



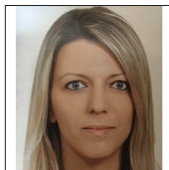
## Case Report

# Acute-onset diplopia from intracranial hypertension due to torcular herophili obstruction by an hemorrhagic intradiploic epidermoid cyst

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## ABSTRACT

**Background:** Epidermoid cysts are benign slow-growing congenital lesions, constituting approximately 1% of all cranial tumors. Most of these lesions are located intradurally, while about 10–25% of them are located within the diploic spaces. Intradiploic epidermoid cysts are usually discovered incidentally and may remain asymptomatic for many years, but in rare instances, they may grow intracranially and produce brain compression. Sometimes, intradiploic epidermoid cysts may occlude the main cranial venous sinuses causing intracranial hypertension.

**Case Description:** We present the case of a 24-year-old male harboring a paramedian right occipital intradiploic cyst with erosion of both outer and inner bony tables, which occluded the torcular herophili producing a worsening symptomatology with acute-onset diplopia from right sixth cranial nerve palsy; the patient also presented bilateral papilledema, but only reported mild headache and dizziness. Neuroradiological studies evidenced a lesion compatible with intradiploic epidermoid cyst with intralesional hemorrhagic component, overlying and almost completely occluding the torcular herophili. Considering the fast worsening of symptomatology and the evidence of intracranial hypertension, the patient was operated on immediately after completion of clinical and radiological assessment. The lesion was radically removed with almost immediate reversal of signs and symptoms. Histopathology confirmed the diagnosis of epidermoid cyst with intralesional hemorrhagic components.

**Conclusion:** Intradiploic epidermoid cysts may cause intracranial hypertension by occlusion of main cranial venous sinuses; intralesional hemorrhage may act as precipitating factor in occlusion of the torcular herophili, producing rapidly worsening intracranial hypertension, which requires prompt surgical treatment to reverse symptomatology. Radical surgical resection is necessary to avoid recurrence.

**Keywords:** Epidermoid cyst, Intracranial hypertension, Intradiploic, Sixth cranial nerve palsy, Torcular herophili

## INTRODUCTION

Intracranial epidermoid cysts are congenital nontumoral lesions, arising from aberrated ectodermal remnants that develop into epithelium-like cells during neural tube closure between the 3<sup>rd</sup> and 5<sup>th</sup> weeks of fetal development. Even though congenital, these lesions have an extremely slow

linear growth rate, which make them become symptomatic late in adult population, most frequently between 20 and 40 years of age. Epidermoid cysts account for approximately 1% of all intracranial space-occupying lesions.<sup>[1,3,8,17,30,31]</sup> The vast majority of these lesions are intradurally located and only 10–25% of them develop extradurally within the cranial diploic spaces.<sup>[3-5,9,20]</sup> Intradiploic epidermoid cysts may remain asymptomatic for many years and are often occasionally discovered on plain skull X-rays, computed tomography (CT) scan, or magnetic resonance imaging (MRI) performed for trauma or other reasons, but they may erode both the inner and the outer bony tables and grow intracranially producing brain compression with consequent neurological deficit and/or intracranial hypertension;<sup>[11,13,16,18,24,33]</sup> intradiploic epidermoid cysts may rarely occlude major venous cranial sinuses, causing secondary intracranial hypertension.<sup>[22,23,28,29,32,36]</sup>

We present the case of a 24-year-old male harboring a paramedian right occipital intradiploic epidermoid cyst, with erosion of both outer and inner bony tables and evidence of intralesional hemorrhage, which occluded the torcular herophili producing a rapidly worsening intracranial hypertension with acute-onset diplopia due to the right sixth cranial nerve palsy; the prompt resection of the lesion allowed immediate flow restoration in the confluence of sinuses, with reversal of symptomatology and recovery from sixth cranial nerve palsy.

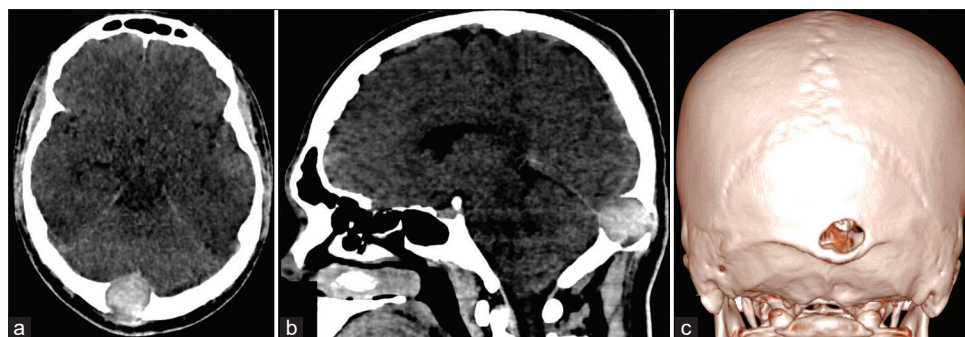
## CASE REPORT

A 24-year-old male presented with a 3-week history of intensifying morning headache, progressively worsening gait, unsteadiness, and blurred vision; he also reported increasing frequent episodes of transient loss of vision for a few seconds each time he stood up quickly, in the week before admission; on the day of admission, he presented acute-onset binocular diplopia.

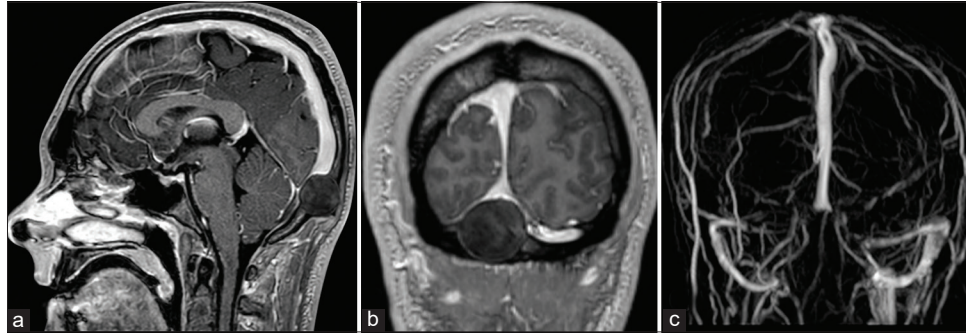
Examination revealed horizontal binocular diplopia; the remainder of the cranial nerve examination findings were

unremarkable; motor strength, sensation, and reflexes resulted normal, while minimal gait unsteadiness was noticed. A nontender and not painful lump, of which the patient was not aware, was palpable at the level of inion. The patient was referred to an ophthalmologist, who performed fundoscopy, which showed gross (Grade III Frisen scale) papilledema, and Hess screen chart, confirming right sixth cranial nerve palsy. A noncontrast-enhanced CT scan demonstrated a paramedian right occipital well-circumscribed, inhomogeneously hyperdense, lesion with sharply demarcated erosion of both tables of the skull and scalloping of sclerotic bony margins [Figures 1a and b]; the 3D CT reconstruction clearly showed the bone defect [Figure 1c]. MRI confirmed the presence of the intradiploic, almost spherical, lesion with a diameter of about 2.6 cm; on T1-weighted images, the lesion appeared hypointense to the brain and slightly hyperintense to the CSF; on T2-weighted images, it appeared inhomogeneously hyperintense to the brain and almost isointense to CSF; diffusion-weighted images were hyperintense with a high value on apparent diffusion coefficient; contrast-enhanced T1-weighted images revealed almost complete occlusion of the venous flow at the confluence of sinuses (torcular herophili) by the lesion itself, which resulted nonenhanced [Figures 2a and b]; MRI venogram confirmed the occlusion of the torcular [Figure 2c]. Neuroimaging was compatible with diagnosis of intradiploic epidermoid cyst with intralesional hemorrhage.

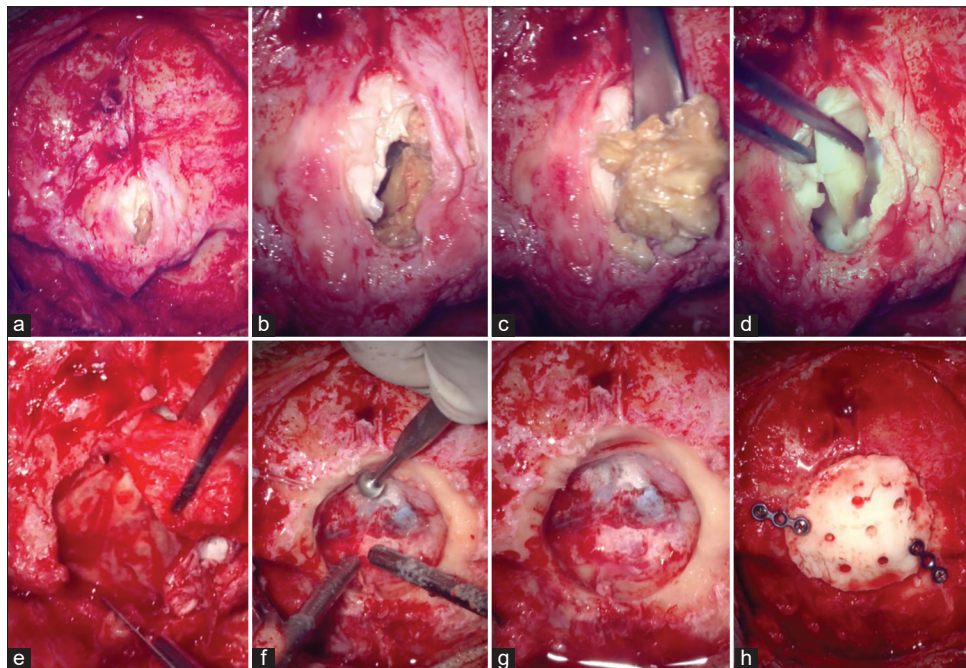
Considering the fast worsening of symptomatology, the presence of evident papilledema and the acute onset of diplopia from the right sixth cranial nerve palsy, all compatible with intracranial hypertension, a rapid restoration of flow in the main venous sinuses was reputed necessary and the patient was operated on immediately after completion of clinical and radiological assessment. A longitudinal median parieto-occipital incision of 8 cm, centered on the inion, allowed skeletonization of the region and exposure of the lesion, which bulged from the eroded outer bone table, harvested by a fibrotic capsule;



**Figure 1:** Preoperative nonenhanced computed tomography (CT) scan demonstrating a paramedian right occipital well-circumscribed, inhomogeneously hyperdense, lesion with sharply demarcated erosion of both tables of the skull in axial (a) and sagittal view (b); preoperative 3D CT scan showing the bone defect (c).



**Figure 2:** Preoperative contrast-enhanced T1-weighted magnetic resonance imaging in sagittal (a) and coronal (b) view demonstrating the hypointense lesion obstructing the confluence of the superior longitudinal and transverse sinuses, confirmed by MRV (c).



**Figure 3:** Intraoperative images: the lesion, exposed through a longitudinal median parieto-occipital incision and skeletonization, bulged from the eroded outer bone table, harvested by a fibrous capsule (a); under microscopic magnification, the capsule was incised evidentiating the cyst content, which had a variegated appearance with some dark areas suggestive for previous subacute intralesional hemorrhage and other areas with the classical pearly aspect of epidermoid cysts (b); the lesion was removed piecemeal (c and d); after evacuation of the cystic contents, the fibrous capsule was separated from the borders of the bone and dissected from the intact dura mater (e); the sclerotic and thickened bony borders were drilled away, highlighting blood flow restoration in the confluence of sinuses (f and g); the bony defect was reconstructed by acrylic resin cranioplasty (h).

incision of the capsule gave rise to a small amount of brownish fluid (collected for analysis) exposing the cyst content, which had a variegated appearance with some dark areas suggestive for previous subacute intralesional hemorrhage and other areas with the classical pearly aspect of epidermoid cysts; under microscopic vision, the lesion was removed piecemeal; the fibrotic capsule was separated from the sclerotic borders of the bone and dissected away from the intact dura mater, with evident blood flow restoration in the confluence of sinuses; thereafter, the sclerotic scalloped bone margins were drilled to reduce

their thickness, for a better reconstruction, which was performed by acrylic resin cranioplasty [Figure 3a-h].

Postoperative course was uneventful; the patient was mobilized on the 1<sup>st</sup> postoperative day and discharged on the 4<sup>th</sup> postoperative day; diplopia progressively improved, and at a 30 days follow-up, the sixth cranial nerve palsy and the papilledema had completely resolved. Postoperative CT scan with 3D reconstruction and MRI with MR venogram, performed during hospitalization, demonstrated the radical exeresis of the lesion, the optimal result of cranioplasty, and

the restoration of flow in the torcular herophili and transverse sinuses [Figures 4 and 5].

Histology of specimen revealed features consistent with a cystic formation with a fibrous wall, outlined by squamous epithelium and containing keratinic material arranged in lamellae, consistent with the diagnosis of epidermoid cyst [Figure 6]. Analysis of the cystic fluid revealed an exudate with erythrocytes, leukocytes, and epithelial cells.

## DISCUSSION

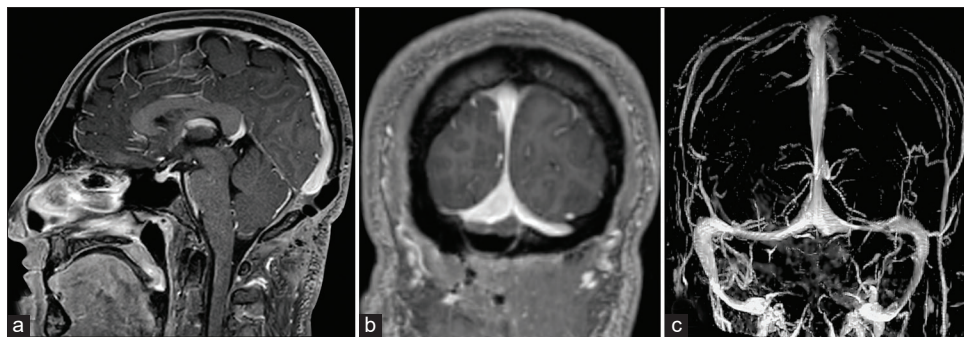
Epidermoid cysts are nontumoral lesions accounting for approximately 1% of all intracranial expansive lesions, originating from ectodermal cell remnants enclosed into the neural tube during the embryonic period, between the 3<sup>rd</sup> and 5<sup>th</sup> weeks of fetal development. These lesions have an extremely low growth rate and thereafter, in most instances, become symptomatic late in adult life, more often between the twentieth and fortieth decade of life.<sup>[1,3,4,8,17,30,31]</sup> Most of these lesions are intradurally located, but 10–25% are located extradurally into the diploic spaces of the skull bones.<sup>[3-5,9,20]</sup> Intradiploic epidermoid cysts may represent an occasional finding during neuroradiological imaging but also may grow eroding both the inner and the outer table of the skull; these lesions may sometimes grow both extracranially and/or intracranially, reaching a giant size.<sup>[11,13,16,19,20,27,35]</sup>

Erosion of the outer table may produce a palpable, usually painless, lump, which may require surgical resection, not only for diagnostic confirmation but also for cosmetic reasons.<sup>[4,16,19,30]</sup> Intradiploic epidermoid cysts may grow intracranially producing brain compression with consequent neurological deficit and/or intracranial hypertension.<sup>[11,13,16,18,24,33]</sup> Intradiploic epidermoids may rarely occlude major venous cranial sinuses, causing secondary intracranial hypertension,<sup>[22,23,28,29,32,36]</sup> in effect, according to the Monro–Kellie doctrine, disturbances in cerebral venous outflow, may result in venous congestion and consequent development of signs and symptoms of increased intracranial pressure.<sup>[34]</sup>

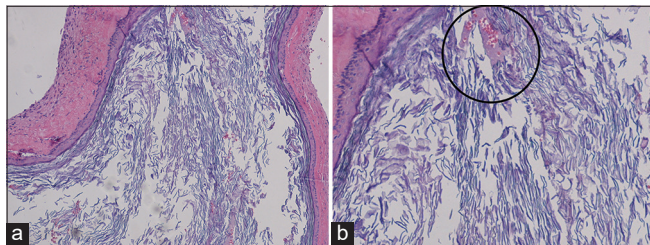
Intradiploic epidermoid cysts typically appear on CT scans as hypodense, nonenhancing lesions, with sharply demarcated bony sclerotic, often scalloped, margins; on MRI, these lesions usually appear slightly hyperintense to the CSF in T1-weighted images and isointense/hyperintense to the CSF in T2-weighted images, are markedly hyperintense on flair and on DWI present characteristic hyperintensity with a high value on apparent diffusion coefficient; on T1-weighted images with paramagnetic contrast administration, epidermoid cysts appear not enhancing.<sup>[3,6,8,10,17,30,31]</sup> In rare instances, intradiploic epidermoid cyst contents are hyperdense on CT scans which may be the consequence of inflammatory or degenerative changes or, more frequently,



**Figure 4:** Postoperative CT scan showing complete removal of the lesion and reconstruction of the bone defect by acrylic resin material cranioplasty in axial (a) and sagittal view (b) and in 3D CT scan (c).



**Figure 5:** Postoperative contrast-enhanced T1-weighted MRI in sagittal (a) and coronal (b) view, demonstrating the complete exeresis of the lesion with resolution of venous sinuses obstruction, as confirmed by MRV (c).



**Figure 6:** Photomicrograph of the specimen shows a cystic formation with a fibrous wall, outlined by squamous epithelium and containing keratinic material arranged in lamellae (a: HE,  $\times 100$ ); foreign body granulations with moderate vascularity and intralesional hemorrhage can be observed at higher magnification (b: HE,  $\times 200$ ; foreign body granulation in the circle).

as in the present case, of intralesional hemorrhage.<sup>[2,25,26,35]</sup> Magnetic resonance venography is the gold standard examination to evaluate flow in cases of epidermoid cysts overlying the torcular or other major sinuses.<sup>[22,23,36]</sup>

Histopathology typically shows a cystic lesion with a fibrous wall containing stratified squamous epithelium and keratinic material arranged in lamellae;<sup>[9,21,33]</sup> it is noteworthy that intracranial intradural epidermoid cysts are lacking fibrous wall and are usually simply outlined by a germinative membrane formed by stratified squamous epithelium; this peculiarity, not reported in the current literature, could be the expression of foreign body reaction by the mesenchymal bony structures, absent in case of intradural involvement of the brain. On the other hand, both in case of intracerebral or in case of intradiploic epidermoid cysts, intralesional hemorrhage has been described; due to the avascular nature of epidermoid cysts, intralesional hemorrhage is rare and only a limited number of cases have been reported. The most accredited hypothesis is that the irritant cyst content leakage may result in foreign body granulation formation not only in the cyst wall but also inside the cyst; foreign body granulations behave mild vascularity, with capillary sized vessels, whose rupture is responsible for the intracystic hemorrhage.<sup>[7,12,14,15]</sup>

The case we present is not unusual in its presentation because produced intracranial hypertension by obstructing the torcular herophili, but because presented an acute-onset diplopia from the sixth nerve palsy and rapidly evolving signs (marked papilledema) and symptoms of intracranial hypertension; in our opinion, this is due to intralesional hemorrhage, which led to a fast and almost complete occlusion the confluence of venous sinuses, presumably only partially compressed before the hemorrhage. The atypical hyperdense aspect of the lesion in preoperative CT scans was actually consequent to the intralesional hemorrhage, as intraoperatively demonstrated.

Resection of the lesion, performed shortly after the symptomatologic onset, allowed prompt and complete

resolution of symptoms and signs of intracranial hypertension, comprising papilledema and right sixth nerve palsy, with recovery of visual acuity and ocular motricity. The use of cautious and precise dissection under microscopic vision allowed preservation of the venous sinuses with immediate restoration of flow.

## CONCLUSION

Intradiploic epidermoid overlying the torcular herophili may compress the confluence of venous sinuses producing intracranial hypertension. Complete neuroimaging study (CT scan; MRI; and MR venogram) allow precise diagnosis and evaluation of venous sinus flow. Intralesional hemorrhage may acutely occlude the venous flow aggravating the symptomatology. Evidence of hyperdensity on CT scan images, actual expression of intralesional hemorrhage, and acute onset of signs and symptoms of hypertension indicate the need for immediate surgical decompression. Resection under microscopic vision and skilled technique warrants for radical removal with preservation of sinuses.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

## REFERENCES

1. Akar Z, Tanriover N, Tuzgen S, Kafadar AM, Kuday C. Surgical treatment of intracranial epidermoid tumors. *Neurol Med Chir (Tokyo)* 2003;43:275-80.
2. Ambo M, Kumakiri M, Chohno Y. Epidermoid cyst with perforation of the skull. *J Am Acad Dermatol* 2003;49:934-6.
3. Arana E, Latorre FF, Revert A, Menor F, Riesgo P, Liano F, *et al.* Intradiploic epidermoid cysts. *Neuroradiology* 1996;38:306-11.
4. Arko L 4<sup>th</sup>, Berry CT, Desai AS, Weaver M. Intradiploic Epidermoid tumors of the cranium: Case report with review of the literature. *J Neurol Surg A Cent Eur Neurosurg* 2017;78:167-79.
5. Bikmaz K, Cosar M, Bek S, Gokduman CA, Arslan M, Iplikcioglu AC. Intradiploic epidermoid cysts of the skull: A report of four cases. *Clin Neurol Neurosurg* 2005;107:262-7.
6. Bulja D, Didgar M, Depreitere B, Wilms G. Intradiploic epidermoid cyst: Report of two cases with typical CT and MR imaging features. *Eur J Radiol Extra* 2011;77:e23-7.
7. Chen CY, Wong JS, Hsieh SC, Chu JS, Chan WP. Intracranial epidermoid cyst with hemorrhage: MR imaging findings.

- AJNR Am J Neuroradiol 2006;27:427-9.
8. Chowdhury FH, Haque MR, Sarker MH. Intracranial epidermoid tumor; microneurosurgical management: An experience of 23 cases. *Asian J Neurosurg* 2013;8:21-8.
  9. Ciappetta P, Artico M, Salvati M, Raco A, Gagliardi FM. Intradiploic epidermoid cysts of the skull: Report of 10 cases and review of the literature. *Acta Neurochir (Wien)* 1990;102:33-7.
  10. Clark MP, Pretorius PM, Beaumont D, Milford CA. Congenital cholesteatoma of occipital bone or intradiploic epidermoid cyst? One and the same disease. *J Laryngol Otol* 2009;123:673-5.
  11. Duan ZX, Chu SH, Ma YB, Zhang H, Zhu JL. Giant intradiploic epidermoid cyst of the occipital bone. *J Clin Neurosci* 2009;16:1478-80.
  12. Fox BD, Smitherman SM, Amhaz H, Ruiz MI, Rouah E, Radaideh M, *et al.* A supratentorial, hemorrhagic, intraparenchymal epidermoid cyst. *J Clin Neurosci* 2009;16:1101-5.
  13. Gollapudi PR, Musali SR, Mohammed I, Pittala SR. A frontal giant intradiploic giant pearl (epidermoid cyst) with intracranial and extracranial extension: A rare entity. *J Pediatr Neurosci* 2018;13:480-2.
  14. Hsieh CH, Huang KM, Kao MC, Peng S, Wang CC. Hemorrhage in intracranial epidermoid cyst. *J Formos Med Assoc* 1996;95:173-5.
  15. Inoue Y, Ohata K, Nakayama K, Haba T, Shakudo M. An unusual middle fossa interdural epidermoid tumor. Case report. *J Neurosurg* 2001;95:902-4.
  16. Jaiswal AK, Mahapatra AK. Giant intradiploic epidermoid cysts of the skull. A report of eight cases. *Br J Neurosurg* 2000;14:225-8.
  17. Kato K, Ujiie H, Higa T, Hayashi M, Kubo O, Okada Y, *et al.* Clinical presentation of intracranial epidermoids: A surgical series of 20 initial and four recurred cases. *Asian J Neurosurg* 2010;5:32-40.
  18. Khan AN, Khalid S, Enam SA. Intradiploic epidermoid cyst overlying the torcula: A surgical challenge. *BMJ Case Rep* 2011;2011:bcr0820103245.
  19. Krupp W, Heckert A, Holland H, Meixensberger J, Fritzsche D. Giant intradiploic epidermoid cyst with large osteolytic lesions of the skull: A case report. *J Med Case Rep* 2012;6:85.
  20. Kumaran SP, Gupta K, Priyadarshini H, Ghosal N. Giant intradiploic epidermoid cyst of occipital bone. A case report and literature review. *Neuroradiol J* 2010;23:707-10.
  21. Lakshmi V, Thirumurthy KS, Rabia S, Chaitra W. Intradiploic epidermoid cyst-a pearl within the skull. *J Diagn Pathol* 2014;9:38-42.
  22. Lam C, Solomon R, Clark H, Casey S. Reversal of increased intracranial pressure with removal of a torcular epidermoid: Case report. *Neurosurgery* 2001;48:929-32.
  23. Lee G, Seex K, Scott G. Pseudotumour cerebri due to a torcular epidermoid cyst. *ANZ J Surg* 2001;7:385-8.
  24. Moreira-Holguin J, Medélez-Borbonio R, Quintero-Lopez E, García-González U, Gómez-Amador J. Intradiploic epidermoid cyst with intracranial hypertension syndrome: Report of two cases and literature review. *Int J Surg Case Rep* 2015;16:81-6.
  25. Nambu A, Imanishi Y, Iwasaki Y, Fujikawa M, Hayashi T, Shinagawa T, *et al.* Intradiploic epidermoid cyst with focal internal enhancement. *Radiat Med* 2006;24:224-7.
  26. Ng WH, Mitchell P, Tickell L, Kaye AH. A case of hyperdense diploic epidermoid cyst on CT scan mimicking meningioma. *J Clin Neurosci* 2004;11:930-2.
  27. Oommen A, Govindan J, Peroor DS, Azeez CR, Rashmi R, Jalal MJ. Giant occipital intradiploic epidermoid cyst. *Asian J Neurosurg* 2018;13:514-7.
  28. Park BH, Lee HS, Lee JS. Occipital intradiploic epidermoid cyst with intracranial hypertension. *J Korean Neurosurg Soc* 2006;40:377-80.
  29. Powers JM, Schnur JA, Baldree ME. Pseudotumor cerebri due to partial obstruction of the sigmoid sinus by a cholesteatoma. *Arch Neurol* 1986;43:519-21.
  30. Reddy MP, Jiacheng S, Xunning H, Zhanlong M. Intracranial epidermoid cyst: Characteristics, appearance, diagnosis, treatment and prognosis. *Sci Lett* 2015;3:102-10.
  31. Rehman L, Bokhari I, Siddiqi SU, Bagga V, Hussain MM. Intracranial epidermoid lesions: Our experience of 38 cases. *Turk Neurosurg* 2018;28:763-7.
  32. Rengachary S, Kishore PR, Watanabe I. Intradiploic epidermoid cyst of the occipital bone with torcular obstruction. Case report. *J Neurosurg* 1978;48:475-8.
  33. Wang B, Zhang F, Cheng H, Hong W. Study of intradiploic epidermoid cyst in the right occipital bone. *Int J Clin Exp Med* 2019;12:9483-6.
  34. Wilson MH. Monro-Kellie 2.0: The dynamic vascular and venous pathophysiological components of intracranial pressure. *J Cereb Blood Flow Metab* 2016;36:1338-50.
  35. Xiao G. Giant hyperdense intradiploic epidermoid cyst of the skull with extra and intracranial extension: Report of two cases and literatures review. *Chin J Cancer Res* 2005;17:154-6.
  36. Yokota H, Noguchi H, Yokoyama K. Epidermoid cyst with torcular herophili obstruction and unusual venous drainage. *World Neurosurg* 2019;128:14-7.

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