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Intracranial Off-Midline Mature Teratoma and Pneumosinus Dilatans: A Unique Clinical Report

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Abstract

Teratomas typically arise as midline lesions in the suprasellar and pineal regions. Pneumosinus dilatans is a rare condition characterized by the expansion of one or more of the paranasal sinuses and thinning of their bony walls with a normal covering mucosa. It usually involves the sphenoid and posterior ethmoid sinuses and has been associated with meningiomas and arachnoid cysts. Off-midline mature teratomas are uncommon, and no reports have described an association with pneumosinus dilatans. We present a rare association between an intracranial off-midline mature teratoma and pneumosinus dilatans in an 18-year-old male patient who presented with a second episode of a left-sided seizure, which has not yet been reported in the literature.

Keywords

- mature teratoma
- pneumosinus dilatans
- Intracranial
- magnetic resonance imaging
- computed tomography
- differential diagnosis

Case History

An 18-year-old man was hospitalized for a second episode of left-sided seizure. Magnetic resonance imaging revealed an extra-axial, $35 \times 40 \times 43$ mm off-midline mass in the right frontal region. The mass was heterogeneous and hypointense on the T2-weighted (T2W) images. This included high signals on T1W images. A partial diffusion abnormality was observed on diffusion-weighted image (DWI). Susceptibility-weighted images demonstrated low signal intensity related to calcification and/or hemorrhage. On fat-saturated contrast-enhanced T1W images, there was no enhancement in the mass, but areas of fat saturation (►**Fig. 1**). Contrast-enhanced computed tomography examination demonstrated a mass with marked hypodense areas characteristic of fat, calcification, and enhanced solid components. There was asymmetric expansion of the sphenoethmoidal paranasal sinus adjacent to the intracranial tumor without any bone overgrowth (►**Fig. 2**). Surgery was performed with total removal of the tumor (►**Fig. 3**). The pathological diagnosis was an intracranial mature teratoma (►**Fig. 4**).

Discussion

Intracranial teratomas account for less than 0.6% of all intracranial tumors.[1] They usually arise in midline structures

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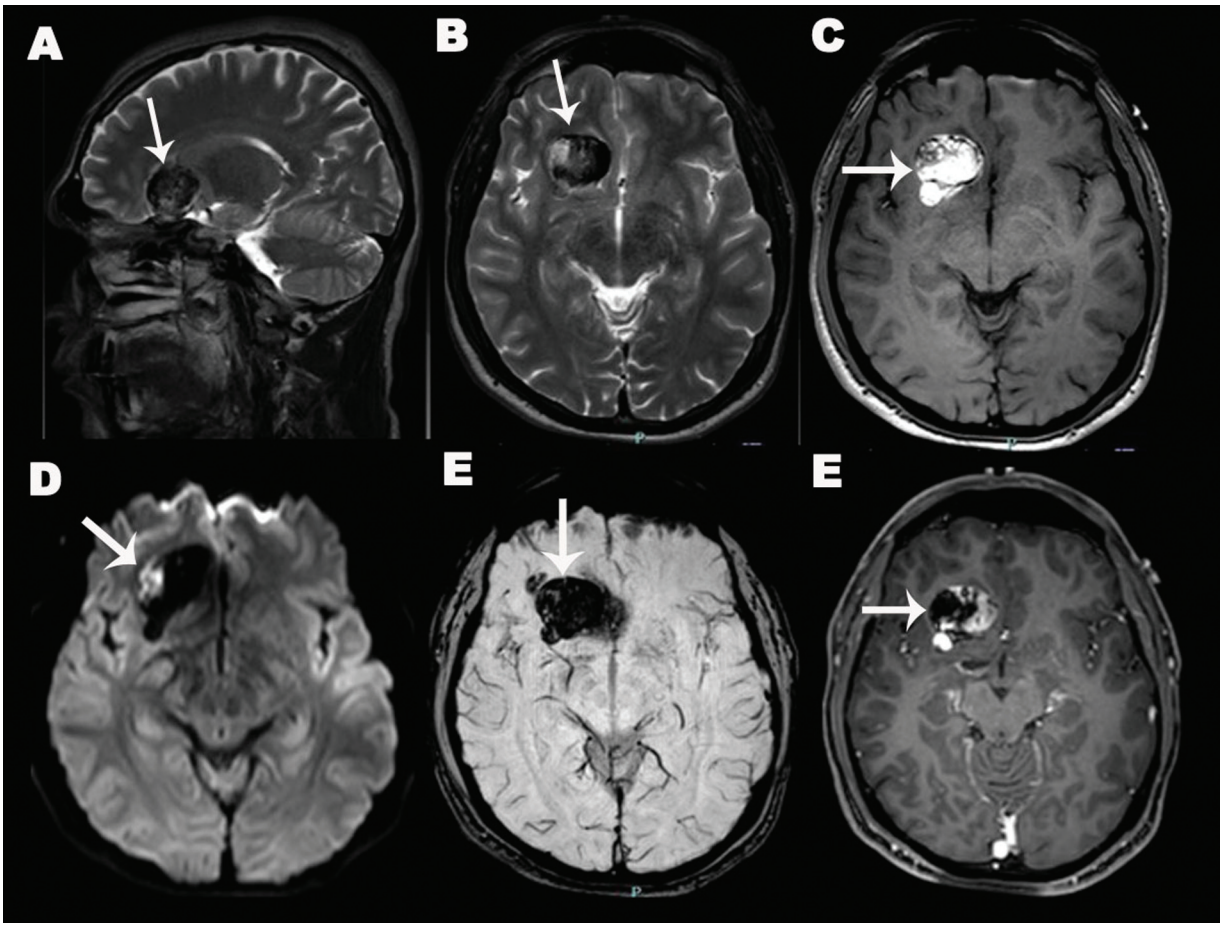

Fig. 1 Preoperative brain magnetic resonance imaging (MRI). (**A**, **B**) Sagittal and coronal fat-saturated T2-weighted (T2W) images showing an off-midline extra-axial heterogeneous mass (arrows) with mixed signal intensities in the left frontal region. No perifocal edema was observed. (**C**) Axial TIW image without fat saturation showing prominent hyperintense signals in the mass (arrow), suggesting fat. (**D**) There is a focal hyperintense diffusion abnormality in the lateral part of the mass (arrow) on diffusion-weighted image. (**E**) Susceptibility-weighted image clearly shows hypointense signals within the mass due to calcification or hemorrhage (arrow). (**F**) Axial contrast-enhanced T1-weighted image with fat saturation showing suppression of the fat signal intensity (arrow).

such as the suprasellar and pineal regions.[2] Mature teratomas are benign tumors that consist of completely differentiated ectodermal, mesodermal, and endodermal elements.[3] Only 13 cases of intracranial off-midline mature teratomas have been reported in the literature. Six of the patients were male, and seven were female. The mean age at presentation was 27.7, with a range of 0.1 to 70. Nine of the masses were located to the left and four to the right of the midline. There were two intra-axial masses. Clinical presentations were dependent on mass location and size and, as a result, were variable. An overview of pertinent findings in intracranial off-midline mature teratoma patients is summarized in ►**Table 1**.[4–16]

On imaging, the majority of intracranial mature teratomas tend to be heterogeneous due to their extremely variable histological components. They usually have solid and cystic components, with some fat, calcifications, and multilocularities. Due to the existence of tumors with capsules and an intact blood–brain barrier, the lesions typically do not show perilesional cerebral edema on imaging studies. Solid components may show enhancement on contrast-enhanced examination.[17] These lesions may show restricted diffusion on DWI.[5,9] Mature cystic teratomas of the temporal lobe may resemble epidermoid tumors, gangliogliomas, or dysembryoplastic neuroepithelial tumor.[5]

Pneumosinus dilatans (PSD) is characterized by the expansion of one or more paranasal sinuses beyond their normal anatomic limits and was first named by Benjamins in 1918.[18] To date, there have been reports that PSD is associated with meningiomas and arachnoid cysts.[19–21] PSD can be divided into two categories: primary, in which no underlying structural abnormalities are present; and secondary, in which other underlying causes may be identified. It is proposed that PSD should always be thoroughly investigated for skull base meningiomas and that bone remodeling is caused by focused dural tension on an adjacent sinus.[22–25] Thus, we considered lipomatous meningioma as a differential imaging diagnosis in the current case due to the knowledge of the relationship between PSD and meningioma. Lipomatous meningioma is an extremely rare subtype of meningioma and is classified as a metaplastic meningioma according to the World Health Organization classification, which may involve all mesenchymal tissues, including osseous, cartilaginous, lipomatous, and myxoid tissues.[26]

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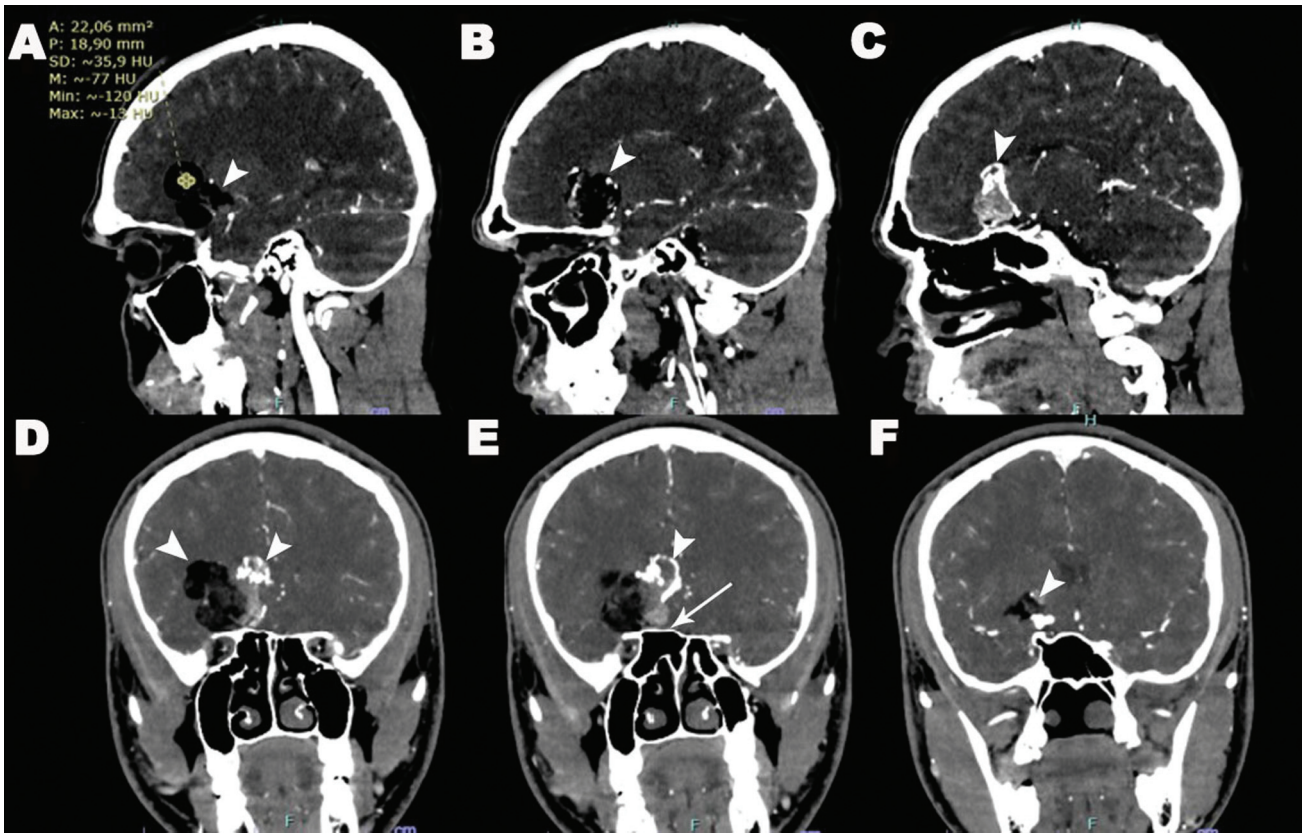

Fig. 2 Diagnostic preoperative contrast-enhanced sagittal (**A**–**C**) and coronal (**D**–**F**) computed tomography (CT) reformatted images show a mass with areas of marked hypoattenuation (mean −77 HU) characteristic of fat, calcification, and enhancing solid components (arrowheads). There is sphenoethmoidal pneumosinus dilatans (PSD) on the right side adjacent to the intracranial tumor (arrows).

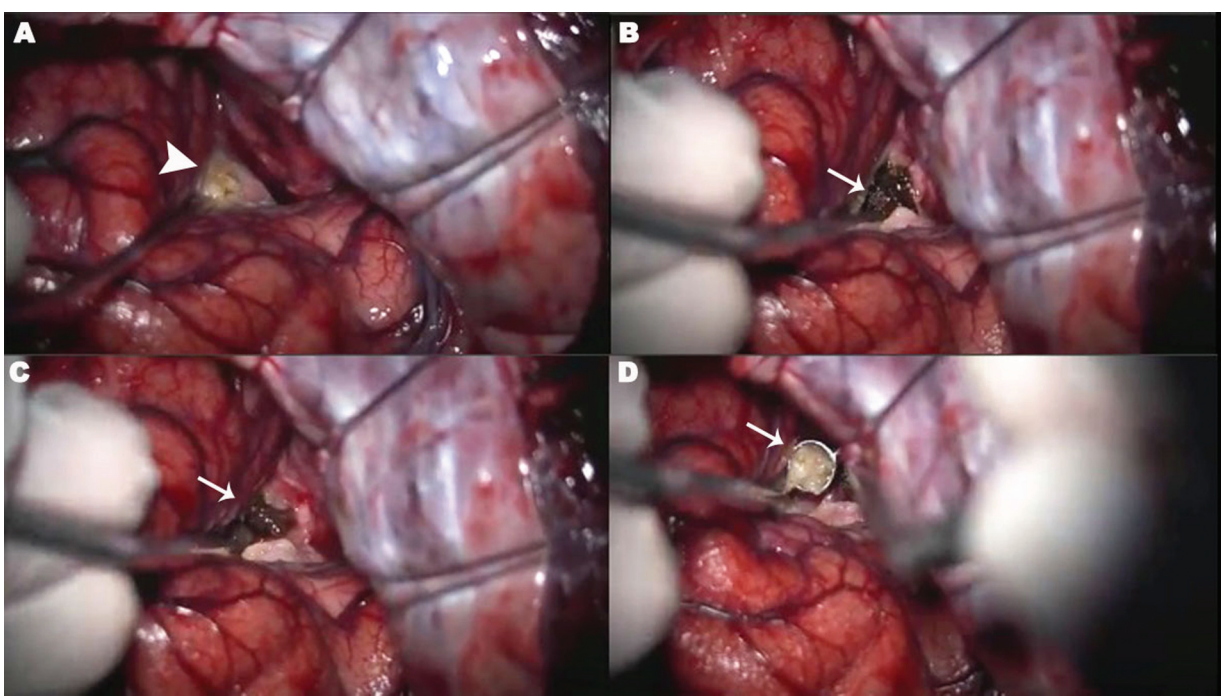

Fig. 3 Selected intraoperative photographs showing an oval or lobulated creamy-yellow mass with a smooth surface (**A**). The lesion contains heterogeneous solid soft tissues and a cystic area with fat, calcification, and hair shafts (**B**–**D**).

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Table 1 Intracranial off-midline mature intracranial teratomas

Authors, year/reference	Age (y)	Sex	Presenting symptoms	Preoperative diagnosis	Intracranial location	Treatment
Nishigaya et al, 1994[6]	67	Male	No	NA	Left Sylvian fissure	NA (found at autopsy)
Lee et al, 1996[7]	35	Male	Dysphagia	Teratoma	Left middle cranial fossa	Surgical resection
Phadke et al, 2004[8]	25	Male	Bony swelling, paresis of cranial nerves	Aneurysmal bone cyst, giant cell tumor, or epidermoid cyst	Right middle cranial fossa	Surgical resection
Khan et al, 2013[9]	11	Female	Left-sided facial weakness	Large temporal bone mass, epidermoid cyst	Left temporal bone and cerebellopontine angle	Surgical resection
Dimov et al, 2013[10]	24	Male	Headache, nausea, vomiting, and fatigue, right-sided weakness, proptosis of the eye	NA	Left fronto-temporo-basal and left orbital	Surgical resection
Hoyer et al, 2013[11]	38	Male	Generalized seizure	NA	Left frontal and temporal lobes	NA (found at autopsy)
Zhang et al, 2012[12]	70	Female	Headache, vomiting, and gait disturbance	Enhancing mass with calcification, fat, and hemorrhage	Right cerebellopontine angle	Surgical resection
Waters et al., 1986[13]	0.1	Female	İntermittent headaches and neck stiffness	A cerebellopontine mass with soft tissue, calcium, and fat densities	Right cerebellopontine angle	Surgical resection
Yin and Guo, 2021[14]	10	Female	Poor appetite, sleep disorder, and poor defecation	Irregular cystic patches of mixed long and short T1 signals and mixed long and short T2 signals - teratoma	Left cerebellopontine angle	Surgical resection
Rijal et al, 2022[5]	17	Male	Abnormal movement of the lips and tongue	Cystic septated mass with restricted diffusion (epidermoid cyst, ganglioglioma, DNET)	Right temporal lobe	Surgical resection
Marques et al, 2023[15]	36	Female	Chronic periorbital headache associated with photophobia, nausea, and vomiting	Meningioma	Left temporal fossa	Surgical resection
Inojie et al, 2021[4]	26	Female	Asymptomatic-incidental	Fat-containing nonenhancing mass	Over the left anterior clinoid process	Surgical resection
Sharma et al., 2019[16]	0.9	Female	A progressively increasing swelling in the left supra-auricular region	A heterogeneous relatively well-defined mass with macroscopic fat content and a few hypodense areas with fluid attenuation and chunky peripheral calcification	Left petrous temporal bone	Surgical resection

Abbreviations: DNET, dysembryoplastic neuroepithelial tumor; NA, not applicable.

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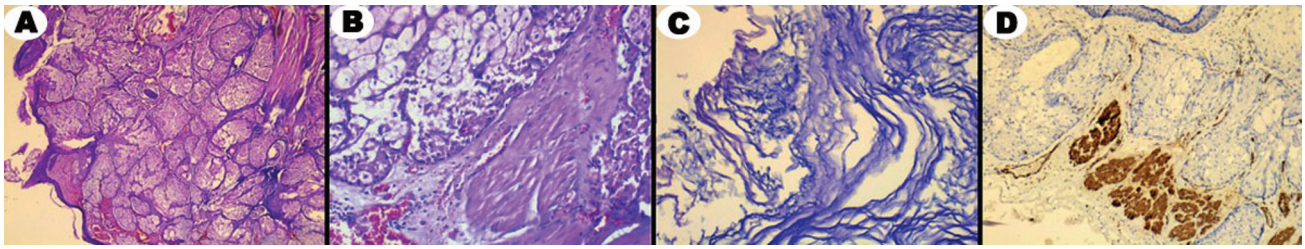

Fig. 4 Photomicrographs of the tumors. (**A**) ×100, hematoxylin and eosin (H&E), showing cyst wall lined by squamous epithelium, hair follicles with sebaceous glands, and smooth muscle fibers (upper right corner). (**B**) ×200, H&E, showing sebaceous glands (upper left) and smooth muscle fibers (central right). (**C**) ×400, H&E staining, showing loose keratin flakes. (**D**) ×100, smooth muscle actin (SMA), confirms the existence of the smooth-muscle cell component.

Conclusion

This is the first time that an intracranial off-midline mature teratoma with PSD has been reported, highlighting this unexpected association. The recognition of an extra-axial location, macroscopic fat, calcification, and a solid-enhancing component of an off-midline tumor with PSD association should also suggest a mature teratoma in the differential diagnosis besides a meningioma.

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Conflict of Interest
None declared.

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