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Case Report

Acute headache and seizures in psychiatric patient revealing atypical location of a ruptured dermoid cyst ☆

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ABSTRACT

Intracranial dermoid cysts are rare slow-growing cystic lesions. They are frequently extra-axial, intra-axial localization is very rare. These benign congenital ectodermal inclusions cysts have a rare risk of rupture. Ruptured dermoid cysts can manifest with headache, epilepsy seizure, cerebral infarction, meningitis, and hydrocephaly. Neuroimaging features are quite characteristic. We report a case of a 30-year-old male who presented to the emergency room with subacute-onset headaches. CT scan and MRI show a ruptured intracranial dermoid cyst.

Introduction

Intracranial dermoid cysts are very rare, they occur in just 0.04%–0.6% of intracranial tumors. These congenital tumors often derive from ectopic cell remnants, which are incorporated into the neural tube. They are usually located along the midline. Dermoid cysts usually remain asymptomatic. Ruptured dermoid cysts can cause various clinical aspects such as epileptic seizures, meningitis, focal neurological deficits, and hydrocephalus. Sudden death has also been reported.

Case report

We report a case of a 30-year-old male, followed in the psychiatric department for bipolar disorder, who presented to the emergency room with subacute-onset headaches that did not respond to medical treatment. An MRI was requested, which identified a lesion in the left frontal horn that was roughly oval-shaped, well-defined, and showed hyperintensity on T1 and heterogeneous hyperintensity on T2-weighted sequences without restriction on diffusion sequence (Figs. 1 and 2).

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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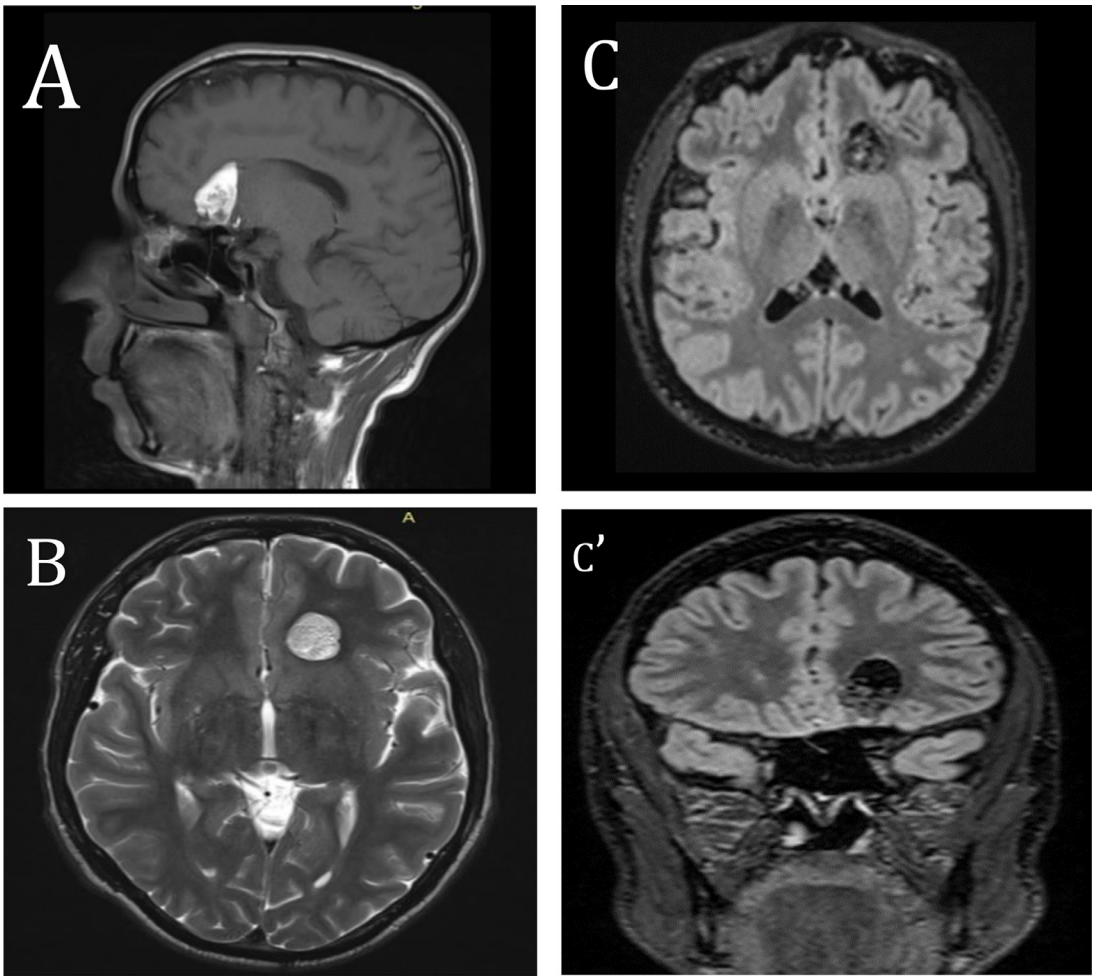

Fig. 1 – Images showing intra-axial lesion with a hypersignal on T1-weighted image on sagittal section (A), with hypersignal on axial section on T2-weighted image (B) and heterogenous hypersignal on FLAIR on coronal and axial sections (C and C').

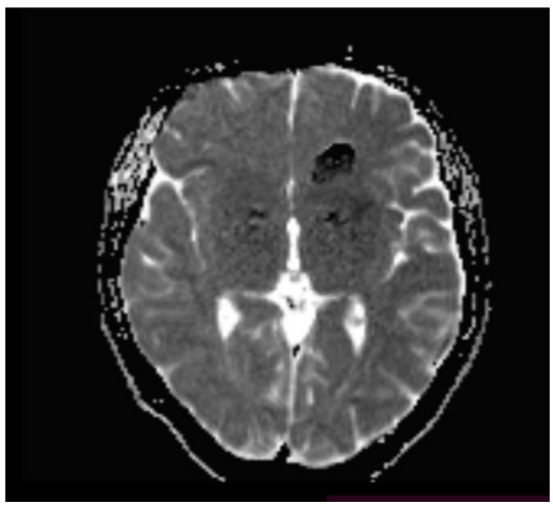

Fig. 2 – Axial diffusion image showing the absence of restriction at B1000.

The lesion partially disappeared on fat saturation sequences (Fig. 3) and contained calcifications (Fig. 4) on the inversion recovery sequence. This lesion extended intimately into the frontal subarachnoid spaces and the opto-chiasmatic cistern and was in close contact with the left optic nerve in its prechiasmatic segment (Fig. 5). Several pinpoint-like formations with hypersignal on T1, a signal on SWI with blooming effect were also noted in the lateral ventricle and the subarachnoid spaces bilaterally, predominantly in the basal frontal region, and significant of a ruptured dermoid cyst (Fig. 6). Our patient received conservative treatment and was set for a neurology follow-up postsurgery. The patient did not attend any subsequent appointments at the neurology clinic and did not request a follow-up brain MRI.

Discussion

Dermoid cysts are uncommon, noncancerous growths that consist of mature squamous epithelium, along with apocrine,

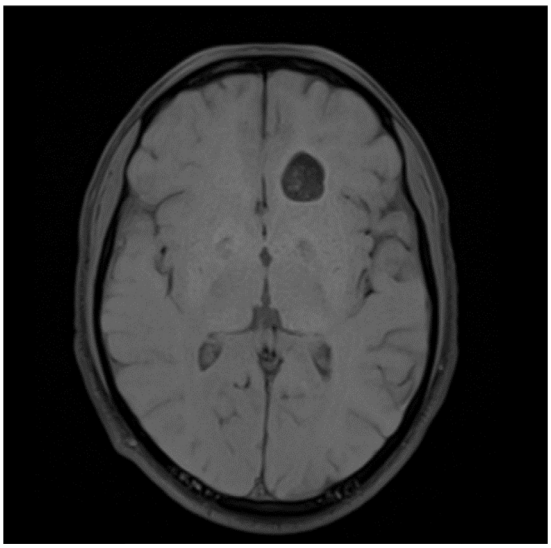

Fig. 3 – The axial section of T1 with fat suppression shows suppression of the internal fatty component.

eccrine, and sebaceous glands, as well as other ectodermal structures like hair follicles and teeth [1].

They represent approximately 0.04%-0.6% of all intracranial tumors [2] with slightly female predominance [3]. Typically, dermoid cysts are diagnosed in individuals during the first 3 decades of life [3].

These extra-axial tumors are mostly found in the midline suprasellar region, followed by the posterior fossa (vermis and fourth ventricle), frontobasal, or temporobasal regions, and occasionally in the spine [4]. Intra-axial localization is very rare, it might result from the proliferation of multipotent embryonic cells, followed by lateral migration of ectodermal remnants along the pathways of development of the optic vesicles and neurovascular structures that could lead them to their final location along the Virchow and Robin spaces, where they become associated with vascular structures [5]. As for spinal dermoid cysts, 60% are located in the lumbosacral region, 25% in the sacrococcygeal region, 10% in the dorsal region, and 5% in the cervical region [6].

Dermoid cysts develop from embryonic remnants of ectodermal tissue that are implanted in an abnormal manner (totipotent ectodermal cells) and become incorporated into the neural tube during its closure, typically occurring between the third and fifth week of embryonic life [7].

Histologically, the cyst wall of dermoid cysts is characterized by a thick, layered squamous epithelium capsule that contains dermal elements, such as sebaceous glands, sweat glands, and hair follicles [8,9]. Dermoid cysts usually grow slowly and collect thick, yellowish material composed of shed epithelial cells, sebaceous gland secretions, fat, oil, and hair [9,10].

Clinically, the symptoms of dermoid cysts depend on their location, size, and whether they have ruptured. Most often, dermoid cysts grow slowly and can reach a significant size before causing symptoms. They can be the cause of headaches, visual disturbances, increased intracranial pressure due to hydrocephalus in cases of dermoid cysts in the posterior cranial fossa, and seizures [11]. The growth of dermoid cysts is associated with the accumulation of shed epithelial cells and secretions from sebaceous glands, which can vary with hormonal changes related to age [7]. While malignant transformation into squamous cell carcinoma is exceedingly rare, has been documented [12].

It is believed that dermoid cysts may rupture as a result of the production of hair and oils from internal dermal elements, leading to increased pressure [13]. However, rupture can also be preceded by cranial and medullar trauma [14]. A rupture of the cyst can manifest as a sudden headache, seizure, or, in more severe cases, complications like chemical meningitis, vasospasm, and cerebral infarction [15].

Diagnosis of ruptured dermoid cysts is based on imaging, including both computed tomography (CT) and magnetic resonance imaging (MRI).

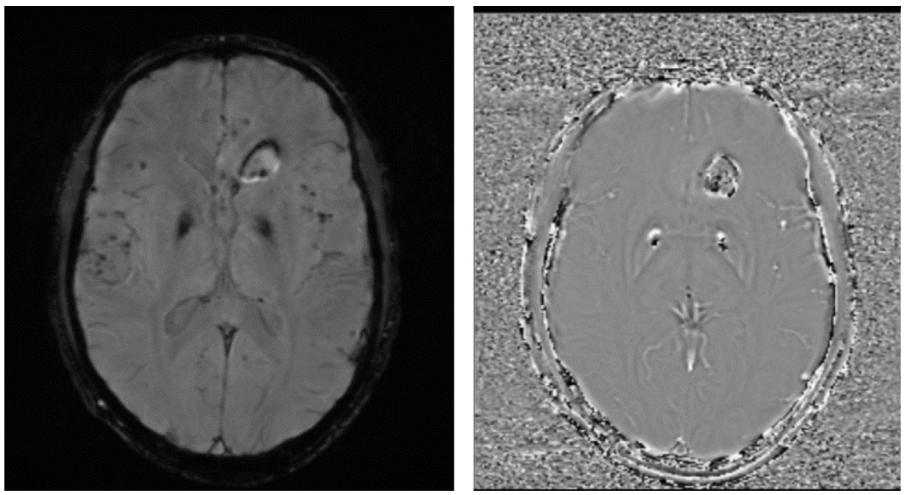

Fig. 4 – Axial SWI image showing heterogenous high and low T2 signal, with hyposignal component on phase posttreatment, related to the presence of calcifications.

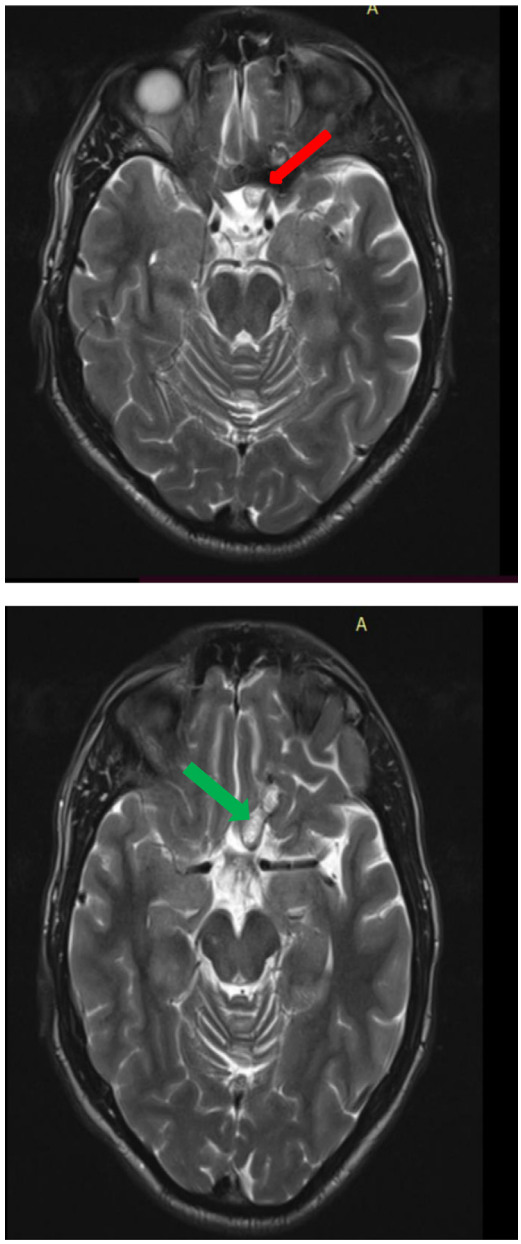

Fig. 5 – Axial sections of T2-weighted sequence showing that the lesion extended intimately into the frontal subarachnoid spaces (Red arrow) and the opto-chiasmatic cistern (green arrow) and was in close contact with the left optic nerve in its prechiasmatic segment.

In a CT scan, the appearance of an intracranial dermoid cyst is highly distinctive. These cysts are usually well-defined and appear as low-attenuation lesions because of their fat content. However, they may contain varying proportions of fat, hair, and epidermal debris. Calcifications can also be observed, with reports of them occurring in about 20% of cases [16].

In MRI, dermoid cysts, due to their fat content, typically appear as high-signal mass lesions on T1-weighted, variable signal on fast spin echo T2-weighted, and FLAIR (fluid-attenuated inversion recovery) sequences, with no restriction on diffusion sequence [15]. However, in some cases, these lesions may ex-

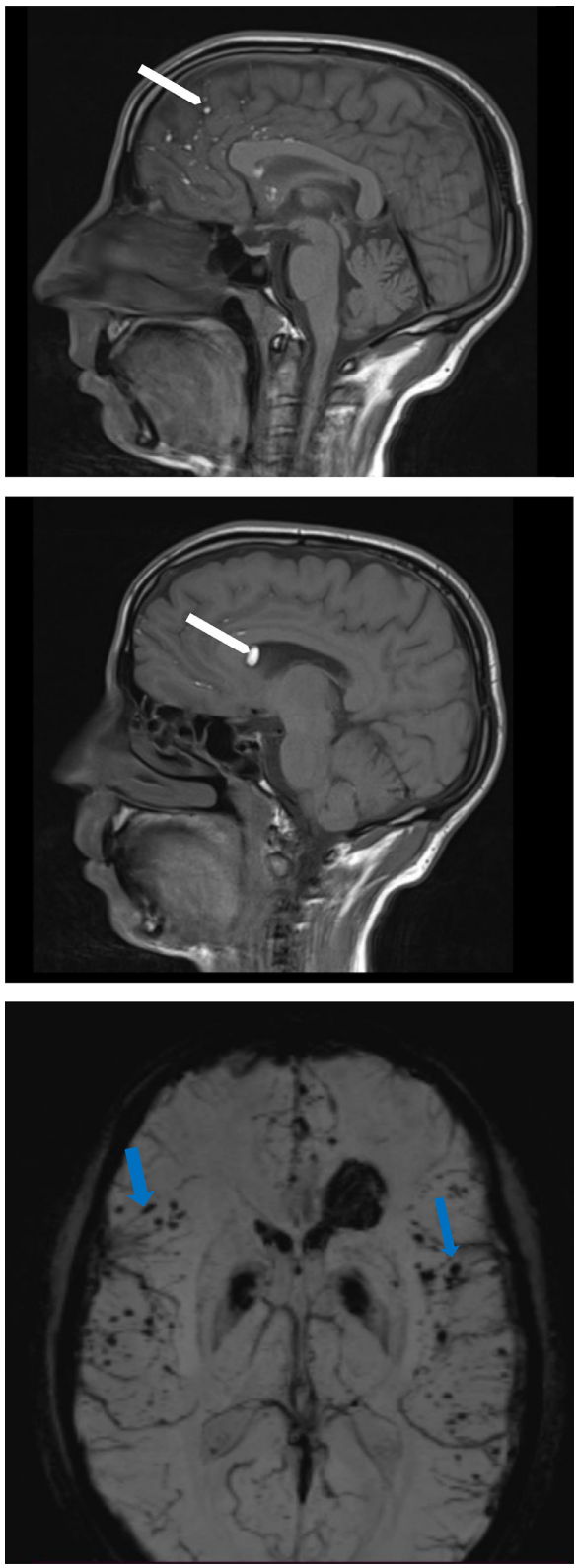

Fig. 6 – Sagittal section on T1-weighted sequence showing several pinpoint-like formations with hypersignal (White arrow), and with a signal on SWI with<<blooming effect >> (blue arrow) in the lateral ventricle and the subarachnoid spaces bilaterally, significant of a ruptured dermoid cyst.

hibit a more varied appearance with minimal enhancement. This variability can be attributed to the presence of calcifications, hair, epithelial debris, and sebaceous secretion within the cyst, which can result in a heterogeneous appearance on imaging [4].

Ruptured dermoid cyst occurs in the dispersion of lipid droplets in the ventricle, subarachnoid spaces, and cortical sulci. It appears with a hyperintensity signal on T1-weighted images while their signal is suppressed on fat-suppression sequences. On T2* and susceptibility-weighted imaging (SWI) sequences those lipid droplets cause blooming artefact [15]. This blooming effect is a susceptibility artefact defined by areas of low signal due to the presence of paramagnetic substances [17].

Intraventricular fat-fluid level and leptomeningeal enhancement due to chemical meningitis can also be found in the ruptured dermoid cyst. [4,15] Malignant transformation to carcinoma is extremely rare [18].

The main differential diagnosis of unruptured dermoid cysts is primarily made with tumors showing hypersignal on T1-weighted images.

The epidermoid cyst is the flagship of these tumors; it is an extra-axial tumor rich in mucus that appears as hyperintense on T1 with heterogeneous signal on T2, and fluid attenuation similar to LCR. On diffusion-weighted sequences, it appears hyperintense, which is the key element for distinguishing it from the dermoid cyst [4,15].

Other tumors with fatty content can also pose differential diagnostic challenges, such as immature teratomas and intracranial lipomas.

Intracranial lipomas are congenital lesions, usually asymptomatic, widely distributed in an intracranial compartment, with predilection in pericallosal region (50%) where they are often associated with corpus callosum agenesis [19]. On MRI, they appear hypersignal on T1 and T2 due to the fat content, with low signal on low signal on fat saturated sequences, and a blooming effect on susceptibility-weighted imaging (SWI) [19].

Regarding ruptured dermoid cysts, the main differential diagnosis is primarily made with lesions that generate a blooming effect on SWI (susceptibility-weighted imaging) or T2* sequences. Magnetic susceptibility sequences have sensitivity to compounds with paramagnetic, diamagnetic, and ferromagnetic properties, allowing them to distort the local magnetic field and therefore create the blooming effect [17].

Several substances can be responsible for this artefact; the deposition of hemosiderin in the context of acute subarachnoid hemorrhage or the presence of multiple intraparenchymal cavernomas can mimic the rupture of a dermoid cyst. Also, cerebral gas emboli and calcifications can also distort the diagnosis. [15,17]

When it is symptomatic, surgical resection is the treatment of choice to relieve the mass effect causing neurological deficit. The surgical approach for dermoid cysts entails 3 primary steps: incising the cyst's capsule, extracting its contents to reduce its size internally, and microsurgically dissecting the capsule from any adhering or nearby neurovascular structures [10].

In the case of a ruptured cyst, the dispersion of fat droplets is extensive, and there is neither a necessity nor a feasibility to extract all disseminated fat droplets. An MRI is essential prior to surgical treatment due to its capacity to reveal the extent of the lesion and its proximity to the local neurovascular structures [4,20].

Ruptured intracranial dermoid cysts are relatively rare, which can cause not only headaches and seizures but also some serious complications such as chemical meningitis, vasospasm, cerebral infarction, and death.

MRI is obligatory to assess the diagnosis. Magnetic susceptibility sequences, such as SWI or T2*, generating the blooming effect, related to disseminated fat droplets in the subarachnoid space or ventricles, is quite characteristic of ruptured intracranial dermoid cysts.

Patient consent

Written informed consent for publication was obtained from the patient.

REFERENCES

[1] Muçaj S, Ugurel MS, Dedushi K, Ramadani N, Jerliu N. Role of MRI in diagnosis of ruptured intracranial dermoid cyst, case report. Acta Inform Med 2017;25:141–4.

[2] Guidetti B, Gagliardi FM. Epidermoid and dermoid cysts. Clinical evaluation and late surgical results. J Neurosurg 1977;47:12–18.

[3] Mehta, Minesh P. Principles and practice of neuro-oncology: a multidisciplinary approach. Demos Medical Publishing; 2010.

[4] Liu JK, Gottfried ON, Salzman KL, Schmidt RH, Couldwell WT. Ruptured intracranial dermoid cysts: clinical, radiographic, and surgical features. Neurosurgery 2008;62(2):377–84.

[5] Osborn AG, Preece MTT. Intracranial cysts: radiologic-pathologic correlation and imaging approach. Radiology 2006;239(3):650–64.

[6] Calabro F, Capellini C, Jinkins JR. Rupture of spinal dermoid tumors with spread of fatty droplets in the cerebrospinal fluid pathways. Neuroradiology 2000;42:572–9.

[7] Stendel R, Pietila TA, Lehmann K, Kurth R, Suess O, Brock M. Ruptured intracranial dermoid cysts. Surg Neurol 2002;57:391–8.

[8] Lunardi P, Missori P. Supratentorial dermoid cysts. J Neurosurg 1991;75:262–6.

[9] Lunardi P, Missori P, Rizzo A, Gagliardi FM. Chemical meningitis in ruptured intracranial dermoid. Case report and review of the literature. Surg Neurol 1989;32:449–52.

[10] Yas¸argil MG, Abernathey CD, Sarioglu AC. Microneurosurgical treatment of intracranial dermoid and epidermoid tumors. Neurosurgery 1989;24:561–7.

[11] Chen JC, Chen Y, Lin SM, Tseng SH. Sylvian fissure dermoid cyst with intratumoral hemorrhage: case report. Clin Neurol Neurosurg 2005;108:63–6.

[12] Ciurea AV, Coman T, Tascu A, Ionescu V. Intradural dermoid tumor of the posterior fossa in a child with diastematobulbia. Surg Neurol 2005;63:571–5.

[13] Osborn AG, Preece MT. Intracranial cysts: radiologic-pathologic correlation and imaging approach. Radiology 2006;239:650–64.

[14] Esquenazi Y, Kerr K, Bhattacharjee MB, Tandon N. Traumatic rupture of an intracranial dermoid cyst: case report and literature review. Surg Neurol Int 2013;4:80.

[15] Jacków J, Tse G, Martin A, Sąsiadek M, Romanowski C. Ruptured intracranial dermoid cysts: a pictorial review. Pol J Radiol 2018;83:e465–70. doi:10.5114/pjr.2018.80206.
[16] Erasmus LJ, Hurter D, Naudé M, Kritzinger HG, Acho S. A short overview of MRI artifacts. SA J Radiol 2004;8:13–17.
[17] Mittal P, Kalia V, Dua S. Pictorial essay: susceptibility-weighted imaging in cerebral ischemia. Indian J Radiol Imaging 2010;20(4):250–3. doi:10.4103/0971-3026.73530.
[18] Wulfovich S, Tucker S, Levy M, Crawford JR. Incidental cerebellar dermoid cyst mimicking low grade glioma in a teenager. BMJ Case Rep 2021;14(2):e241227.
[19] Truwit CL, Barkovich AJ. Pathogenesis of intracranial lipoma: an MR study in 42 patients. AJR Am J Roentgenol 1990;155(4):855–64 discussion 865. doi:10.2214/ajr.155.4.2119122.
[20] Carvalho GA, Cervio A, Matthies C, Samii M. Subarachnoid fat dissemination after resection of a cerebellopontine angle dysontogenic cyst: case report and review of the literature. Neurosurgery 2000;47:760–4.