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Supratentorial intraventricular epidermoids are very rare and midline septal pellucidal epidermoids are even more uncommon with only one case being reported in available literature.^{1,2} We present only the second case of a midline septum pellucidum epidermoid in available literature.

Case Description: A 42-year-old lady with no previous complaints was admitted to the emergency services with history of intermittent headache, vomiting and giddiness of 3 months duration. A cranial computed tomography (CT) revealed a hypodense, non-enhancing intraventricular mass lesion (**Fig. 1A & B**) and cranial magnetic resonance imaging (MRI) demonstrated a non-enhancing mass lesion in the septum pellucidum suggestive of an epidermoid (**Fig. 2A-F**). She underwent endoscopic-assisted surgery via an interhemispheric transcalsal approach. Intra-

Cavum Septum Pellucidum Epidermoid- An Extremely rare occurrence

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Key Words: Cavum Septum Pellucidum, Epidermoid, Intraventricular

operatively, the lesion was located in the enlarged cavum septum pellucidum and was removed totally. The histology confirmed the diagnosis as an epidermoid (**Fig. 3A,B**). The patient recovered uneventfully and was discharged. Post-operative CT showed no residual lesion. (Figure 4)

Discussion:

Epidermoid cysts account for 1% of all intracranial tumours.¹ They are thought to occur in early embryonic life, during the period of neural tube closure during the third to fifth weeks. These are presumed to result from embryonic cell inclusions at closure of the neural tube, to produce a mass resembling stratum corneum of the epidermis and containing cholesterol crystals. These trapped committed cutaneous cells may give rise to

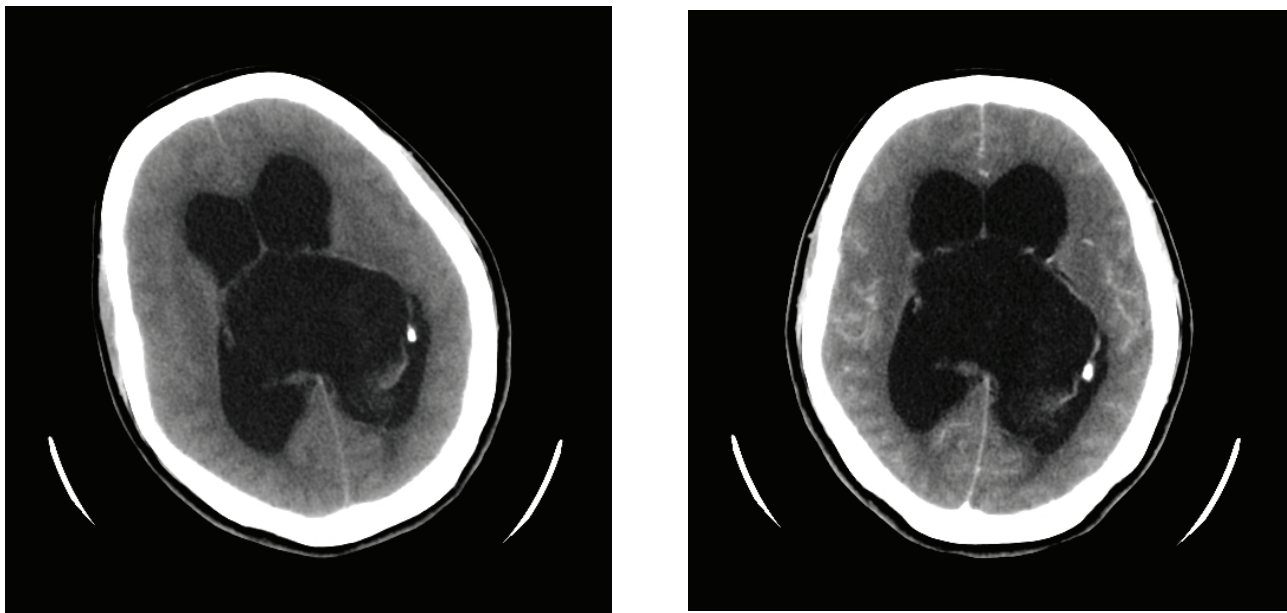


Figure 1 [A-F]: MRI (Brain) [A- Axial T1W plain, B- Axial T2W, C- Sagittal T2W, D- Coronal T2W, E- Axial T1W-post contrast, F- Axial diffusion weighted images] showing a non-enhancing mass lesion in the septum pellucidum suggestive of an epidermoid. Note the septum pellucidum (Fig 2B- Black arrow)

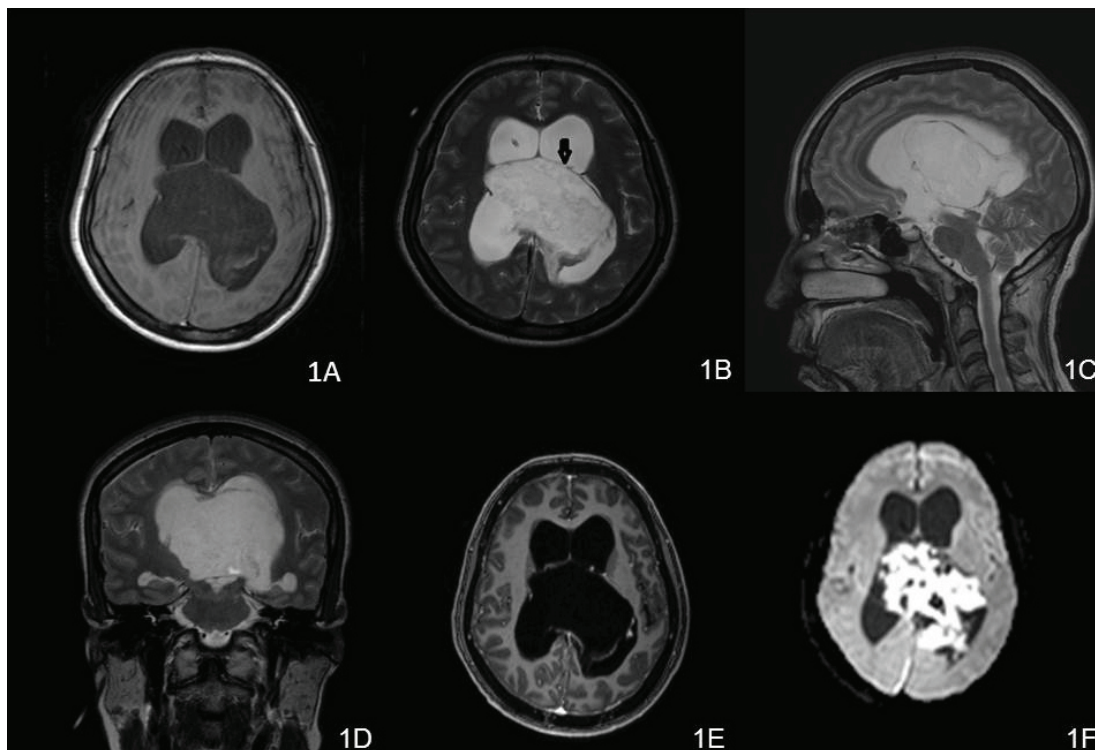


Figure 2A: Photomicrograph showing a cyst wall lined by stratified squamous epithelium and containing keratin in the lumen (H&E, X 100) B: Photomicrograph highlighting the lamellated keratin (H&E, X 100).

epidermoid or dermoid cysts.¹ They are mostly laterally placed probably due to displacements occurring during the closing of the otic and optic vesicles.¹ The preferred sites of formation of intracranial epidermoids include the cerebellopontine angle and perimesencephalic cisterns. Though midline epidermoids are unusual, the 4th

ventricle and pineal region are the preferred locations.¹ Supratentorial intraventricular epidermoids are very rare and midline septal pellucidal epidermoids are even more uncommon with only one case being reported in available literature.^{1,2}

The cavum septum pellucidum forms during the fourth

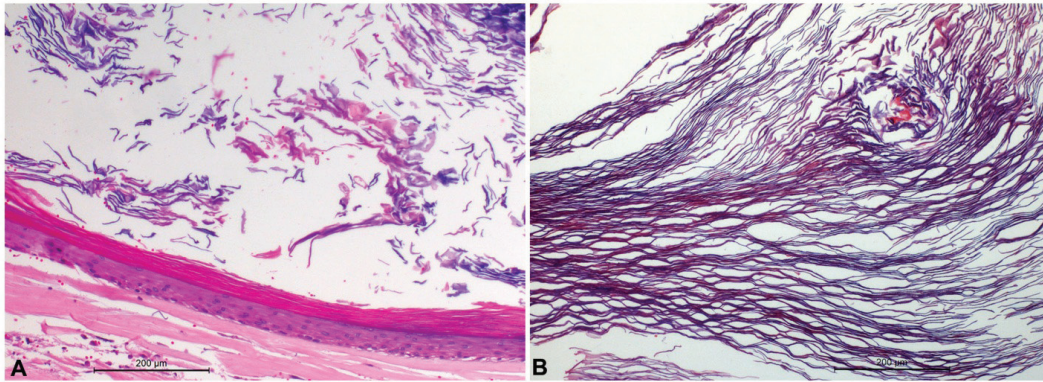


Figure 3: Postoperative CT (brain) (plain): showing complete tumor removal.

month of gestation and is present in nearly all premature infants and 15% of 6-month-old infants which is almost equal to the incidence in adults.² Although it is incorrectly called the 5th ventricle, in truth it has no communication with either the lateral or 3rd ventricles. Also, its origin is different from the ventricles and is not lined with ependymal or choroid plexus cells. Incidental (asymptomatic) cavum septum is demonstrated in almost 1% of cranial CTs. Although true cysts³ as well as empyema of the cavum septum pellucidum have been described in literature, this is only the second reported epidermoid in this location. We believe that late closure of the septum pellucidum and trapping of the epidermoid remnants within the cavum is the possible mechanism in the pathogenesis.

Supratentorial intraventricular epidermoids are very rare. Bhatoe et al in their extensive review could find only 9 cases of intraventricular epidermoids but none in the midline or in the septum pellucidum.^{1,4-7} We performed an extensive Medline and PubMed search using the keywords (epidermoid, cavum septum) and without setting any time limits and using the links provided. The search returned one case report by Bikmaz et al in 2007.² This report is the first documented case in the English language medical literature.

The diagnosis is predominantly radiological as the clinical presentation is predominantly as raised intracranial pressure along with other bizarre clinical symptoms. The management is surgical and endoscopic assistance is often required to confirm total excision. Though the pathogenesis of septum pellucidum epidermoid is the same as that of the other sites, it is rather a mystery as to the extreme rarity of epidermoid at this site. Probably a much delayed closure of the septum with entrapped epidermoid remnants could be the explanation.

Disclosure:

The authors have no conflicts of interest to disclose.

There is no financial interest to disclose.

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There are no directly related manuscripts or abstracts, published or unpublished, by any authors of this paper.

References:

1. Bhatoe HS, Mukherji JD, Dutta V. Epidermoid tumour of the lateral ventricle. *Acta Neurochir (Wien)* **148**: 339–342, 2006
2. Bikmaz K, Dinc C, Cakbay M, Iplikcioglu AC. Epidermoid cyst of the cavum septum pellucidum, *Acta Neurochir (wien)* **149**: 1271–1272, 2007
3. Lancon JA, Haines DE, Raila FA, Parent AD, Vedanarayanan VV. Expanding cyst of septum pellucidum. *J Neurosurg* **85(6)**: 1127–1134, 1996
4. Bayindir C, Balak N, Karasu A. Microinvasive squamous cell carcinoma arising in a pre-existing intraventricular epidermoid cyst. *Acta Neurochir (Wien)* **138**: 1008–1012, 1996
5. Eekhof JL, Thomeer RT, Bots GT. Epidermoid tumor of the lateral ventricle. *Surg Neurol* **23**: 189–192, 1985
6. Higashi K, Wakuta Y. Epidermoid tumour of the lateral ventricle. *Surg Neurol* **1976;5**: 363–365
7. Koot RW, Jagtap AP, Akkerman EM, Den Heeten GJ, Majole CB. Epidermoid of the lateral ventricle: evaluation with diffusion-weighted diffusion tensor imaging. *Clin Neurol Neurosurg* **105**: 270–273, 2003