European Review for Medical and Pharmacological Sciences 2024; 28: 1970-1975

Atypical dermoid cyst of the corpus callosum: a case report

A. SPALLONE[1,2], K. IVANOVA[1], L. FERRANTE[2], A.A. BELOGUROV JR.[1,3]

[1]Shemyakin and Ovchinnikov Institute of Bioorganic Chemistry, Russian Academy of Sciences, Moscow, Russian Federation
[2]Neurosciences Department, Neurological Centre of Latium, Rome, Italy
[3]Department of Biological Chemistry, Evdokimov Moscow State University of Medicine and Dentistry, Moscow, Russian Federation

Abstract. – BACKGROUND: Intracranial dermoid cysts (DCs) represent an infrequent subset of congenital ectodermal inclusion cysts predominantly observed near the midline structures. In spite of their benign nature, they can cause clinical manifestations, necessitating surgical removal as the main therapeutic measure.

CASE REPORT: We present here an extremely rare case characterized by a radiologically atypical dermoid cyst located within the corpus callosum, an extremely rare location for such tumors. Successful surgical excision resulted in good clinical outcomes.

CONCLUSIONS: This paper underscores the importance of a timely, proper radiological diagnostic process, which sees magnetic resonance imaging (MRI) as the main step, as well as the fact that interpretation of MRI data can sometimes be challenging, as it was in the patient of this report.

Key Words:
Intracranial dermoid cysts, Corpus callosum.

Background

Intracranial dermoid cysts (DCs) are congenital ectodermal inclusion cysts originating from ectopic epithelial cells that are part of the neural tube. This origin explains their typical location near the midline. They are rare, constituting 0.3-0.7% of intracranial tumors, making them less common than epidermoid cysts[1,2].

DCs present as well-defined, lobulated, "pearly" masses of variable size. They are pathologically characterized by a thick, stratified squamous epithelium cyst wall that contains dermal elements[3]. DCs increase in size through glandular secretion and epithelial desquamation. Though benign and slow-growing, they can cause focal neurological signs through encroachment on neurovascular structures and, occasionally, through rupture. Generally, DCs carry a favorable prognosis, with surgical resection being the treatment of choice[4]. Intracranial DCs typically occur in the midline, involving the sellar, parasellar, and frontonasal regions. Some may be located in the posterior fossa, either as vermian lesions or within the fourth ventricle. Radiologically, they usually appear hyperintense on T1, hypo- to hyperintense on T2, and do not exhibit contrast enhancement[5]. The symptoms of patients with intracranial dermoid cysts depend on the tumor's location, depth, size, and whether it has ruptured[6]. In this report, we describe a radiologically atypical, substantial dermoid cyst situated in the corpus callosum, which was surgically excised with favorable results.

Case Presentation

A 22-year-old right-handed female was admitted to our center following a new onset of Jacksonian seizure accompanied by loss of consciousness. Upon admission, the patient had no memory of the event and exhibited post-ictal confusion and urinary incontinence. When properly questioned a few hours later, she reported experiencing left-sided paresthesias, including in her face, and a persistent headache, which had gradually intensified over the preceding two months. Both general and neurological examinations were within normal limits. Complete blood count, serum electrolytes, and coagulation studies revealed no abnormalities.

The x-rays of the skull showed no significant abnormalities. Subsequent magnetic resonance imaging (MRI) (Figure 1) of the brain using

Corresponding Author: Aldo Spallone, MD; e-mail: aldo.spallone@nclroma.it

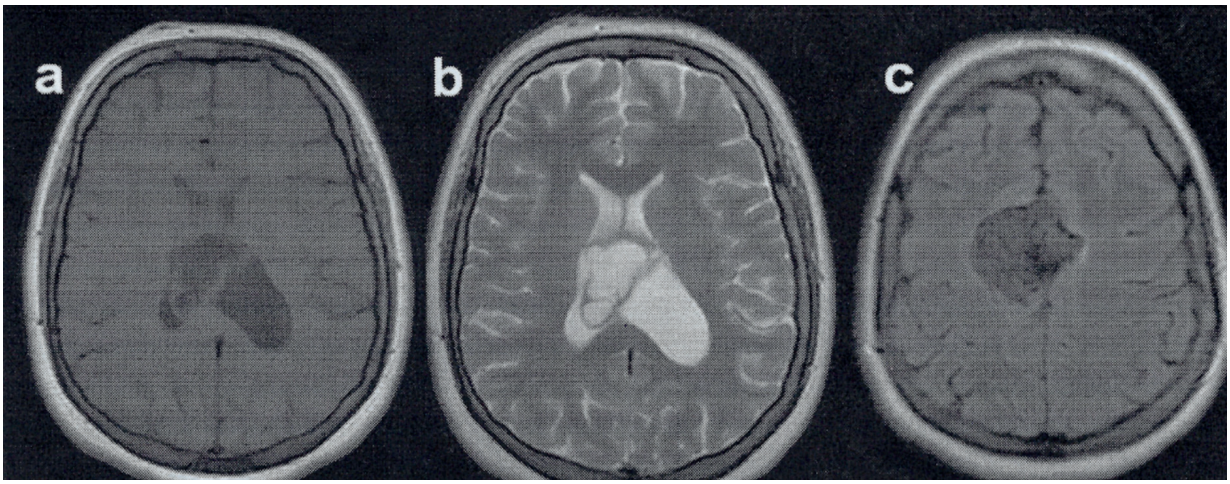

Figure 1. MRI conventional sequences of the present case. **a**, T1 Axial: displays a heterogeneously hypointense lobulated lesion located over the body of the corpus callosum. **b**, T2 Axial: reveals an irregular hyperintense mass, complemented by a hypointense, thick capsule. **c**, FLAIR Axial: illustrates a heterogeneously hypointense mass with an indication of incomplete signal suppression.

conventional sequences [T1, T2, fluid-attenuated inversion recovery (FLAIR)] revealed a non-enhancing, substantial (5×4×5.5 cm), supratentorial expansive lesion situated in the body of the corpus callosum, slightly more to the right. The right lateral ventricle was compressed and displaced, and the contralateral ventricle was enlarged, with initial signs of trans-ependymal edema in the left occipital horn. The cystic lesion appeared hypointense on T1 and hyperintense on heterogeneous T2-weighted images and demonstrated incomplete suppression on FLAIR images. A hypointense rim was also noted at the lesion's periphery, potentially indicating the capsule and/or the dural layer. There was neither surrounding edema nor gadolinium enhancement. Additional sequences were employed to define this lesion, including DWI with ADC maps and T1 3D Fat Sat PG (Figure 2). The mass was hyperintense on DWI but displayed an ADC similar to brain parenchyma.

Total removal of the lesion was achieved *via* posterior frontal parasagittal craniotomy. Histopathological examination of the surgical specimen confirmed the presence of a dermoid cyst with lining epithelium, abundant fat, and scattered hair. A postoperative CT scan confirmed the total removal of the mass (Figure 3).

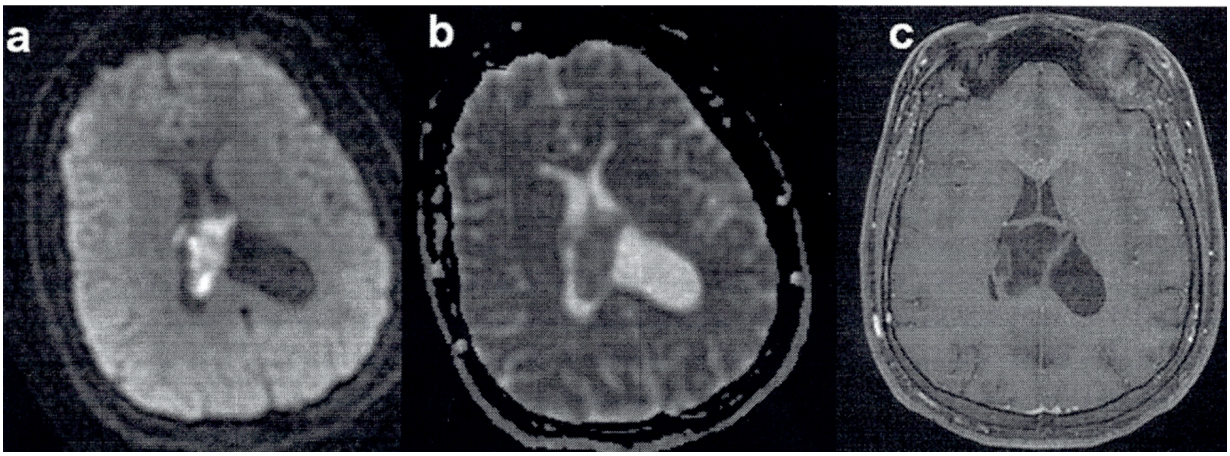

Figure 2. Imaging sequences. **a**, DWI Sequences: present markedly restricted diffusion with hyperintensity. **b**, Relative ADC Maps: exhibit the lesion as isointense to parenchyma. **c**, T1 3D Fat Sat PGR: specific details can be described here depending on the context of the image.

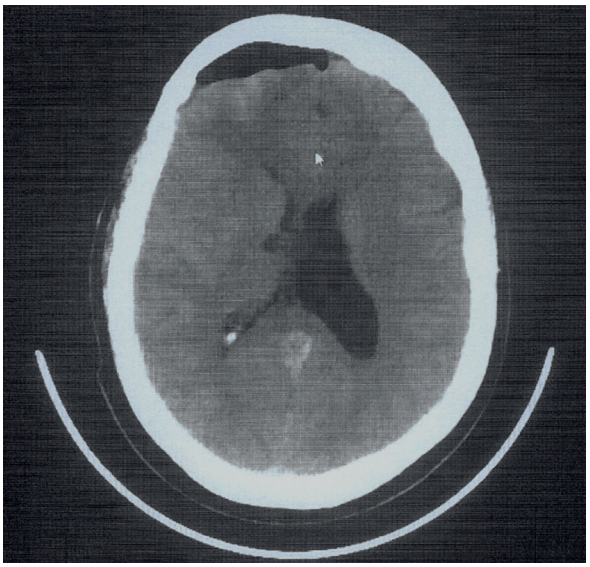

Figure 3. Early postoperative CT scan: illustrates complete removal of the mass, with a few blood remnants observed within the right lateral ventricle and the posterior paraglider region.

Following surgery, the patient exhibited slight left hemiparesis, which fully resolved on the 4th postoperative day. She was discharged on the 6th postoperative day with no neurological deficits. The patient began anticonvulsant therapy, and at the 6-month follow-up she was neurologically intact with no further seizures.

Discussion

Intracranial dermoid cysts (IDCs) are rare, dysembryogenetic cysts that originate from ectodermal inclusions of primitive pluripotent cells[7]. These congenital and gradually progressive tumors constitute approximately 0.3 to 0.7% of all intracranial tumors, and they typically become symptomatic between the third and fifth decades of life[1,2].

Dermoid cysts (DCs) are predominantly situated in the midline, particularly in the cisternal space. The corpus callosum (CC) is the primary interhemispheric commissure connecting homologous territories of both cerebral hemispheres, and its most prevalent congenital abnormalities are total or partial agenesis. Lipomas and, less frequently, arachnoid cysts represent the most recurrent developmental and benign tumors in this location. Dermoid cysts are exceptionally rare in this region, notwithstanding that the CC is a midline structure[8]. To the best of our knowledge, this is among the first few reports of a dermoid cyst located in the corpus callosum[9,10].

IDCs are pathologically identified by the presence of a capsule comprised of simple epithelium bolstered by collagen. In thicker areas, the lining is augmented with a dermis containing hair follicles, sebaceous glands, and apocrine glands[5]. Generally, intact dermoid cysts grow insidiously, expanding in the subarachnoid space, enveloping critical neurovascular structures long before clinical manifestation[11]. Intact IDCs commonly present with headaches, visual deficits, and seizures[1]; ruptured dermoid cysts might manifest with symptoms such as meningitis, acute hemiparesis, cheiro-oral syndrome, arachnoiditis, ventriculitis, hydrocephalus, and cerebral ischemia[12]. A pure lesion of the corpus callosum is likely asymptomatic, with seizures and mental disorders often attributed to accompanying nervous tissue anomalies; however, epilepsy remains a frequent symptom[8].

The clinical diagnosis of IDCs is complex and relies on imaging studies such as computed tomography (CT) or MRI. On MRI, unruptured dermoid cysts share imaging characteristics with fat due to their liquid cholesterol content. They often appear hypointense on T1 with no enhancement and heterogeneous on T2, with intensity ranging from hypo- to hyperintense depending on fat content[5]. However, some dermoid cysts may not exhibit homogeneous hyperintensity on T1, as fat may be absent or other elements like hair or debris might predominate[13]. The cysts typically do not show suppression on FLAIR images and appear hyperintense on DWI and isointense to brain parenchyma on ADC, which assists in differentiating them from other cystic masses. Upon rupture, the presence of T1 hyperintense droplets and leptomeningeal enhancement may be observed, making MRI the optimal imaging modality for diagnosing this rare entity[3]. Unruptured dermoid cysts often exhibit atypical radiological findings, such as heterogeneous, hypointense signals on T1-weighted and hyperintense signals on T2-weighted images[14-19]. All these studies explained the signal intensity characteristics with the amount of "fat" in the cysts, referring to that on T1-weighted sequences.

To quote in particular a few examples, a 10-year-old girl from Morocco previously had an intradiploic dermoid cyst in the left pterion[20]. It was probably formed as a result of insufficient closure of the neural tube during intrauterine development or traumatic migration of skin com-

ponents. Recovery was achieved after surgical resection of the damaged part of the pterion. In another case, a 42-year-old Hispanic woman with an intracranial dermoid cyst underwent craniotomy and craniectomy to resect posterior and anterior tumors[21]. She was discharged from the hospital a week later and then lost in follow-up. A pathomorphological examination of the cyst showed a laceration of the cyst with fat necrosis, hair follicles, keratin, and sebaceous glands. A clinical case with a 34-year-old woman with cerebellar convexity DC with both interdural and extradural components was also reported[22]. The successful treatment consisted of microsurgical decompression. The removed tumor included adipose tissues with desquamated debris and hairs. The patient has effectively recovered.

There are data about a 40-year-old woman with an intracranial dermoid cyst on the left side of the interpeduncular cistern extending to the prepontine cistern[23]. The cyst was surgically removed using a craniotomy. Interestingly, a 54-year-old patient with a cystic lesion with thin marginal enhancement in the anterior and middle cranial fossa with midline shift was described[24]. She also had a craniotomy, and the cyst was removed. After surgery, there were no neurological complications, and the patient was able to perform most of her work. The pathomorphological picture of the cyst showed stratified squamous epithelium linked with fat tissue, calcification, keratinous debris, and cholesterol pockets.

Another case occurred with a 7-month-old boy who was diagnosed with an intracranial nasofrontal dermoid cyst characterized by a hair in the nose tip without a sinus tract identified[25]. At the age of 32 months, the craniotomy was performed, and the cyst was excised. The surgeons used a pericranial flap, and the nose was reconstructed by a temporoparietal fascia graft. The post-operative period included no complications, uneventful, and after a course of antibiotics, the boy was discharged and he was put under otorhinolaryngology and neurosurgery observation. A case report[26] on the treatment of a 22-year-old patient with a posterior fossa dermoid cyst has been published. The therapy involved surgically removing the cyst, resulting in the stabilization of the patient's condition. Consequently, nine months following the surgery, the patient was able to move in a wheelchair due to preexisting cerebral palsy. The pathomorphological picture of the cyst showed that it is benign and includes stratified squamous epithelium with adnexal structures.

Intracranial dermoid cyst rupture is also described in the literature. In a 14-year-old girl, a neoplasm located near the foramen ovale with intra and extradural extensions ruptured and led to the spread of fat molecules into the third ventricle and subarachnoid spaces[27]. As a result, the tumor was resected and the girl was discharged home 7 days later without complications. A similar case occurred with a 24-year-old girl who had an IDC rupture[28]. The cyst was located in the suprasellar space, and lipids were found in the subarachnoid space. Surgical intervention was performed, and the cyst was excised. The girl's headaches and paresthesias were treated symptomatically. Finally, a case report[29] has been published on the occurrence and treatment of an intracranial dermoid cyst within the midline posterior fossa with an occipital transdiploic linear channel in a 39-year-old woman. She had a craniotomy followed by cyst removal, which led to her recovery. Histological examination of the cyst showed that it contains hair shafts, cholesterol molecules, and keratin inclusions.

Summarizing, complete surgical resection following thoughtful dissection from adjacent neurovascular structures is the treatment of choice for symptomatic dermoid cysts, which aims to eliminate the possible complications of its growth, such as its rupture and consequent dissemination along the CSF pathways, recurrent meningitis, while at the same time, it eliminates the compressive-irritative effect on the surrounding structures[4]. However, when the cyst wall adheres strongly to vital or functionally critical structures, part of the lesion may be left *in situ*, increasing the likelihood of recurrence and the need for more challenging and riskier redo surgical interventions. This emphasizes the importance of attempting radical removal whenever feasible[1,4,12,30,31]. This was our intent in the present case, and this was eventually successfully achieved in the present patient. According to the long-term follow-up, excellent radiological and clinical results and the disappearance of symptoms and signs were confirmed.

Conclusions

Early diagnosis of cysts using MRI is extremely important to prevent their rupture and subsequent complications like chemical meningitis. The optimal way to treat the formed tumor is its complete surgical removal, after which the most likely outcome is complete remission of patients.

Informed Consent
Written informed consent was provided by the patient for permission to receive therapy and to publish this case report.

Ethics Approval
This study was conducted in accordance with the Declaration of Helsinki of 1975 (as revised in 2013), and the protocol was reviewed and approved by the Institutional Review Board of Neurological Centre of Latium (project identification number RM2-NCL 2021-000137-12 Rif 046/21).

ORCID ID
Aldo Spallone: 0000-0002-7017-1513
Luigi Ferrante: 0000-0001-9444-972X
Alexey A. Belogurov Jr.: 0000-0002-2033-9621

Authors' Contributions
All authors contributed to the study. A. Spallone made substantial contributions to the conception and design of the study, as well as acquisition of data, analysis, interpretation of data and supervision. Alexey A. Belogurov Jr. made critical revisions related to the relevant intellectual content of the manuscript. L. Ferrante and K. Ivanova performed validation and final approval of the version of the article to be published.

Funding
The publication of the research is supported by the Ministry of Science and Higher Education of the Russian Federation contract No. 075-15-2021-1033 (13.2251.21.0111).

Availability of Data and Materials
Data sharing does not apply to this article as no datasets were generated or analyzed during the current study.

Conflict of Interest
Alexey A. Belogurov Jr, Luigi Ferrante, Karina Ivanova, and Aldo Spallone declare that they have no conflicts of interest in relation to this article.

References

1) Rubin G, Scienza R, Pasqualin A, Rosta L, Da Pian R. Craniocerebral epidermoids and dermoids. A review of 44 cases. Acta Neurochir (Wien) 1989; 97: 1-16.
2) Pant I, Joshi SC. Cerebellar intra-axial dermoid cyst: a case of unusual location. Childs Nerv Syst 2008; 24: 157-159.
3) Liu JK, Gottfried ON, Salzman KL, Schmidt RH, Couldwell WT. Ruptured intracranial dermoid cysts: clinical, radiographic, and surgical features. Neurosurgery 2008; 62: 377-384.
4) Yaşargil MG, Abernathey CD, Sarioglu AC. Microneurosurgical treatment of intracranial dermoid and epidermoid tumors. Neurosurgery 1989; 24: 561-567.
5) Osborn AG, Preece MT. Intracranial cysts: radiologic-pathologic correlation and imaging approach. Radiology 2006; 239: 650-664.
6) Chen JC, Chen Y, Lin SM, Tseng SH. Sylvian fissure dermoid cyst with intratumoral hemorrhage: case report. Clin Neurol Neurosurg 2005; 108: 63-66.
7) Schijman E, Monges J, Cragnaz, R. Congenital dermal sinuses, dermoid and epidermoid cysts of the posterior fossa. Child's Nerv Syst 1986; 2: 83-89.
8) Bourekas EC, Varakis K, Bruns D, Christoforidis GA, Baujan M, Slone HW, Kehagias D. Lesions of the corpus callosum: MR imaging and differential considerations in adults and children. AJR Am J Roentgenol 2002; 179: 251-257.
9) Singh S, Jain P, Yadav H, Singh I, Sen R. Corpus callosum dermoid cyst: A rare entity. Asian J Neurosurg 2017; 12: 296-297.
10) Dawes L, Yap J, Gaillard F. Intracranial dermoid cyst. Reference article, Radiopaedia.org (Accessed on 30 Jul 2023). Available at: https://doi.org/10.53347/rID-1213.
11) Gormley WB, Tomecek FJ, Qureshi N, Malik GM. Craniocerebral epidermoid and dermoid tumours: a review of 32 cases. Acta Neurochir (Wien) 1994; 128: 115-121.
12) Lunardi P, Missori P. Supratentorial dermoid cysts. J Neurosurg 1991; 75: 262-266.
13) Caldarelli M, Massimi L, Kondageski C, Di Rocco C. Intracranial midline dermoid and epidermoid cysts in children. J Neurosurg 2004; 100: 473-480.
14) Wallace D, Tress B, Kwan PF. Radiologically atypical congenital posterior fossa dermoid cyst presenting late in life. J Clin Neurosci 2008; 15: 835-838.
15) Shen WC, Chiou TL, Lin TY. Dermal sinus with dermoid cyst in the upper cervical spine: case note. Neuroradiology 2000; 42: 51-53.
16) Lee JK, Kim JH, Kim JS, Kim TS, Jung S, Kim SH, Kang SS, Lee JH. Cervical dermal sinus associated with dermoid cyst. Childs Nerv Syst 2001; 17: 491-493.
17) Brown JY, Morokoff AP, Mitchell PJ, Gonzales MF. Unusual imaging appearance of an intracranial dermoid cyst. AJNR Am J Neuroradiol 2001; 22: 1970-1972.
18) Drolshagen LF, Standefer M. Dense dermoid cyst of the posterior fossa. AJNR Am J Neuroradiol 1991; 12: 317.
19) Canbaz B, Kemerdere R, Ocal E, Tanriverdi T. Intracranial dermoid cyst mimicking a giant thrombosed aneurysm. Neurol India 2004; 52: 524-525.

20) Abbas S, Jaradat T, Boutarbouch M, Cherradi N, El Ouahabi A. Dermoid cyst, unusual location of the pterion: About a case and review of literature. Radiol Case Rep 2024; 19: 863-866.

21) Shalmiyev R, Devine A, Gonzalez S, Danckers M. Late-onset rupture of an intracranial dermoid cyst: a case report. J Med Case Rep 2024; 18: 9.

22) Mishra MK, Yadav V, Pradhan RS, Prasad RS. Intracranial interdural and extradural cerebellar convexity dermoid cyst: a case report of rare tumor at the rarest location. Asian J Neurosurg 2023; 18: 631-635.

23) Saifi I, Kashikar SV, Dhande R, Pavanan A, Reddy SNG. Magnetic resonance imaging findings in a rare case of a ruptured intracranial dermoid cyst. Cureus 2023; 15: e43316.

24) Emmanuel S, Inban P, Akuma O, Nouman Aslam M, Talat F, Nizamani A, Chenna VSH, Romain EM, Chu Carredo CK, Khan A. An atypical case of intracranial dermoid cyst in an adult female: a case report and literature review. Cureus 2023; 15: e39807.

25) Tam AKY, Zhu XL, Chow SMW, Lee SKJ, Lai CCF, Chan KY, Chu WCW, Tong MCF. Intracranial nasofrontal dermoid cyst and a hair in the nose tip without a sinus tract identified with single-staged combined approach excision done. Clin Case Rep 2023; 11: e7414.

26) Bahakeem B, Alqahtani TF, Darwish A, Alkhotani A, Rawah E. Dermoid cyst in the posterior fossa associated with dermal sinus and abscess formation: a case report. Cureus 2023; 15: e38032.

27) Aktham A, Morita S, Takeuchi S, Ismail M, Hoz SS, Numazawa S, Watanabe S, Mori K. Traumatic rupture of intracranial dermoid cyst with continuous fat droplet migration. Surg Neurol Int 2023; 14: 39.

28) Taha A, Abdelrazek MA, Manalo N, Elsadek R, Morrin SJ, Brodski A, Augustynowiczd A, Mollashahi RS, Shenoy A. Traumatic rupture of a skull base dermoid cyst mimicking chronic meningitis. Cureus 2022; 14: e25066.

29) Delgado-Muñoz P, Oliver-Ricart MS, Labat-Alvarez EJ. A rare case of an intracranial dermoid cyst with atypical appearance on computed tomography and magnetic resonance imaging. Am J Case Rep 2022; 23: e935115.

30) Fornari M, Solero CL, Lasio G, Lodrini S, Balestrini MR, Cimino C, Visintini S, Pluchino F. Surgical treatment of intracranial dermoid and epidermoid cysts in children. Childs Nerv Syst 1990; 6: 66-70.

31) Orakcioglu B, Halatsch M-E, Fortunati M, Unterberg A, Yonekawa Y. Intracranial dermoid cysts: variations of radiological and clinical features. Acta Neurochir (Wien) 2008; 150: 1227-1234.