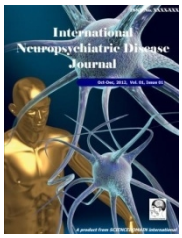

International Neuropsychiatric Disease Journal
2(2): 68-77, 2014

SCIENCEDOMAIN *international*
www.sciencedomain.org

A Rare Case of Third Ventricular Epidermoid Cyst as Seen by Newer MRI Sequences

Ralph Xiu-Gee Man[1], Avinash Kumar Kanodia[2*], Richard Stephen Nicholas[3] and Gavin Main[2]

[1]Elderly medicine (G level) Southampton general hospital, UK.
[2]X-ray Department, Ninewells Hospital, Dundee, UK.
[3]Medical Physics, Ninewells Hospital, Dundee, UK.

Authors' contributions

This work was carried out in collaboration between all authors. Authors RXGM and AKK wrote the manuscript. Author AKK performed the literature search with inputs from authors RSN and GM. All authors read and approved the final manuscript.

Case study

Received 20th September 2013
Accepted 22nd November 2013
Published 13th December 2013

ABSTRACT

Aim: We describe a 52 year old gentleman who had a third ventricular lesion, presumed to be epidermoid cyst, which is very rare. We also describe the appearances of this rare condition on newer MRI sequences.
Presentation of Case: Third ventricle is a rare site of epidermoid cysts and difficult to visualise specially in routine MRI sequences. We used 3D FLAIR sequence as part of routine MRI protocol that was helpful in raising suspicion initially, aided by clinical presentation. Subsequent 3D T2 SPACE sequence confirmed the presence of abnormality.
Discussion: The case highlights the importance of high resolution 3D FLAIR sequence as part of initial MRI protocol that is devoid of CSF flow artefacts and therefore helpful for small intraventricular lesions. Importance of 3D T2 SPACE is also highlighted which is another new MRI sequence. It has excellent spatial resolution without artefacts that is helpful in clearly delineating the presence of small lesions such as epidermoid cyst and its relationship with surrounding structures and increasing diagnostic certainty.
Conclusion: We present a rare case of presumed third ventricular epidermoid cyst and potential role of newer MRI sequences in small intraventricular lesions.

**Corresponding author: Email: ak_kanodia@rediffmail.com;*

Keywords: Third ventricle; epidermoid; 3D FLAIR; SPACE.

1. INTRODUCTION

Intracranial epidermoid cysts (ECs) are rare slow growing congenital lesions, accounting for 0.2% to 1.8% of all intracranial primary tumours [1]. In the literature, 11 cases of ECs have been reported within the region of the third ventricle [2-13]. We present a rare case of third ventricular EC as seen on newer magnetic resonance imaging (MRI) sequences.

2. PRESENTATION OF CASE

A 52 year old gentleman was referred to ophthalmology outpatients by his optician following an incidental finding of bilateral inferior quadrantanopia. There was no history of trauma or significant family history. On examination, visual acuity was normal and no other focal neurological defects were identified. MRI was requested to rule out organic causes of his visual field defect.

The initial unsupervised MRI of the brain showed no obvious abnormality in T1 sagittal and T2 axial images (Fig. 1). However, the three-dimensional Fluid Attenuated Inversion Recovery (3D FLAIR) sequence, performed routinely in almost all patients in our institution, showed an area of heterogenous signal in the suprasellar region within the third ventricle (Fig. 2). This prompted a recall for a repeat supervised MRI using high-resolution 3D Sampling perfection with application optimized contrast using flip angle evolution with strong T2 weighting (3D T2 SPACE), that we use for assessment for Internal acoustic meatus (IAM) and inner ear (Fig. 3), diffusion weighted imaging (DWI) (Fig. 4) and T1-weighted sequences before and after gadolinium enhancement (Fig. 5).

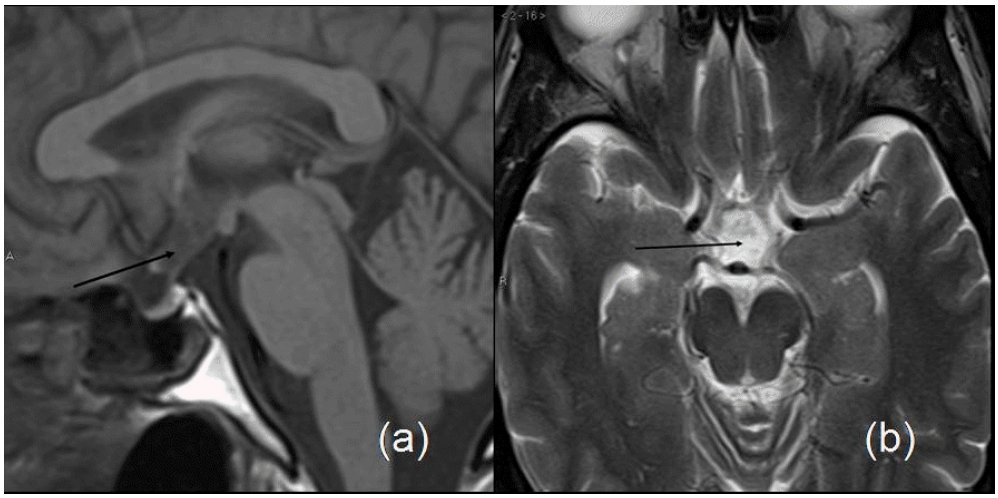

Fig. 1. (a) T1 weighted sagittal and (b) T2 weighted axial images show the position of epidermoid cyst (black arrow) as an area of low signal on T1 and high signal on T2 weighted images.

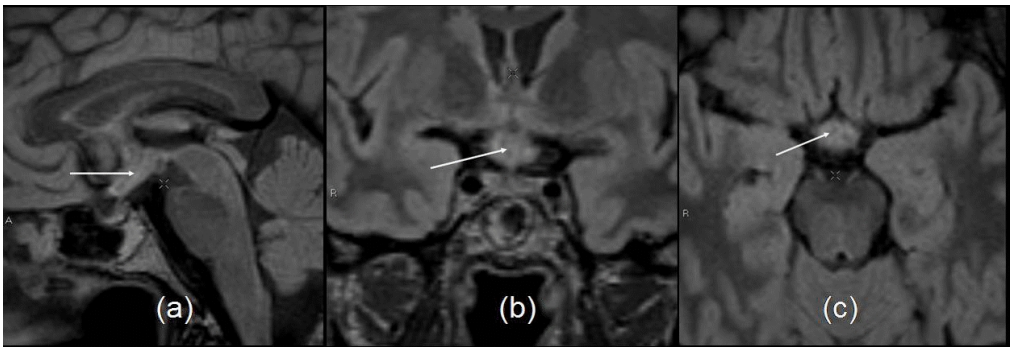

Fig. 2. 3D FLAIR images in (a) sagittal, (b) coronal and (c) axial reconstructions, showing epidemoid cyst (white arrow) as an area of intermediate signal, within the third ventricle.

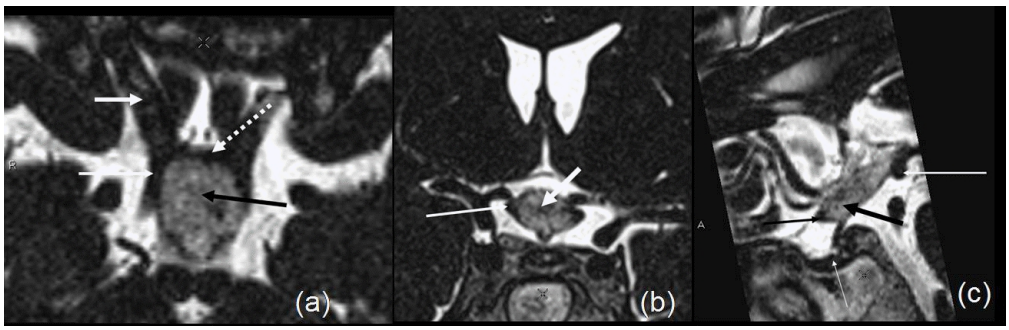

Fig. 3. 3D T2 SPACE sequence (a) Axial reconstruction showing right optic nerve (thick white arrow), optic tract (thin white arrow), optic chiasm (dotted white arrow) and epidermoid cyst (black arrow). (b) Coronal reconstruction showing optic tract (thin white arrow) and epidermoid cyst (thick white arrow). (c) Sagittal reconstruction showing epidermoid cyst (thick black arrow), optic chiasm (thin black arrow), mamillary body (long white arrow) and pituitary gland (small white arrow).

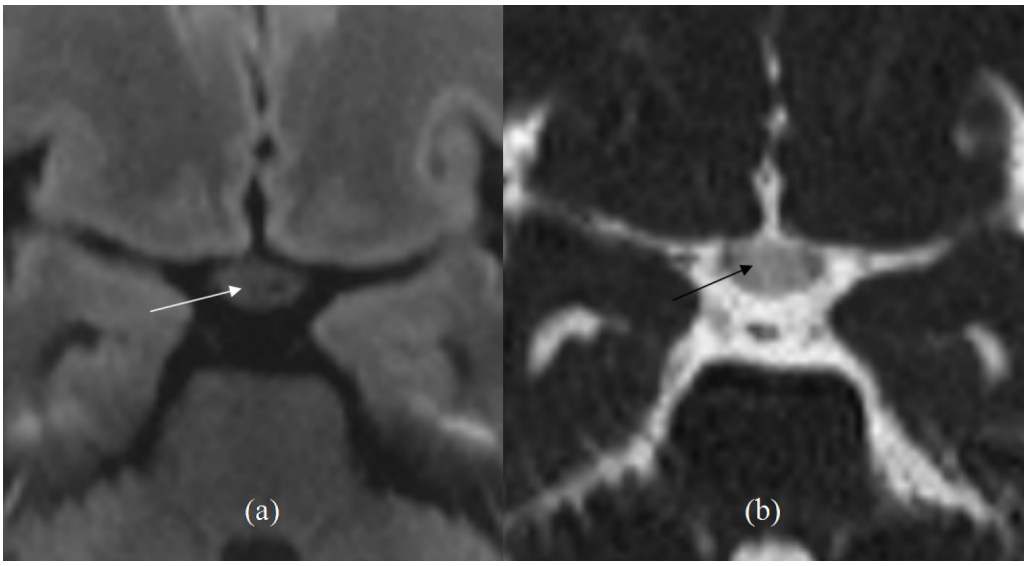

Fig. 4. Diffusion-weighted Imaging (a) Axial DWI (b=1000s/mm^2) and (b) ADC map showing the epidermoid cyst (arrows). The DWI signal is higher than CSF and similar to white matter. On ADC the signal is intermediate between CSF and white matter.

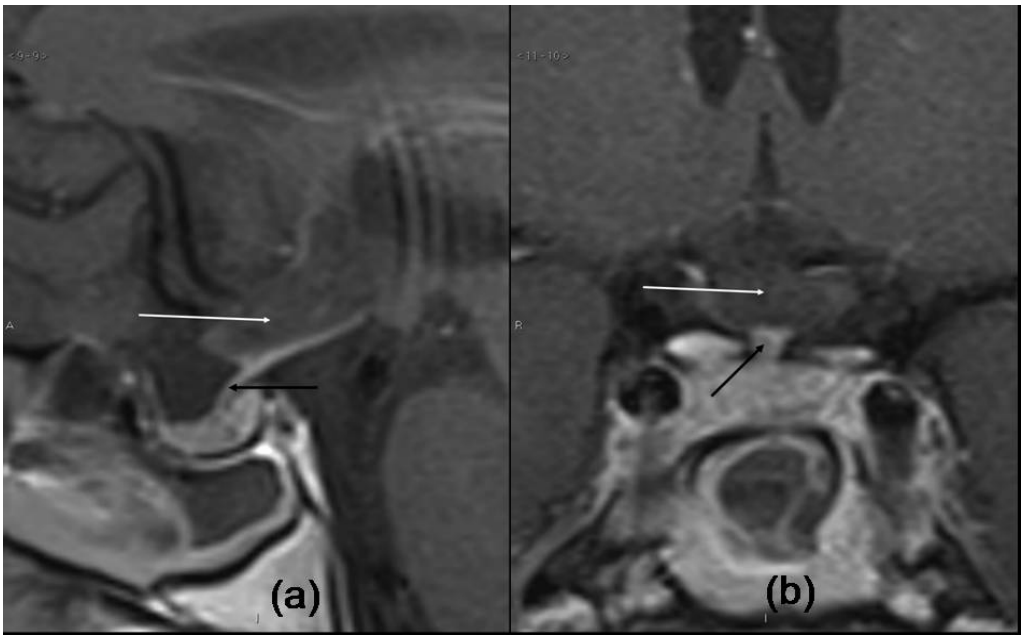

Fig. 5. (a) Sagittal and (b) Coronal T1-post contrast images showing the epidermoid cyst (white arrow) and enhancing pituitary stalk (black arrow).

On 3D T2 SPACE sequence (Fig. 3), a mass measuring 14 x 20 x 7 mm transversely, anterior-posteriorly and supero-inferiorly respectively was demonstrated to be present at the floor of third ventricle mainly on the anterior/antero-inferior aspect including supraoptic

recess, abutting the supero-medial aspect of the optic tracts and optic chiasm but reaching posteriorly upto mamillary bodies. The lesion took the shape of the third ventricle and had no significant mass effect. It had heterogeneously low signal on T1 weighted images and high signal on T2 weighted images. On 3D T2 SPACE, the lesion was hypointense compare to adjacent CSF but significantly brighter than the surrounding cerebral parenchyma. The lesion did not enhance with contrast. On DWI sequence, on the strong diffusion-weighted (b-value = 1000 s/mm^2) images, the lesion was of higher signal than CSF and similar to the white matter signal. On apparent diffusion coefficient (ADC) map, the lesion had intermediate signal between CSF and white matter.

The findings were thought to represent an unusual intra-ventricular EC at the floor of third ventricle, including the supraoptic recess, abutting the optic tracts and chiasm. On 2 month follow up MRI, the lesion showed no interval change. Following multi speciality review, it was decided to keep the patient under further follow up and currently, the patient is being closely monitored in an outpatient setting.

3. DISCUSSION

ECs arise from dermal inclusion during neural tube closure in the third to fifth week of embryogenesis. Islands of ectodermal tissue arise from failure of total separation from the neurotube. Other theories have suggested that invagination or abnormal sequestration of ectoderm from dermal fusion sites such as the eyes and ears [3,12,14,15]. Acquired ECs have been thought to be caused by surgical or post traumatic implantation [16].

Histologically, ECs resembles the linear growth rate of skin [17]. They are composed of a thin of squamous epithelium lining which macroscopically appear white, pearly, smooth, nodular or lobulated [16,18]. There is an internal core of desquamated epithelial keratin and high content of cholesterol giving the distinctive characteristic MR imaging appearance in different radiological modalities.

Intracranial ECs tend to be located off the midline. The most common location, 40-50% of these are within the region of cerebellopontine angle (CPA) [19]. ECs are the third most common tumour in the CPA region, following vestibular schwannoma and meningioma. Approximately 100 cases of ECs have been reported in the fourth ventricle (17%) and the parasellar and sellar region (10-15%) [18,20]. Less common location include intraparenchymal, including all lobes of the cerebral hemisphere, brainstem and spine [16]. Third ventricular ECs are very rare. After careful search, we could find only 11 cases of ECs within the third ventricle in the literature [2-13].

ECs are known to be ‘soft’ tumours due to their content and tend to take the shape of available space, rather than compressing adjacent structure significantly. Masses related to anterior recesses or the floor of the third ventricle tends to manifest with dysfunction of the hypothalamic-pituitary axis or visual disturbance due to impingement of optic chiasm or optic recesses [21].

In the literature, patients with ECs commonly present with signs and symptoms of gradual onset mass effect. Symptoms such as headache, cranial nerve defects, cerebellar signs, seizures and raised intracranial pressure [1,3,4,6,7,13,15,22-24].

Masses arising from floor of the third ventricular are uncommon. The most frequently seen lesion is hypothalamic hamartoma in young adults. Distortion of third ventricular floor by

extrinsic pathology such as ectasia or basilar artery aneurysm, dermoid, epidermoid or arachoid cysts have also been reported [21]. The differential diagnosis of masses in relation to anterior aspect of the third ventricle in adults is broad, including lymphoma, pituitary macroadenoma, craniopharyngioma, metastases, granulomatous disease, meningioma, giant cell astrocytoma and cystic lesions (such as colloid cysts).

Depending on the anatomical location of the EC within the third ventricle, different manifestations have been described in the literature. These include nausea, vomiting, headache, visual disturbance and mental disturbance [2-8]. Tumours in the anterior and inferior region of the third ventricle can cause endocrine disturbance and visual impairment. In the case of our patient, blood test showed no sign of hypopituitarism or diabetes insipidus. A single case report of an EC of posterior third ventricle presented solely with galactorrhea [11]. Significant complications could occur, such as hydrocephaly following obstruction of foramen of Monro. Rupture and release of intracystic content into cerebral spinal fluid have been reported to cause aseptic meningitis. Post-operative recurrence has been reported [5,9].

On CT imaging, ECs typically presents as well defined, round, lobulated hypoattenuated masses with a density similar to CSF, however, uniformly hyperdense epidermoid have been reported on CTs, which is likely due to high protein content or calcium constitution following saponification in 10% of intracranial ECs [3,25].

On MRI, ECs usually appear hypointense on T1 and hyperintense on T2 weighted imaging. They demonstrate some degree of internal heterogeneity and minimal or no enhancement after gadolinium administration [19].

On FLAIR sequences, the majority of ECs appear hyperintense relative to CSF [26]. Contrast variations within ECs have been shown in FLAIR sequences, due to differences in water content. It is suggested that signals from EC with high water content are suppressed by FLAIR giving intermediate signal intensity in some cases. 2D FLAIR is most commonly used; however, the 3D FLAIR is a relatively new sequence that is gaining acceptance for neuroimaging in multiple sclerosis and other situations. It is based around a SPACE acquisition with near-isotropic resolution allowing multi-planar reformatting of the resulting images. 3D FLAIR suppresses CSF signal over the whole brain volume and is therefore less susceptible to CSF flow artefacts than 2D FLAIR. This makes 3D FLAIR ideal to detect subtle pathology within the ventricular cavities. To the best of our knowledge, an example of an EC on 3D FLAIR has not been reported in the literature.

Steady-state gradient-echo sequences such as CISS (Constructive Interference in the Steady State), FIESTA (Fast Imaging Employing Steady State Acquisition) or b-FFE (balanced Fast Field Echo), have been used for assessment of acoustic schwannomas and $7/8^{th}$ root complexes. Using these sequences, ECs typically appear heterogeneous and hypointense compare to CSF and hyperintense compared with brain parenchyma with clear demarcation between EC and CSF [27]. CISS can also demonstrate insinuating margins of EC and has been shown to be one of the best sequences for ECs. However, these sequences are now being replaced by strongly T2 weighted 3D SPACE sequence. 3D SPACE sequence has limited references in literature [28]. Various types of contrasts can be produced such as T2, FLAIR, T1, strongly T2 weighted (for inner ear), MRCP etc. A study by Watanabe, et al. found 3D SPACE superior to CISS for MR cisternography [29]. The 3D T2 SPACE uses a turbo-spin-echo acquisition with an extended echo train. Strong T2 contrast is generated by manipulating the refocusing RF pulses along the train. This sequence

produces high resolution images in a reasonable scan time without the banding artefacts sometimes seen on steady-state gradient echo. The appearance of ECs on 3D SPACE is expected to be similar to CISS and, as expected, the lesion was of slightly lower signal than CSF. Because of its excellent resolution and lack of artefacts, the lesion was very well visualised and its relationship with surrounding structures was clearly delineated, in keeping with the clinical features.

The appearances of ECs are also well described on DWI. The signal intensity of various structures on DWI depends upon the ADC of the tissue and T2 relaxation time. On DWI, the ECs are variably bright, mainly thought to be due to their high T2 relaxation time, although it has been debated in the literature that there is a component of restricted diffusion responsible for the appearance. The signal of ECs on DWI can vary between quite bright to almost isointense to white matter. The ADC of ECs has been described as significantly higher than white matter and lower than CSF [30]. In the present case, the signal on DWI was on lower end of the spectrum appearing rather isointense to white matter. The ADC was intermediate between that of CSF and white matter being more typical. More importantly, DWI and ADC are most helpful in differentiating between ECs and their closest differential, arachnoid cyst, which are typically CSF isointense and show the same signal on DWI and ADC as surrounding CSF. It is clear that the DWI and ADC appearance in the present case is inconsistent with CSF and more likely to represent EC (Fig. 4).

In the present case, a provisional diagnosis of EC was made after taking several important features into consideration. It was a small intraventricular mass present at the floor of third ventricle that appeared to conform to the shape of the ventricle with insinuating margins rather than have significant mass effect. Its appearance on DWI was not that of arachnoid cyst that tends to be the commonest differential. While the appearance on 3D FLAIR and 3D T2 SPACE have not been described in literature, it was assumed that it would be somewhat similar to routine 2D FLAIR and CISS/FIESTA, where it has been commonly described. Its non-enhancing nature meant that the neoplastic and inflammatory diseases mentioned earlier are unlikely as they are known to usually enhance on contrast to variable extent and also on other imaging appearances.

Intraventricular ECs are very rare, particularly in third ventricle and no standard guidelines have been described in literature for management of these lesions, although these have been excised when symptomatic [5,9]. In the more usual location, namely the CPA cistern, the symptomatic lesions are usually excised. A total excision is usually attempted although can sometimes be difficult and may result in recurrence. They can remain stable or grow at a slow rate. These are usually followed up on MRI [31,32].

Following multispeciality review, it was decided that the lesion can be followed up without intervention especially as the patient was asymptomatic and showed no change on initial follow up. If the patient remains asymptomatic and the size stable on future MRI, it is anticipated that no intervention may be necessary, although the long term prognosis is unclear at this point. However, we fully accept that the diagnosis remains provisional in absence of surgical confirmation which is a drawback of this case report.

The current case demonstrates the imaging features of a small third ventricular lesion, particularly on 3D FLAIR and 3D T2 SPACE, presumed to be an EC. These sequences appear to be best for visualising such small intraventricular lesions. We also emphasize that these lesions can be quite subtle on standard MRI and stress upon the importance of using 3D FLAIR sequence in standard brain protocols. Unless these sequences are used and the

lesions are carefully looked for based on clinical features, these lesions can be easily missed and their presence remains unknown.

4. CONCLUSION

We have presented a rare case of third ventricular lesion presumed to be EC as seen on MRI. In particular, we have described the appearances on 3D FLAIR and 3D T2 SPACE that have not been previously described in literature, to the best of our knowledge. We highlight the importance of using 3D FLAIR in routine imaging protocols, instead of 2D FLAIR. We also highlight the superiority of 3D T2 SPACE sequence in properly describing presence of intraventricular lesions and their relationship to surrounding structures.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

Not necessary for this case report.

ACKNOWLEDGEMENTS

No funding required.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Kavar B, Kaye AH. 43 - Dermoid, epidermoid, and neurenteric cysts. Brain Tumors (Third Edition). Edinburgh: W.B. Saunders. 2012;831-48.
2. Gelabert-Gonzalez M, Garcia-Allut A, Gonzalez-Garcia J, Martinez-Rumbo R. Epidermoid cyst of the third ventricle. Neurocirugia. 2002;13(5):389-92.
3. Giannotta SL, Pauli F, Farhat SM. Epidermoid cyst of the third ventricle. Surgical neurology. 1976;5(3):164-6.
4. Rivierez M, Ridarch A, Landau-Ossondo M, Randrianbololona J. Epidermoid cyst of the third ventricle. Report of a case. Neuro-Chirurgie. 1998;44(4):283-6.
5. Shimizu Y, Aihara H, Fukawa O, Ishii M. Epidermoid of the third ventricle--report of a case. No shinkei geka Neurological surgery. 1985;13(1):71-6.
6. Ryu J, Park J, Lee J, Woo J. A Case of Epidermoid in the Third Ventricle: Case Report. . J Korean Neurosurg Soc. 1983;12(3):407-14.
7. Kadoya S, Kwak R, Emori T. Epidermoid of the third ventricle (author's transl). No shinkei geka Neurological surgery. 1980;8(3):221-7.
8. Kontoangelos K, Maltezoua M, Economou M, Kandarakia A, Papadimitriou GN. Depressive symptomatology and pineal epidermoid cyst: a case report. Acta Neuropsychiatrica. 2013;25(4):240-2.
9. Koumtchev YN, Kalnev BM, Dimitrov ZI, Gozmanov GR, Zaprianov ZN. Recurrence of epidermoid cyst in the third ventricle. Case report. Folia medica. 2002;44(1-2):97-9.

10. Sorgel HJ, Schreiber D, Heidrich R. Epidermoid of the third ventricle with recurring hemorrhage and excessive siderosis of the marginal zone. Psychiatrie, Neurologie, und medizinische Psychologie. 1972;24(2):82-9.
11. Santosh IP, Rajshekhar V. Galactorrhea as the sole presenting symptom of a posterior third ventricular epidermoid cyst. Surgical neurology. 2001;55(1):46-9; discussion 9.
12. Chen CY, Wong JS, Hsieh SC, Chu JS, Chan WP. Intracranial epidermoid cyst with hemorrhage: MR imaging findings. AJNR American journal of neuroradiology. 2006;27(2):427-9.
13. Iaconetta G, Samii M. Third ventricle epidermoid cyst. British journal of neurosurgery. 2001;15(6):529-30.
14. Li F, Zhu S, Liu Y, Chen G, Chi L, Qu F. Hyperdense intracranial epidermoid cysts: a study of 15 cases. Acta Neurochir (Wien). 2007;149(1):31-9; discussion 9.
15. Mermuys K, Wilms G, Demaerel P. Epidermoid cyst of the fourth ventricle: diffusion-weighted and flair MR imaging findings. JBR-BTR : organe de la Societe royale belge de radiologie. 2008;91(2):58-60.
16. Kaido T, Okazaki A, Kurokawa S, Tsukamoto M. Pathogenesis of intraparenchymal epidermoid cyst in the brain: a case report and review of the literature. Surgical neurology. 2003;59(3):211-6.
17. Elvord EC Jr. Growth rates of epidermoid tumours. Ann Neurol. 1977;2(5):367-70.
18. Osborn AG, Preece MT. Intracranial cysts: radiologic-pathologic correlation and imaging approach. Radiology. 2006;239(3):650-64.
19. Kallmes DF, Provenzale JM, Cloft HJ, McClendon RE. Typical and atypical MR imaging features of intracranial epidermoid tumors. AJR American journal of roentgenology. 1997;169(3):883-7.
20. Tancredi A, Fiume D, Gazzeri G. Epidermoid cysts of the fourth ventricle: very long follow up in 9 cases and review of the literature. Acta Neurochir (Wien). 2003;145(10):905-10; discussion 10-1.
21. Glastonbury CM, Osborn AG, Salzman KL. Masses and malformations of the third ventricle: normal anatomic relationships and differential diagnoses. Radiographics. 2011;31(7):1889-905.
22. Costa F, Fornari M, Felisati G, Maccari A, Bauer D, Lasio G. Epidermoid Cyst of the Pituitary Stalk: Case Report and Review of the Literature. Neurosurgery Quarterly. 2013;23(2):108-11 10.1097/WNQ.0b013e318266c267.
23. Meng L, Yuguang L, Shugan Z, Xingang L, Chengyuan W. Intraventricular epidermoids. J Clin Neurosci. 2006;13(4):428-30.
24. Nagasawa D, Yew A, Safaee M, Fong B, Gopen Q, Parsa AT, et al. Clinical characteristics and diagnostic imaging of epidermoid tumors. Journal of clinical neuroscience. 2011;18(9):1158-62.
25. Aribandi M, Wilson NJ. CT and MR imaging features of intracerebral epidermoid--a rare lesion. The British journal of radiology. 2008;81(963):e97-9.
26. Chen S, Ikawa F, Kurisu K, Arita K, Takaba J, Kanou Y. Quantitative MR evaluation of intracranial epidermoid tumors by fast fluid-attenuated inversion recovery imaging and echo-planar diffusion-weighted imaging. AJNR American journal of neuroradiology. 2001;22(6):1089-96.
27. Ikushima I, Korogi Y, Yamashita Y, Yi T, Hamatake S, Sugahara T, et al. Evaluation of intracranial lesions with inversion recovery half-Fourier single-shot turbo spin-echo MR: initial observations. AJNR American journal of neuroradiology. 1997;18(3):421-6.
28. Yamazaki R, Uchikoshi M, Hiura Y, Tsuji A, Nishiki S. T2 image contrast evaluation using three dimension sampling perfection with application optimized contrasts using different flip angle evolution (3D-SPACE). Nihon Hoshasen Gijutsu Gakkai Zasshi. 2011;67(12):1515-22.

29. Watanabe Y, Makidono A, Nakamura M, Saida Y. 3D MR cisternography to identify distal dural rings: comparison of 3D-CISS and 3D-SPACE sequences. Magnetic resonance in medical sciences. 2011;10(1):29-32.
30. Hakyemez B, Aksoy U, Yildiz H, Ergin N. Intracranial epidermoid cysts: diffusion-weighted, FLAIR and conventional MR findings. Eur J Radiol. 2005;54(2):214-20.
31. Chowdhury FH, Haque MR, Sarker MH. Intracranial epidermoid tumor; microneurosurgical management: An experience of 23 cases. Asian J Neurosurg 2013;8:21-8
32. Safavi-Abbasi S, Di Rocco F, Bambakidis N, Talley MC, Gharabaghi A, Luedemann W, et al. Has management of epidermoid tumors of the cerebellopontine angle improved? A surgical synopsis of the past and present. Skull Base. 2008;18(2):85-98.

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sciencedomain.org/review-history.php?iid=371&id=29&aid=2718