarct (arrow) due to angioinvasive acute fungal disease, with corresponding hypointense signal on ADC map confirming hemorrhagic infarction (not shown).

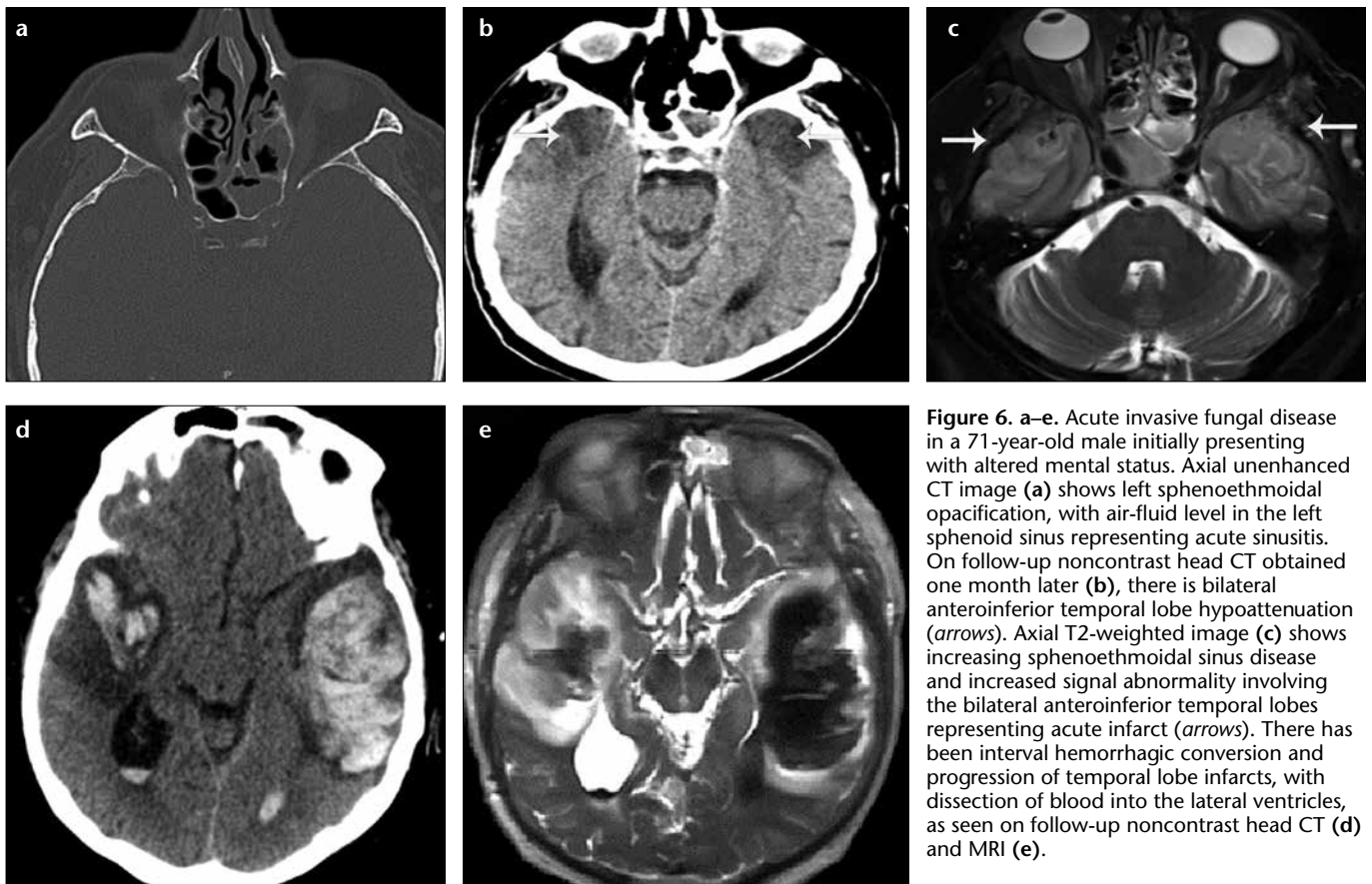


Figure 6. a–e. Acute invasive fungal disease in a 71-year-old male initially presenting with altered mental status. Axial unenhanced CT image (a) shows left sphenothmoidal opacification, with air-fluid level in the left sphenoid sinus representing acute sinusitis. On follow-up noncontrast head CT obtained one month later (b), there is bilateral anteroinferior temporal lobe hypoattenuation (arrows). Axial T2-weighted image (c) shows increasing sphenothmoidal sinus disease and increased signal abnormality involving the bilateral anteroinferior temporal lobes representing acute infarct (arrows). There has been interval hemorrhagic conversion and progression of temporal lobe infarcts, with dissection of blood into the lateral ventricles, as seen on follow-up noncontrast head CT (d) and MRI (e).

the maxillary sinus in the setting of reactive osteitis may suggest chronic invasive FDPNS.

Chronic invasive FDPNS has a tendency to invade adjacent structures, such as the orbit, cavernous sinus, and anterior cranial fossa. This may result in epidural abscess, parenchymal cerebritis or abscess, meningitis, cavernous sinus thrombosis, osteomyeli-

tis, mycotic aneurysm, or stroke. Differentiation between chronic invasive FDPNS and sinonasal malignancies may not be possible based on imaging findings (3, 8).

Chronic invasive granulomatous FDPNS

This condition is a slowly progressive form of fungal infection characterized by a chronic granulomatous

process with a time course of longer than 12 weeks, which is similar clinically and radiographically to chronic invasive FDPNS (1–4). The disease has been reported almost exclusively in immunocompetent patients from North Africa and Southeast Asia with a history of chronic sinusitis, with only scattered reports from the USA. Patients present with symptoms of

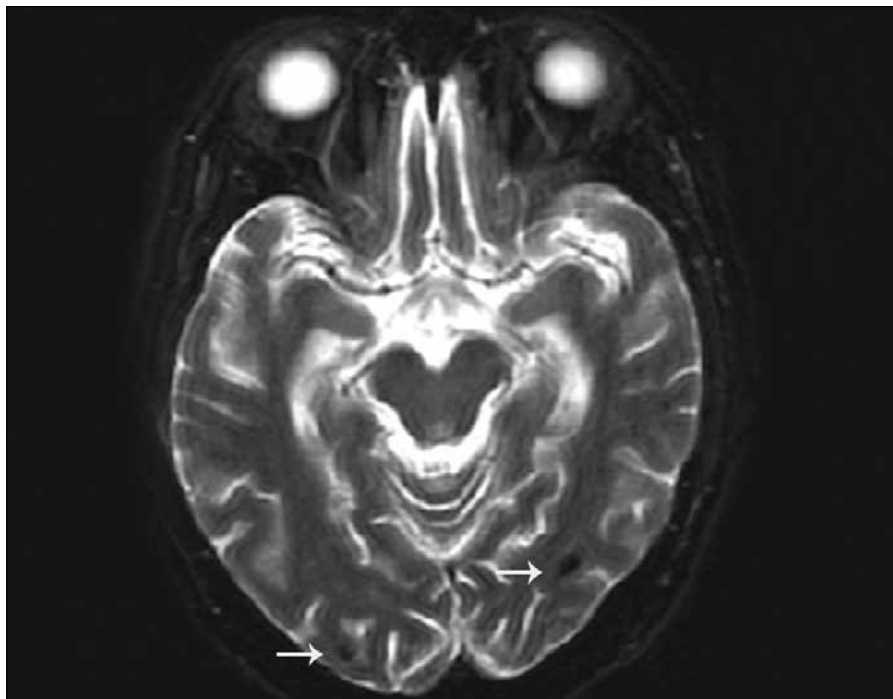


Figure 7. Intracranial aspergillosis due to sinonasal acute invasive disease. Axial T2-weighted image shows typical multifocal nodular hypointense aspergillomas involving the gray-white matter junction (arrows). The patient had acute myelogenous leukemia, presented with altered mental status and died after five days. The route of spread was hematogenous dissemination from the paranasal sinuses.

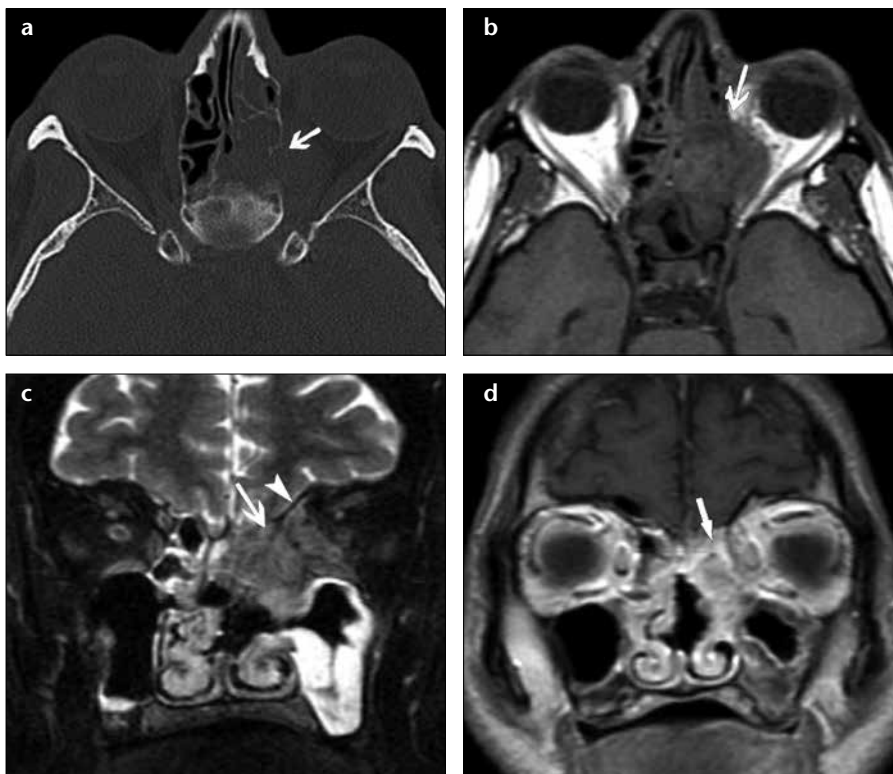


Figure 8. a–d. Chronic invasive fungal sinusitis in a 53-year-old male with orbital pain and double vision on the left. Axial CT image (a) shows left ethmoid soft tissue mass destroying the left medial orbital wall (arrow) and extending into the left orbit. There is an isointense soft tissue mass (arrow) extending from the left ethmoid air cells into the left orbit on axial unenhanced T1-weighted image (b). Coronal T2-weighted image (c) shows relatively T2 hypointense left ethmoid soft tissue mass (arrow) extending into the left orbit (arrowhead). On coronal postcontrast T1-weighted image (d), the mass enhances avidly, making differentiation from sinonasal malignancy difficult (arrow). The nasal septum is absent, secondary to previous sinus surgery.

chronic sinusitis associated with proptosis. Histologically, there are non-caseating granulomas associated with vasculitis, vascular proliferation, and perivascular fibrosis (1, 5). The imaging findings have not been described well, but Stringer and Ryan (8) reported three cases with soft tissue masses eroding the orbital walls, with involvement of the nasal cavity and pterygopalatine fossa.

Allergic FDPNS

This disease is the most common form of FDPNS, and it is believed that fungal allergens evoke immune-mediated mucosal inflammation in an atopic host. Sinonasal inflammation in combination with viscous allergic mucin effectively obstructs the normal drainage pathway (1). The disorder usually occurs in young immunocompetent patients who present with chronic sinusitis and a history of previous sinus surgery (3). Pansinusitis and nasal involvement are common. The imaging findings typically include involvement of multiple paranasal sinuses bilaterally with intrasinus hyperattenuating material on noncontrast CT (Figs. 10 and 11). Expansion of the involved sinuses and remodeling and thinning of the bony sinus walls are common findings, with bone erosion and unilateral involvement rarely seen (3). On MRI, very low T2 signal intensity is seen due to high concentrations of various metals concentrated by the fungal organisms, as well as high protein and low water-free content of the allergic mucin. Although T1 intensity is variable and low signal intensity can also be observed, high signal intensity or mixed low, intermediate, and high signal intensity is frequently observed in allergic fungal sinusitis (3).

Fungus ball

Fungus ball, previously called mycetoma, is described as the presence of a noninvasive, dense conglomeration of fungal hyphae usually in a single sinus cavity (1). The disease tends to occur in older immunocompetent individuals. Patients are usually asymptomatic, although they may present with mild pressure sensation involving one of the paranasal sinuses, nasal discharge, and cacosmia (3).

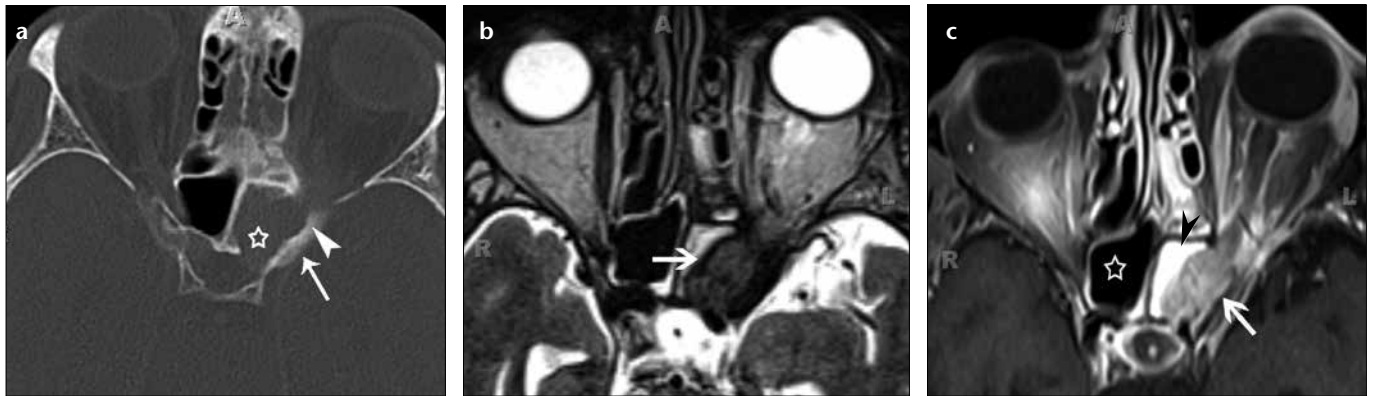


Figure 9. a–c. Chronic invasive fungal disease of the paranasal sinuses in a 62-year-old man who developed pain in the left eye 10 months ago and subsequently noted a blurry spot in the center of his left eye. On axial unenhanced CT image in bone algorithm (a), there is opacification of the left sphenoid sinus (*star*) with surrounding sphenoid septum and lateral wall reactive osteitis (*arrow*). There is erosion of the posterior wall of the sphenoid on the left. Note also left optic canal expansion (*arrowhead*). Axial T2-weighted image (b) shows hypointense soft tissue filling a majority of the left sphenoid sinus (*arrow*), eroding through the sinus wall and involving the left prechiasmatic optic nerve. There is T2 hypointense soft tissue extending through the left optic canal. Axial contrast-enhanced T1-weighted image (c) shows abnormal enhancement and thickening of the left prechiasmatic and intracanalicular optic nerve (*arrow*). The remaining sphenoid sinus is filled with obstructed sinus secretions on the left (*arrowhead*) and air on the right (*star*).

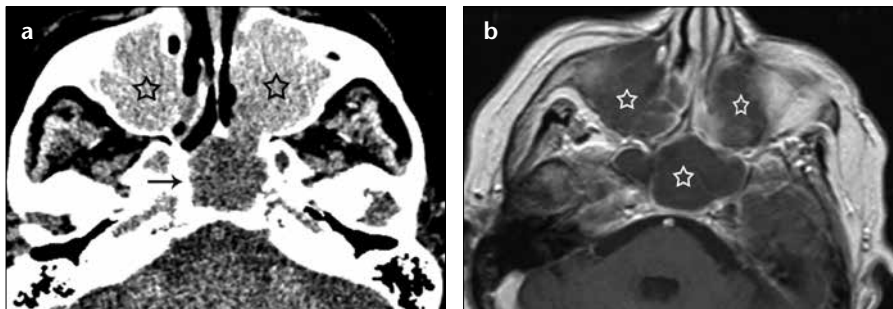


Figure 10. a, b. Allergic fungal sinusitis in an 83-year-old female with nasal obstruction. Axial unenhanced CT image (a) shows bilateral expanded and completely opacified maxillary and sphenoid sinuses with hyperattenuating (*stars*) and isoattenuating (*arrow*) sinus contents. On axial postcontrast T1-weighted image (b), there is no enhancement of mildly expanded paranasal sinus contents (*stars*).

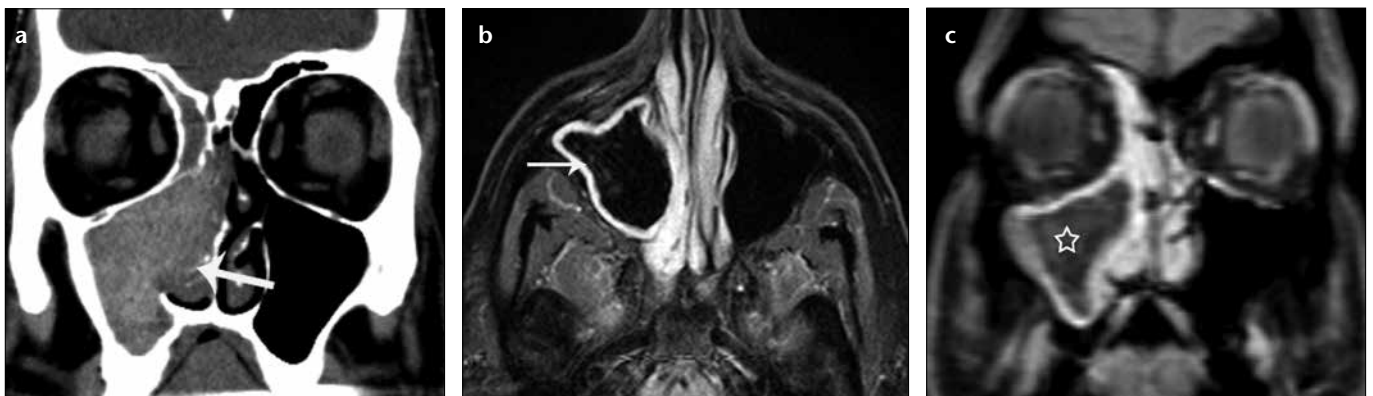


Figure 11. a–c. Allergic fungal sinusitis in a 62-year-old male presenting with sinus pressure. Coronal unenhanced CT image (a) shows complete opacification of the right maxillary sinus with areas of hyperattenuation (*arrow*). Axial fat-suppressed T2-weighted MR image (b) shows pseudo-pneumatized sinus, with circumferential rim to T2 hyperintense thickened mucosa and “dirty” central T2 hypointensity (*arrow*) due to fungal content. Coronal fat suppressed postcontrast T1-weighted image (c) shows no intrinsic enhancement of right maxillary sinus contents (*star*). There is circumferential mucosal enhancement that is mildly thickened.

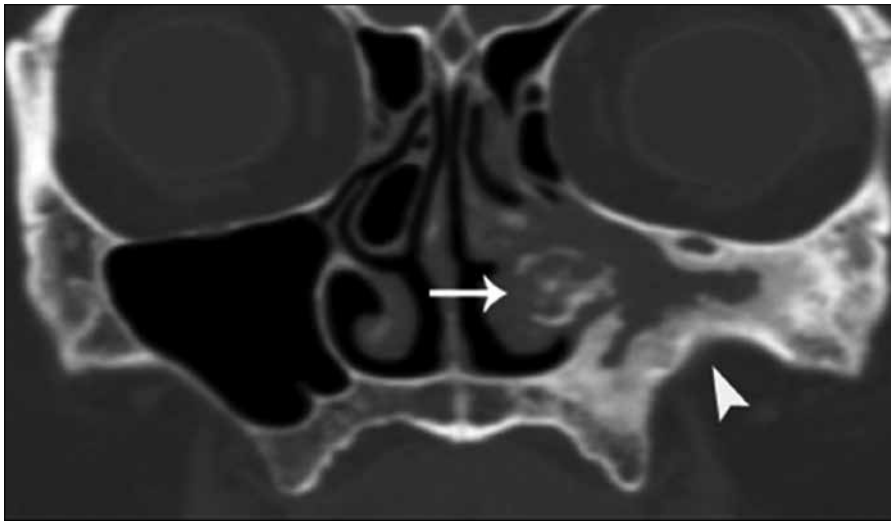


Figure 12. Fungus ball in a 56-year-old man presenting with left nasal obstruction and chronic sinus pain. On noncontrast coronal CT image of the paranasal sinuses, there is complete opacification of the atelectatic left maxillary sinus, with associated thickened walls, which is consistent with chronic sinusitis (*arrowhead*). There is a coarse nodular calcification replacing the left inferior turbinate, representing a fungus ball (*arrow*).



Figure 13. Fungus ball in a 55-year-old male with facial pain. Axial noncontrast CT image shows a coarse calcification in the right middle turbinate (*arrow*), representing a fungus ball.

Noncontrast CT reveals a hyperattenuating mass within the involved sinus, with occasional intralesional calcifications (Figs. 12 and 13). Surrounding circumferential hypoattenuating mucosal thickening is usually present, and there may be reactive osteitis indicating chronic sinusitis (Fig. 12). The bony margins of the involved sinus are usually intact. MRI shows

intermediate T1 and markedly hypointense T2 signal.

Conclusion

Classification of FDPNS into invasive or noninvasive forms is important to guide treatment and determine prognosis. Sinus expansion, smooth sinus wall thinning, intrasinus hyperattenuation on CT and hypointense signal on

T2-weighted MRI in an atopic individual should raise suspicion of allergic FDPNS.

A hyperattenuating mass within an opacified single sinus in a nonatopic immunocompetent patient is consistent with fungus ball. Obliteration of the normal fat density within the periantral regions, osseous erosion, orbital, cavernous sinus and/or brain involvement in an immunocompromised individual should alert the radiologist to the possibility of invasive FDPNS. It should be kept in mind that the initial stages of infection might not reveal any suggestive imaging signs.

In conclusion, the radiologist should be aware of the various subtypes of FDPNS and their particular imaging features. We propose noncontrast CT imaging when either invasive or noninvasive fungal infection is suspected. If spreading beyond the paranasal sinuses is suspected, MRI should be employed.

Conflict of interest disclosure

The authors declared no conflicts of interest.

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