

Case 16443

Eurorad●●

Pneumosinus dilatans associated with fibrous dysplasia

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Section: Musculoskeletal system

Area of Interest: Musculoskeletal bone Neuroradiology brain

Procedure: Diagnostic procedure

Imaging Technique: CT

Imaging Technique: MR

Special Focus: Dysplasias Case Type: Clinical Cases

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Patient: 83 years, female

Clinical History:

An 83-year-old female patient was admitted to the emergency department of our hospital one hour after onset of right-sided hemiparesis and right-sided hemianopia. There was no relevant medical history.

Imaging Findings:

Computed tomography (CT) of the brain revealed no intracranial haemorrhage nor imaging signs of acute ischaemia (Fig. 1). Incidentally, an enlarged left frontal sinus with unilateral osseous expansion of the adjacent frontal bone and the greater wing of the sphenoid was seen. Moreover, the affected bones were sclerotic with areas of interspersed radiolucency (Fig. 1,2).

Subsequent magnetic resonance imaging (MRI) was performed the next day to confirm the clinical suspicion of ischaemic stroke and to exclude haemorrhagic transformation after tissue Plasminogen Activator (tPA) administration. MRI revealed a subtle focus of acute ischaemia in the left lentiform nucleus, visualised as a hyperintense signal on FLAIR with diffusion restriction on diffusion weighted imaging (DWI). The osseous lesion in the left frontal bone demonstrated intermediate signal intensity on T1-weighted imaging and predominantly high signal on T2-weighted imaging. Gadolinium enhancement was inhomogeneous but without any extra-axial nor intra-axial enhancement of the brain (Fig. 3).

Discussion:

Pneumosinus dilatans (PSD) refers to an air-filled paranasal sinus, abnormally enlarged beyond the normal boundaries of the skull bones, and in the absence of osseous erosion, hyperostosis, or mucous membrane thickening [1]. Although the condition was first described by Meyes in 1898, the term 'pneumosinus dilatans' was coined by Benjamins in 1918 [2].

PSD is a rare condition with an unknown true incidence [3]. It occurs most frequently in the frontal sinuses, followed by the sphenoid, ethmoid, and maxillary sinuses [4].

The imaging characteristics for PSD are straightforward on both CT and MRI, i.e. expansion of a paranasal sinus with normal wall thickness [3]. PSD is often an incidental finding [3,5]. Unilateral frontal bossing may be present as a clinical sign [6].

The key role of imaging is to evaluate the presence of underlying conditions [3]. There is a documented association between PSD and fibrous dysplasia [7], meningiomas [3], arachnoid cysts [8], port-wine stains [9], hydrocephalus [10], but it can also be idiopathic [5]. PSD may cause spontaneous pneumocephalus [11].

The pathophysiology of PSD remains unclear, although many hypotheses have been postulated. The most cited theory comprises a one-way valve, creating a pressure gradient, thus increasing the outward pressure on the sinus wall [3,12,13]. A traction phenomenon due to an adjacent meningioma with subsequent bone remodelling has been suggested as well [3]. Whether these theories can be extrapolated to other causes of PSD such as fibrous dysplasia is still debated.

Fibrous dysplasia (FD) is a non-hereditary, benign bone disease. It is characterised by abnormal osteoblastic differentiation and maturation, leading to focal replacement of normal bone tissue by fibrous stroma and islands of immature bone [14]. FD manifests as an expansile bone lesion with smooth cortical contours. Its most common appearance on CT is that of ground-glass density, but it may be homogeneously sclerotic and even cystic [14,15]. MRI appearance is highly variable due to the variation in cellularity of FD. T1-weighted imaging may yield low to intermediate signal intensities, T2-weighted imaging may demonstrate low to high signal intensities. Similarly, gadolinium enhancement is highly variable [15].

Treatment of PSD is directed at surgically correcting the cosmetic deformity of the skull bone, and endoscopic restoration of sinus drainage [5].

In conclusion, the importance of pneumosinus dilatans is to recognise this type of sinus expansion as a clue to potential underlying pathologies.

Written informed patient consent for publication has been obtained.

Differential Diagnosis List: Pneumosinus frontalis dilatans with associated craniofacial fibrous dysplasia, Pneumocele, Hypersinus, Paget's disease of bone, Skull vault haemangioma

Final Diagnosis: Pneumosinus frontalis dilatans with associated craniofacial fibrous dysplasia

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Figure 1

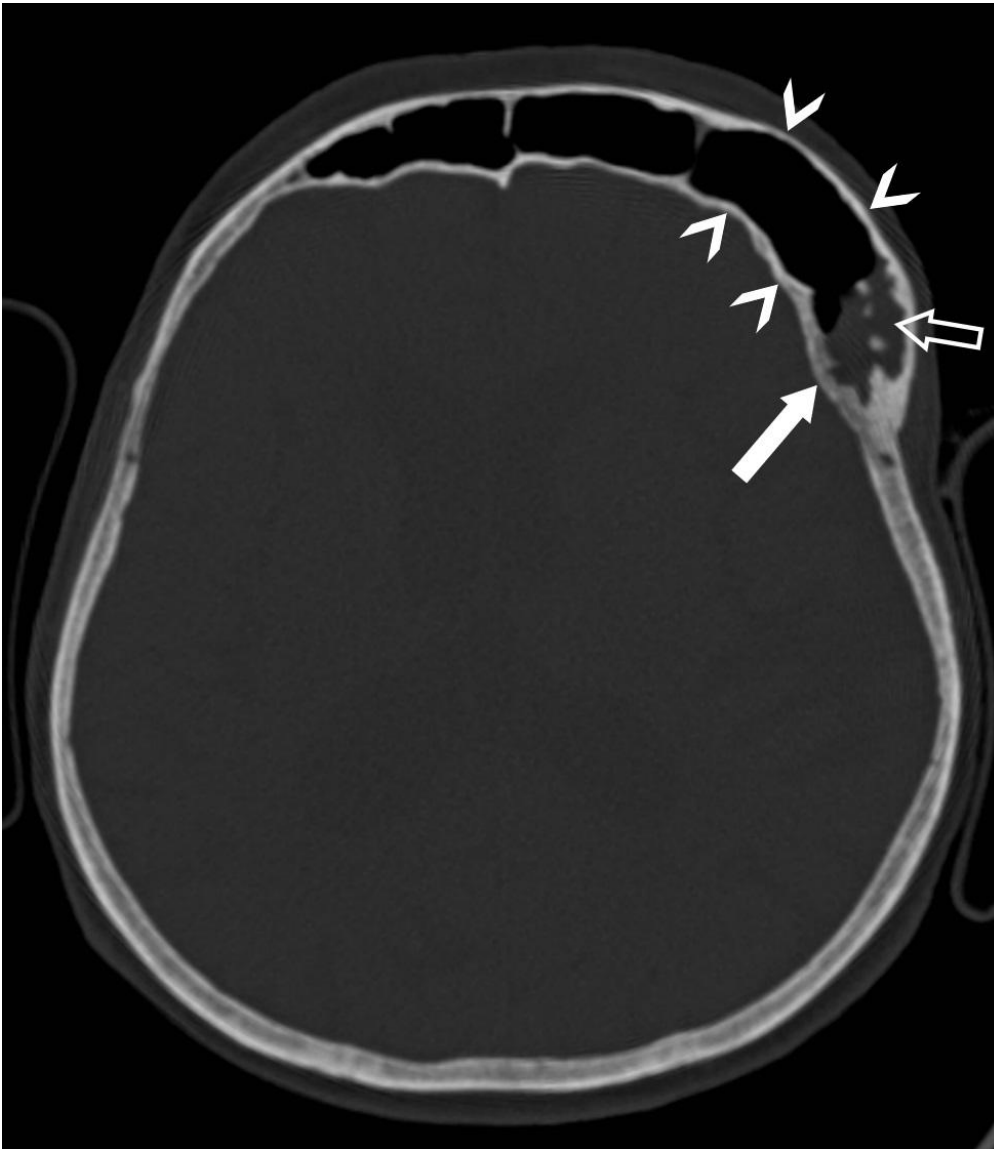
a



Description: Enlarged left frontal sinus (arrowheads) with unilateral osseous expansion of the adjacent frontal bone (arrow). The expanded bone is sclerotic with interspersed radiolucent areas. No evidence of intracranial haemorrhage or acute ischaemia. **Origin:** © Department of Radiology, RZ Heilig Hart Tienen, Belgium 2019.

Figure 2

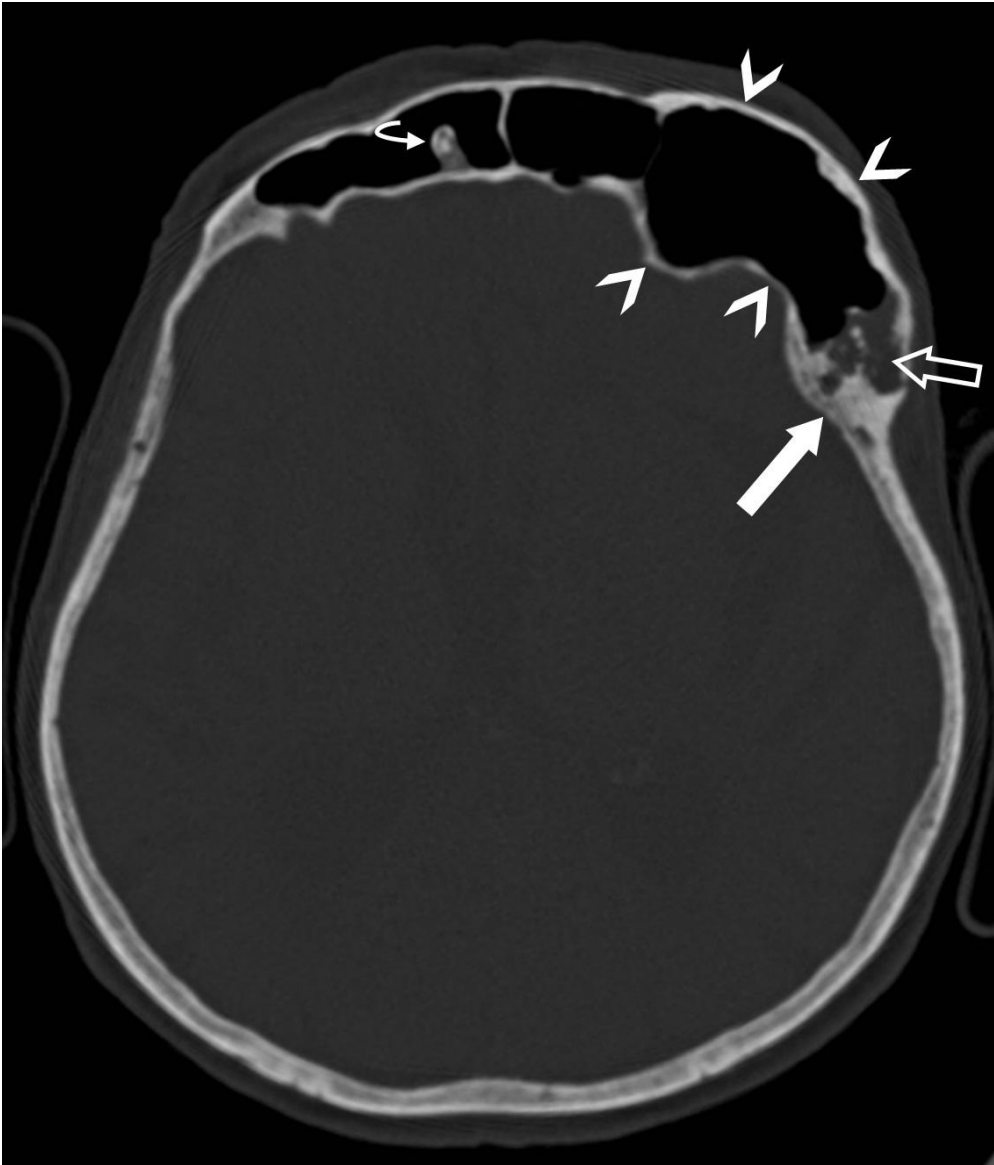
a



Description: Axial view: Enlarged left frontal sinus with normal bony wall thickness (arrowheads). Unilateral osseous expansion of the adjacent frontal bone; sclerotic bone (arrow) with interspersed radiolucent areas (void arrow).

Origin: © Department of Radiology, RZ Heilig Hart Tienen, Belgium 2019.

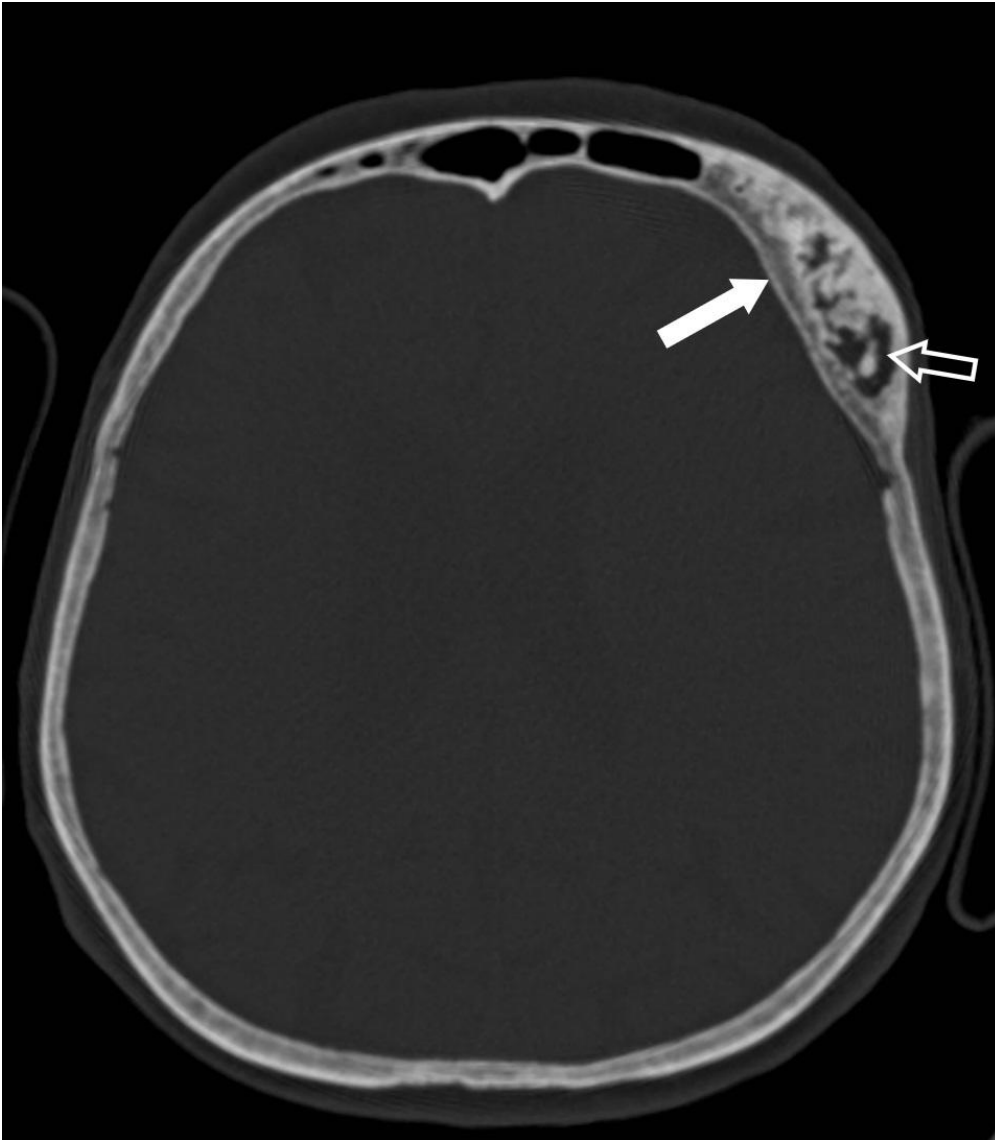
b



Description: Axial view, adjacent slice more caudally: Enlarged left frontal sinus with normal bony wall thickness (arrowheads). Unilateral osseous expansion of the adjacent frontal bone; sclerotic bone (arrow) with interspersed radiolucent areas (void arrow). Small paranasal sinus osteoma in the right frontal sinus (curved arrow).

Origin: © Department of Radiology, RZ Heilig Hart Tienen, Belgium 2019.

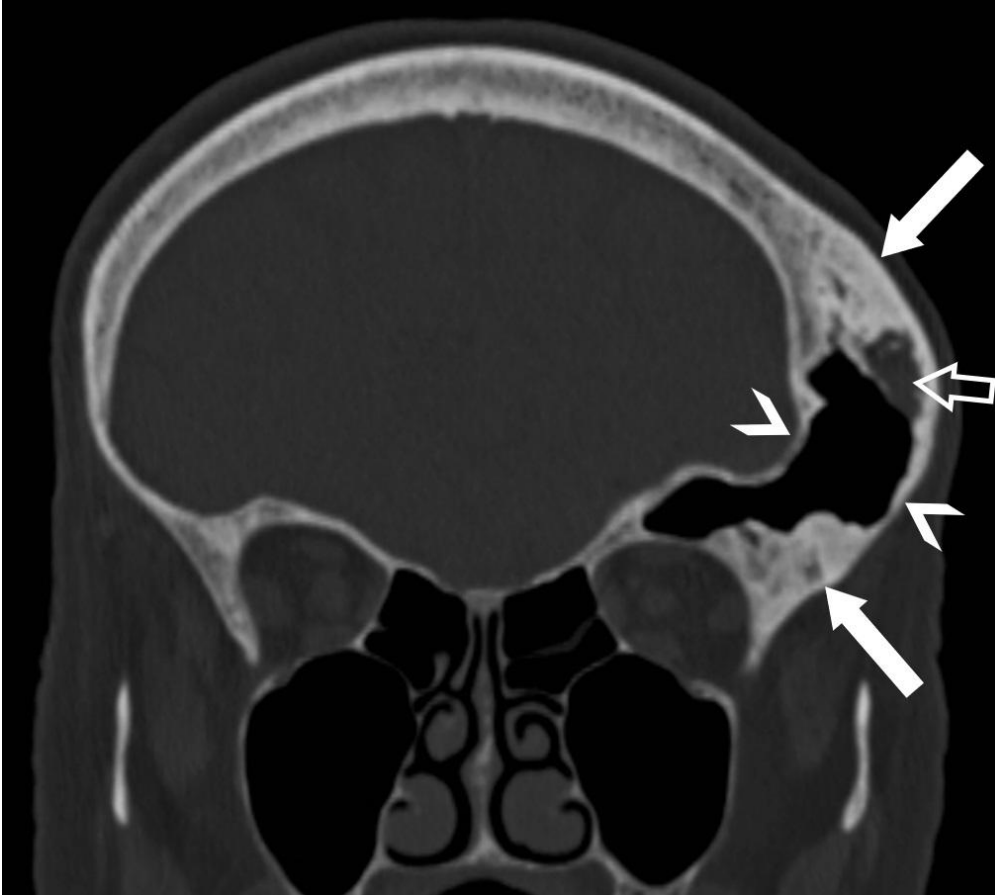
c



Description: Axial view, adjacent slice more cranially: Unilateral osseous expansion and sclerosis of the adjacent frontal bone (arrow) with interspersed radiolucent areas (void arrow).

Origin: © Department of Radiology, RZ Heilig Hart Tienen, Belgium 2019.

d

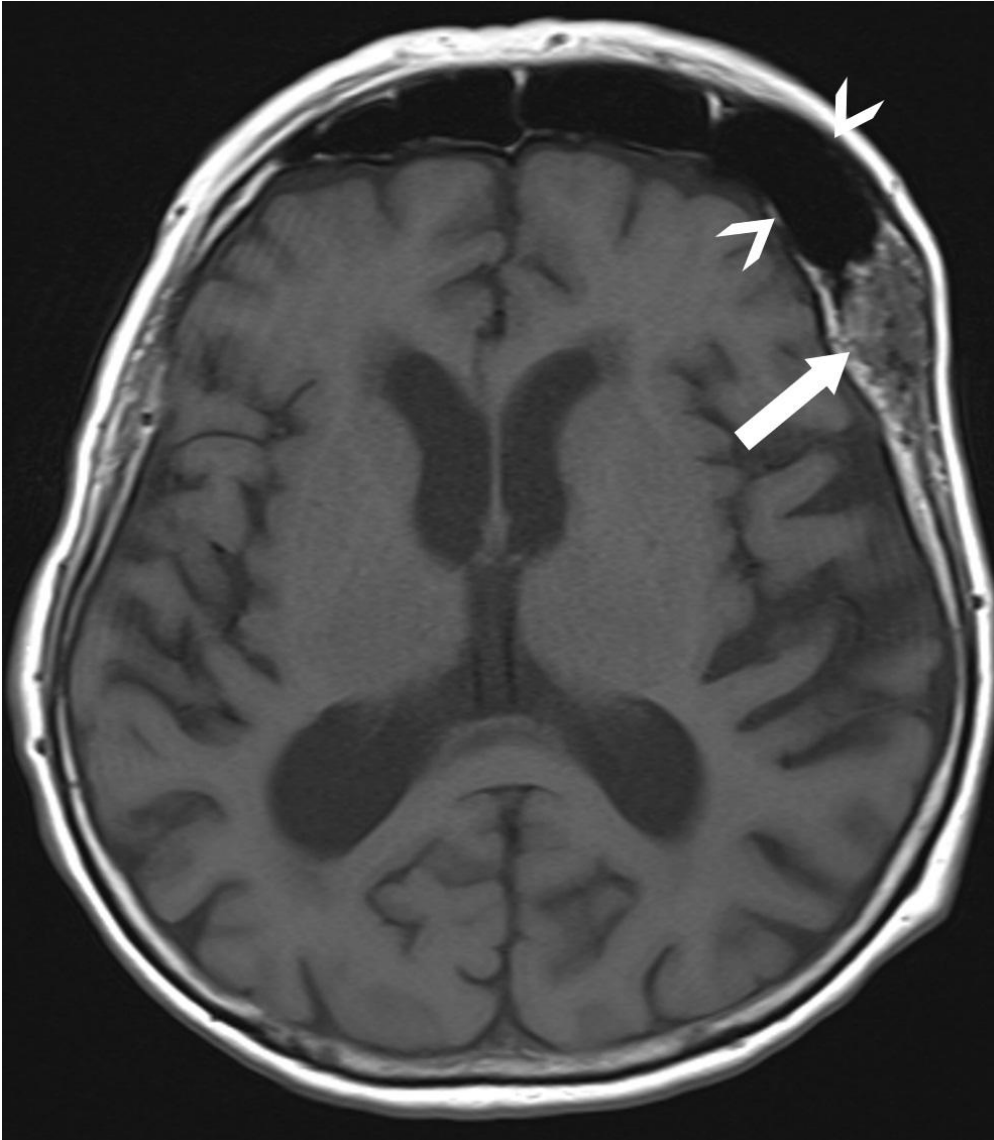


Description: Coronal reconstruction: Enlarged left frontal sinus (arrowheads) with unilateral osseous expansion of the adjacent frontal bone. The expanded bone is predominantly sclerotic (arrows) with interspersed radiolucent areas (void arrow).

Origin: © Department of Radiology, RZ Heilig Hart Tienen, Belgium 2019.

Figure 3

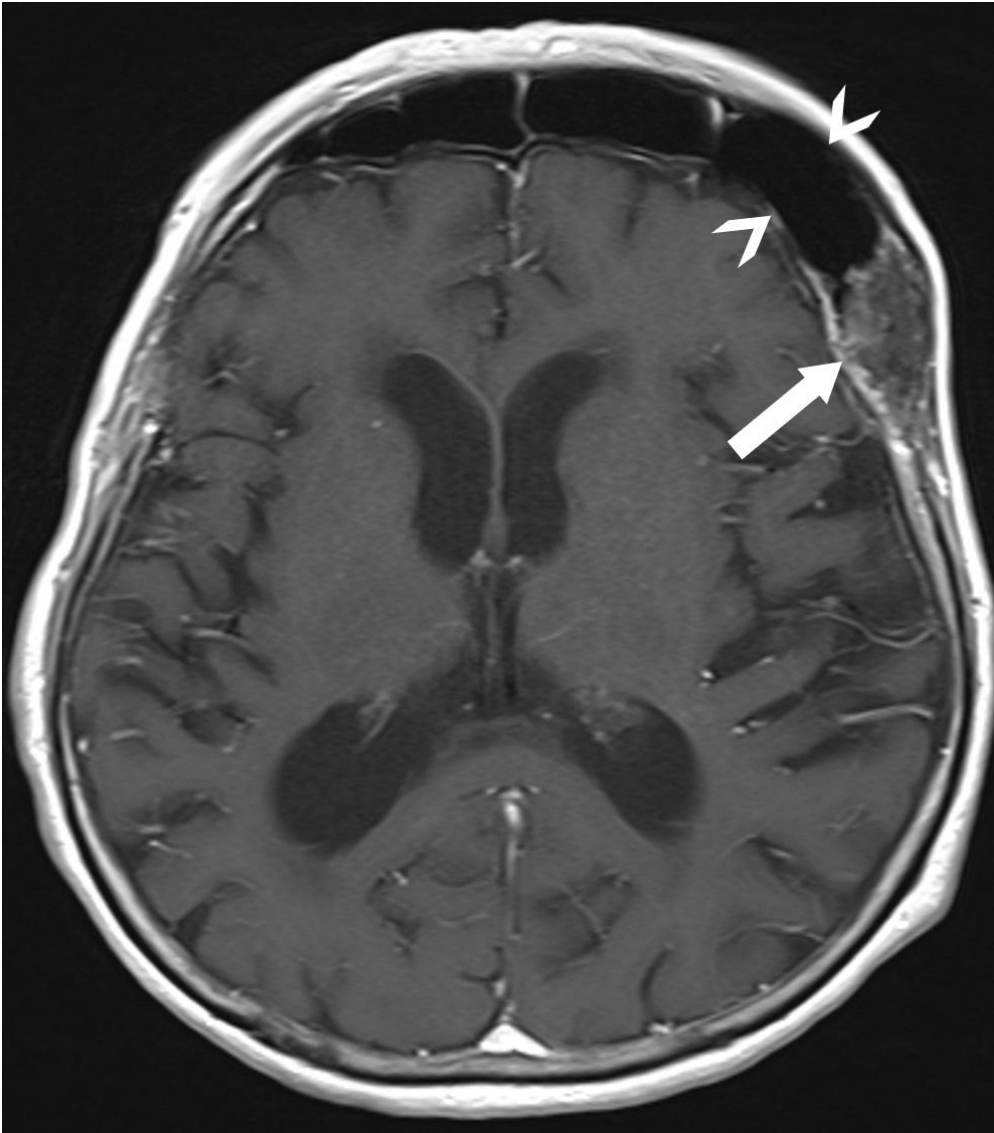
a



Description: Axial T1-weighted image: Enlarged left frontal sinus (arrowheads) with unilateral osseous expansion of the adjacent frontal bone. The osseous lesion is heterogeneous but predominantly iso-intense to muscle tissue (arrow).

Origin: © Department of Radiology, RZ Heilig Hart Tienen, Belgium 2019.

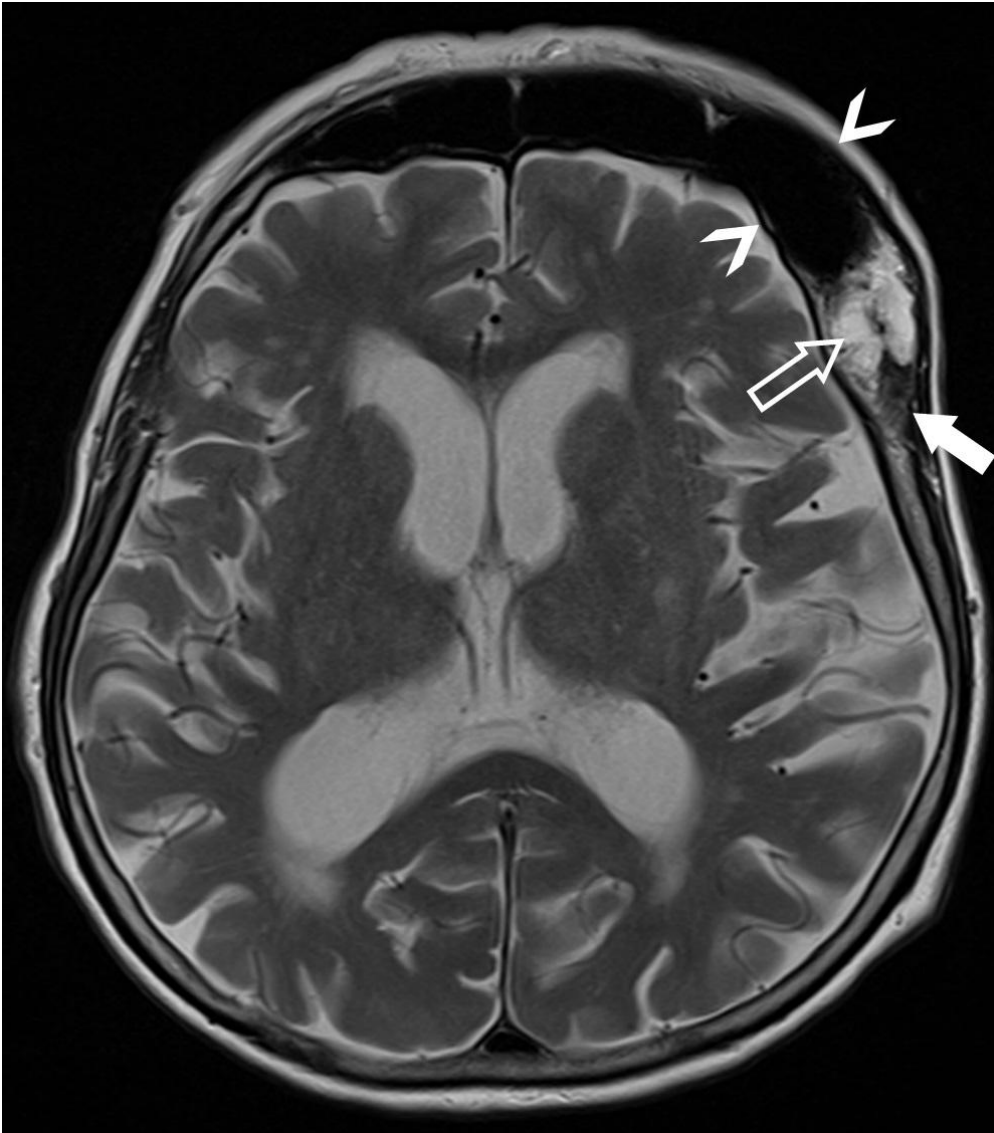
b



Description: Axial T1-weighted image +Gadolinium: Enlarged left frontal sinus (arrowheads) with unilateral osseous expansion of the adjacent frontal bone. The osseous lesion enhances inhomogeneously (arrow). No evidence of extra-axial nor intra-axial enhancement of the brain.

Origin: © Department of Radiology, RZ Heilig Hart Tienen, Belgium 2019.

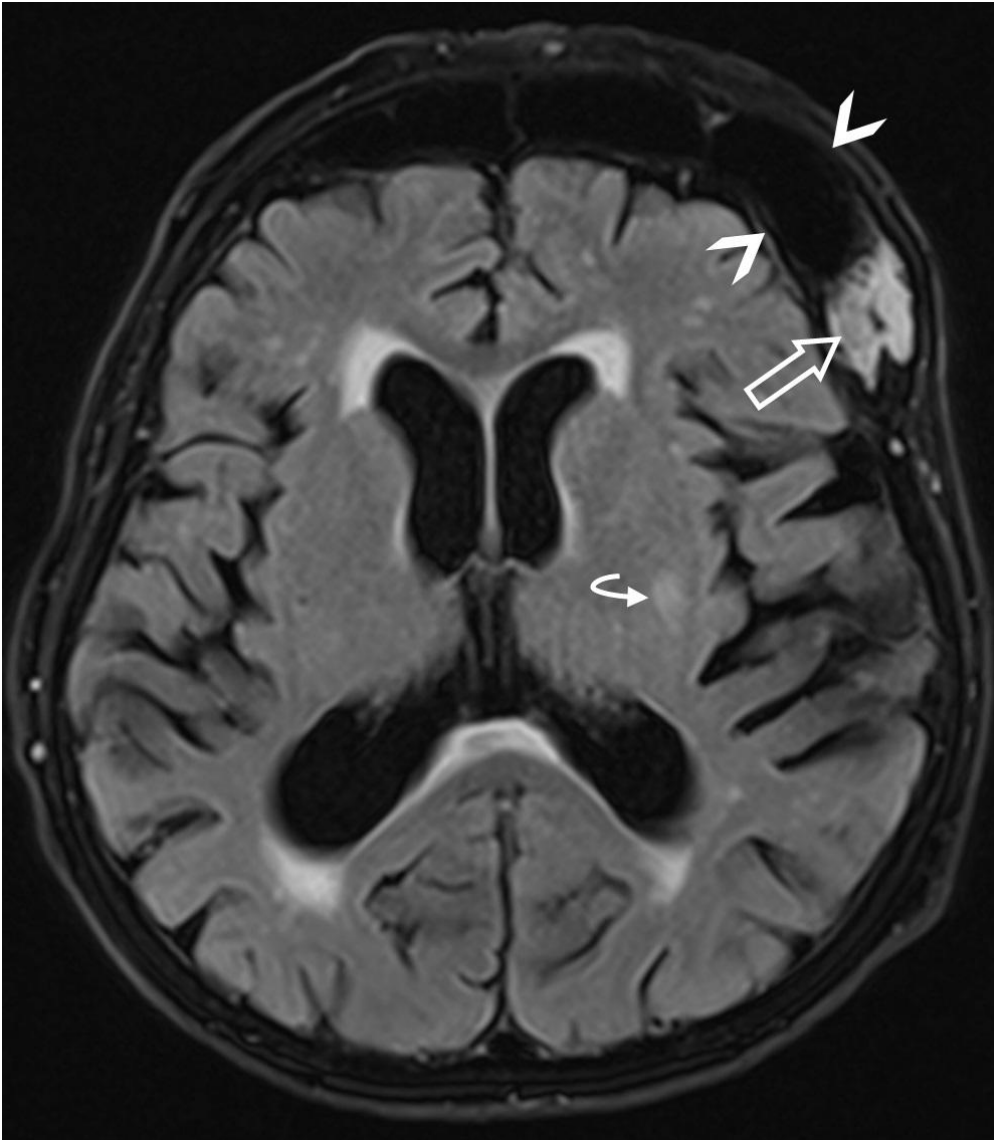
c



Description: Axial T2-weighted image: Enlarged left frontal sinus (arrowheads) with unilateral osseous expansion of the adjacent frontal bone. The osseous lesion is hyperintense, correlating with the areas of interspersed radiolucency seen on CT (void arrows). The low signal intensities seen in the osseous lesion correlate with islands of sclerotic bone (arrow).

Origin: © Department of Radiology, RZ Heilig Hart Tienen, Belgium 2019.

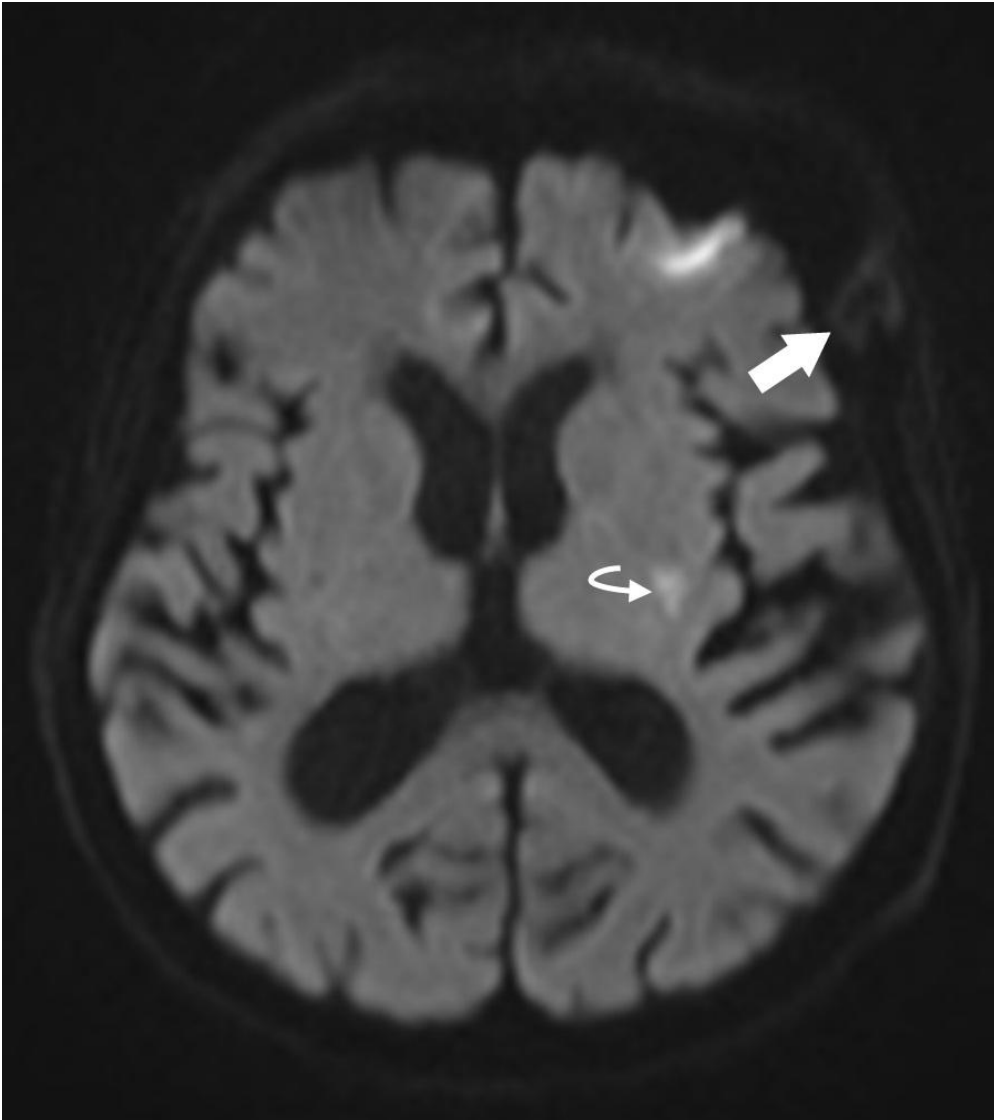
d



Description: Axial FLAIR sequence: Enlarged left frontal sinus (arrowheads) with predominantly hyperintense unilateral osseous expansion of the adjacent frontal bone (void arrow). A subtle focus of hyperintense signal is seen in the left lentiform nucleus (curved arrow).

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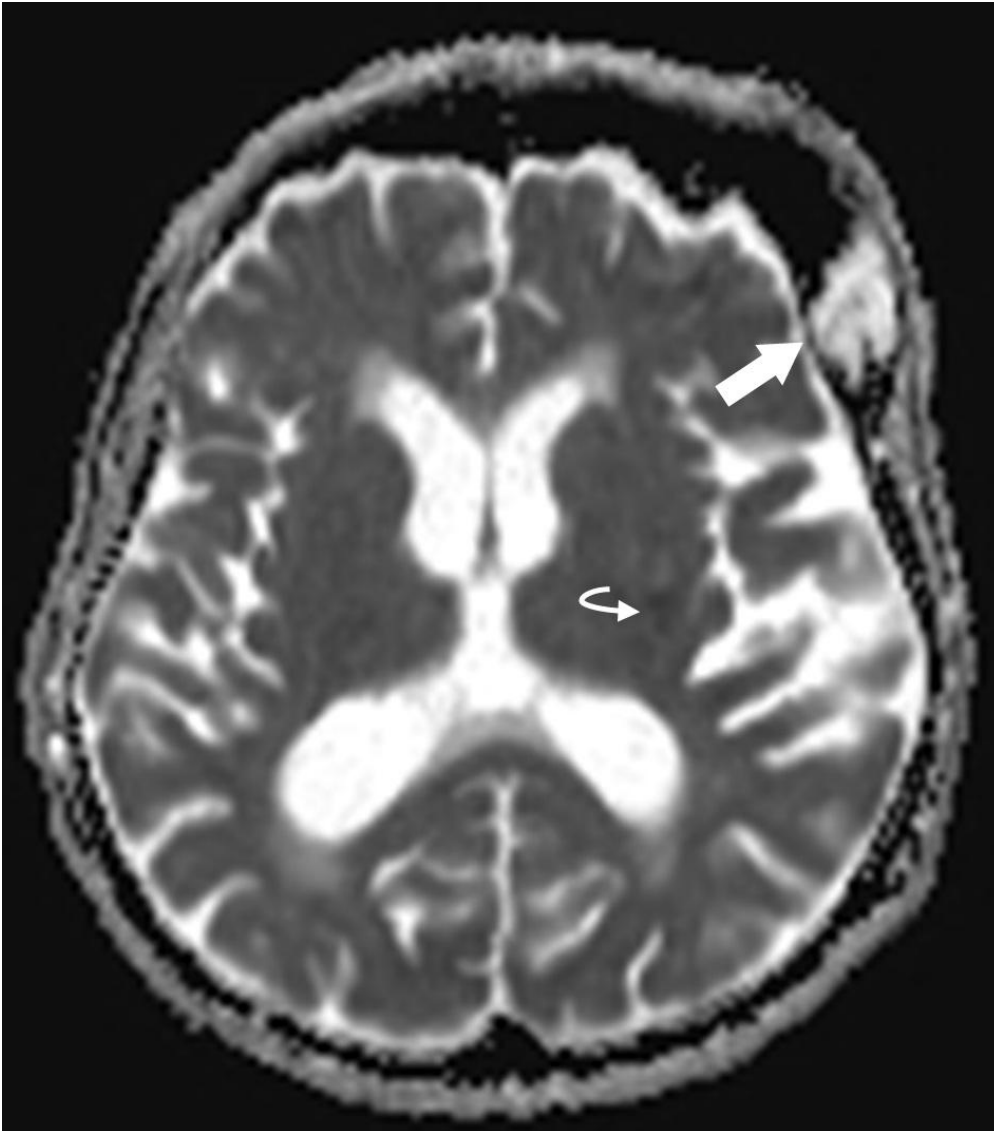
e



Description: Diffusion weighted image (b1000 value): The osseous lesion demonstrates no diffusion restriction (arrow). A small area of restricted diffusion is seen in the left lentiform nucleus (curved arrow).

Origin: © Department of Radiology, RZ Heilig Hart Tienen, Belgium 2019.

f



Description: ADC map: The osseous lesion demonstrates no diffusion restriction (arrow). The lesion seen in the left lentiform nucleus is dark, i.e. low ADC value (curved arrow).

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