DOI: 10.7860/JCDR/2021/50206.15590

Case Report

Pathology Section

Intracranial Dermoid and Epidermoid Cysts: A Case Report

ANMOL TANEJA[1], SAMARTH SHUKLA[2], SOURYA ACHARYA[3], SUNITA VAGHA[4]

ABSTRACT

Dermoid and epidermoid cysts are ectodermally derived neuraxis cysts, both lined by keratinising squamous epithelium. Epidermoid cyst is filled by friable, often lamellated keratinous debris and is devoid of cutaneous-type adnexal structures, as found in dermoid cysts. They are usually located at the cerebellopontine angle, cerebellar vermis, fourth ventricle, parasellar region, and frontal and fronto-temporal cisternal spaces but the cerebellopontine angle being their single most common location of epidermoid cysts whereas dermoid cyst clings tightly to the midline. Hereby, the authors are presenting a case report of two cases; first case is of a one-month-old child presented with 4×4 cm sized cystic swelling over occipital region since birth with redness since 15 days with complaint of pus discharge from the swelling since one day. Cyst with contents was sent for histopathological examination and features suggestive of intracranial dermoid cyst. Second case is of 14-year-old female child who presented with the complaints of headache since six months and imbalance since two months. Magnetic Resonance Imaging (MRI) was suggestive of cerebellopontine angle tumour with hydrocephalus. Tumour was excised and sent for histopathological examination and features suggested of epidermoid cyst. These benign lesions are potentially curable. As these cases were suspected as malignant tumours but as they found out to be diagnosed as benign lesions. Although these lesions are benign but can still lead to some seroius complications. We are reporting these cases for its early diagnosis and thereby better management.

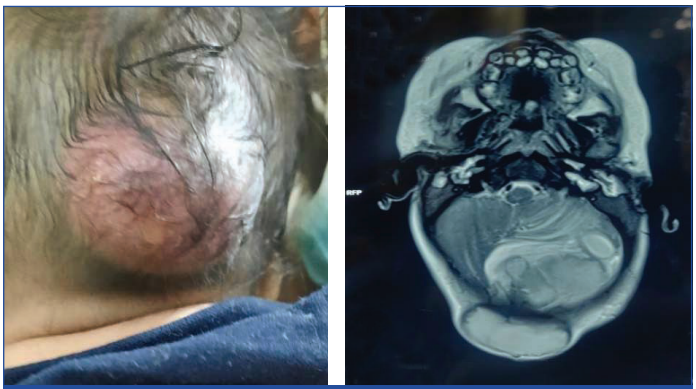
Keywords: Anucleated squames, Cerebellopontine angle, Neuraxis cysts

CASE REPORT

Case 1

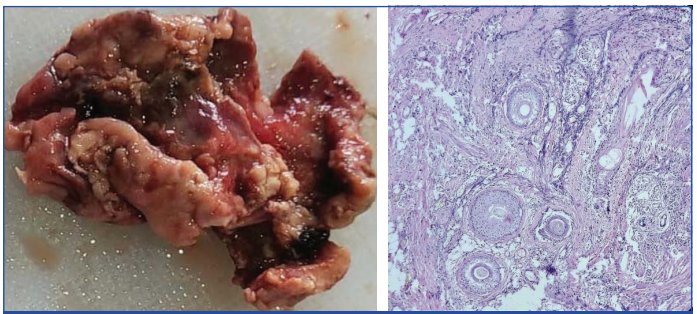
A one-month-old male child presented to Neurosurgery Outpatient Department (OPD) with swelling over occipital region since birth with redness since 15 days with complaint of pus discharge from the swelling since one day. No history of altered sensorium, vomiting, fever, unconciousness was present. No signs of raised intracranial tension were present, as told by mother. No significant past history was present. On clinical examination, swelling was 4×4 cm in size, cystic, fluctuant with raised local temperature, as shown in [Table/Fig-1].

Intraoperatively, subcutaneous abscess was seen with evidence of fibrous tract connecting skin and traversing intracranially. Cyst with contents of pearly white flakes and yellowish discharge along with hair was noted. Magnetic Resonance Imaging (MRI) Brain was suggestive of occipital meningocele, approximately 35 mm in size with small defect in occipital bone, as shown in [Table/Fig-2]. Differential diagnosis included Rathke cleft cysts with extensive squamous metaplasia, arachnoid cysts, inflammatory cysts like neurocysticercosis, neurantric cysts, craniopharyngiomas, acoustic schwannomas. Provisional diagnosis was made possibly as intracranial cystic lesion.



[Table/Fig-1]: Clinical picture depicting occipital swelling.
[Table/Fig-2]: T2 weighted MRI imaging showing occipital meningocele, approximately 35 mm in size with small defect in occipital bone. (Images from left to right)

Cyst with its contents was sent for histopathological examination. Grossly, tissue piece was irregular, brownish white in colour, measuring approximately 6×3.5×1.5 cm. On cut section, reddish velvety areas identified, as shown in [Table/Fig-3]. Microscopically, cyst was lined by keratinised squamous epithelium, deeper tissue piece shows extensive acute on chronic lymphoplasmacytic inflammatory reaction along with granulation tissue formation. At places, hair follicles were also noted, as shown in [Table/Fig-4]. Histopathological features were suggestive of intracranialdermoid cyst. Follow-up was advised and patient was asked to come after 10 days to check whether any signs of raised intracranial tension or recurrence were present.



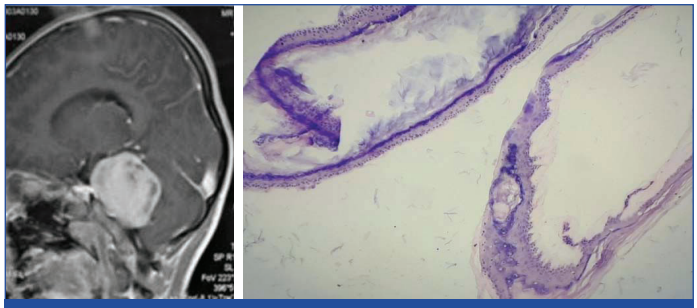
[Table/Fig-3]: Gross appearance of the specimen showing brownish white, irregular areas. **[Table/Fig-4]:** Microscopy (10X magnification), on H&E staining section showing cystic areas with inflammatory infiltrates and hair follicles. (Images from left to right)

Case 2

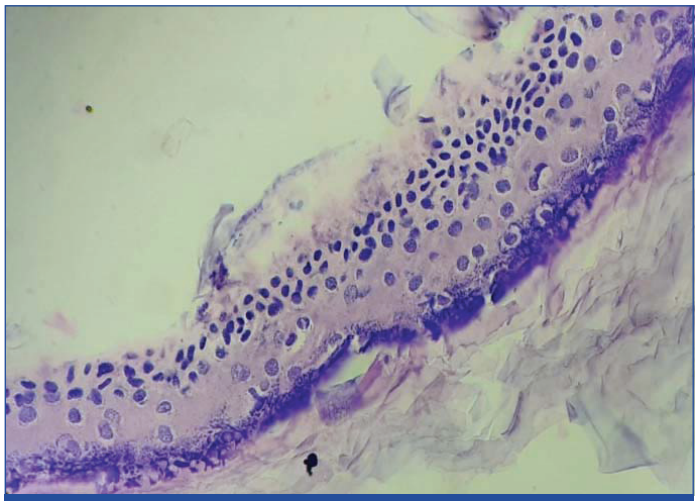
A 14-year-old female child presented to Neurosurgery Outpatient Department (OPD) with the chief complaints of headache since six months with complaint of imbalance since two months. Patient was well oriented to time, place and person lying comfortably on bed. No history of altered sensorium, vomiting, fever, unconsciousness, difficulty in taking food was present. No significant past history was present.

On examination, hydrocephalus was present, but no signs of tremours, stupor, memory loss, tingling sensation, double vision were present. The MRI Brain was suggestive of cerebellopontine angle tumour with hydrocephalus, as depicted in [Table/Fig-5]. Differential diagnosis made were; acoustic neuroma, meningioma, ependymoma, neuroepithelial cyst, arachnoid cyst, epidermoid cyst, schwannoma, gliomas. Intraoperatively, pearly white material inside the tumour capsule was found. Tumour was excised and sent for histopathological examination. Grossly, multiple, irregular, brownish tissue pieces were received aggregating 4×4 cm.

Microscopically, fibrous wall was lined by keratinising squamous epithelium, contained anucleated squames, keratin flakes but no skin adnexae, pilosebaceous glands or hair are found, as depicted in [Table/Fig-6,7]. Features are suggestive of epidermoid cyst. Ventriculoperitoneal shunt was also applied to patient for treatment of hydrocephalus and patient was asked to come after 15 days as follow-up to check whether any signs of raised intracranial tension or recurrence were present.



[Table/Fig-5]: T2 weighted MRI imaging showing mass at cerebellopontine angle, approximately 45 mm in size. **[Table/Fig-6]:** Microscopy (10X magnification) on H&E staining section showing cyst wall lined by keratinising squamous epithelium, contains anucleated squames, keratin flakes. (Images from left to right)



[Table/Fig-7]: Microscopy (40X magnification) on H&E staining section showing cyst wall lined by keratinising squamous epithelium.

DISCUSSION

Intracranial dermoid and epidermoid cysts, both lined by keratinising squamous epithelium [1] is rare, benign, slow growing neoplasms, ectodermally derived [2]. They expand through agglomeration of desquamation products and sebaceous secretions in a cystic cavity rather than via cell division, they are not true neoplasms. They are usually located at the cerebellopontine angle, cerebellar vermis, fourth ventricle, parasellar region, and frontal and frontotemporal cisternal spaces but the cerebellopontine angle being their single most common location of epidermoid cysts whereas dermoid cyst clings tightly to the midline [2] and this is one of the major points which differentiates epidermoid cyst from dermoid cyst.

Common symptoms include headache and seizure and epidermoid cysts mostly affect patients in young adults or middle age whereas dermoid cyst usually present in childhood or adolescence [1]. On MRI, dermoid cysts are hyperintense on T1WI and heterogenous on T2WI [2], hypodence on CT scan [3]. Whereas, epidermoid cysts are slightly hypointense signal intensity on T1-weighted and intermediate to high signal on T2-weighted [4], which is also one of the most important feature to differentiate dermoid cyst from epidermoid cysts. Dermoid cysts are benign formations, and have a generally good prognosis. Surgical treatment is only indicated when dermoid cysts cause mass effect and serious neurological deficits [2]. However, spontaneous rupture of these benign lesions is reported in only a small percentage of cases [5] and due to which there is often dissemination of their intracystic contents into the subarachnoid space and ventricles [6]. Other complications associated are bacterial meningitis, cerebellar abscesses, neoplastic transformations [1]. Ammor H et al., conducted a study and found that when dermoid cysts become extensive, they lead to compression of neurologic and vascular structures, producing cerebral hypertension [2]. Tandon N et al., conducted a study and found that traumatic rupture of an intracranial dermoid cyst was an extremely rare event and this was only the fourth such case reported in the literature and they concluded that this rupture occurred due to sudden shifts in the cyst sac, which was adherent to nearby partially mobile intracranial contents [5].

Conditions which may mimic include; Rathke cleft cysts with extensive squamous metaplasia, arachnoid cysts, inflammatory cysts like neurocysticercosis, neurantric cysts, craniopharyngiomas, acoustic schwannomas [1].

Often there is a difficulty to cure these lesions because of their sliding growth into different spaces and cisterns, also due to involvement of cranial nerves and vessels, radical excision is difficult [7]. These benign lesions are potentially curable [8], but it is best to avoid surgery while the cyst is actively inflammed due to a higher risk of infection, wound dehiscence and cyst recurrence. Ruptured intracranial epidermoid cyst can be treated by use of steroids and antibiotics with combination of surgical resection along with irrigation with saline during surgery [9]. Radical resection is the choice of treatment but sometimes, cyst capsule adheres firmly to vital structures, so radical removal can be dangerous. In that cases subtotal resection may be a wise option [10]. The recurrence rate of the epidermoid cysts is found to be in between 1% and 54%. In cases of recurrence, re-operation should be performed when the patient again becomes symptomatic [11]. In cases of intracranial dermoid cysts, recurrance rate of 9% has been reported by some studies [12]. Following the resolution of the infection, the lesion is excised [4]. Present cases included intracranial benign lesions, but can lead to compression of neurologic and vascular structures, nearby vital structures, if become extensive can also produce cerebral hypertension, if not diagnosed and managed properly.

CONCLUSION(S)

Although these lesions are benign but can still lead to some serious complications. Authors reported these cases for its early diagnosis and thereby better management. The MRI is superior than CT in diagnosisng these lesions as they provide complete information of the extent of these lesions. Complete surgical resection is the choice, but must be thinking about against the risk for injury to nearby vital structures. Complications associated should be managed and monitored closely in the perioperative period. The recurrence rate following resection is extremely low, but incompletely resected cysts should be followed on a regular basis.

REFERENCES

[1] JuanRosai; Rosai and Ackerman's Surgical Pathology: Tenth Edition, Volume 2 Central Nervous System: Page 1954.
[2] Ammor H, Addou O, Boujarnija H, Lamrani Y, Boubbou M, Maâroufi M, et al. A cause of seizure: intracranial dermoid cysts: Eurorad. (2014, Jun. 14).
[3] Sanchez-Mejia R, Limbo M, Tihan T, Galvez M, Woodward M, Gupta N. Intracranial dermoid cyst mimicking hemorrhage. J Neurosurg. 2006;105(4):311-14.

[4] Hoang V, Trinh C, Nguyen C, Chansomphou V, Chansomphou V, Tran T. Overview of epidermoid cyst. Eur J Radiol Open. 2019;6:291-301.
[5] Tandon N, Esquenazi Y, Kerr K, Bhattacharjee M. Traumatic rupture of an intracranial dermoid cyst: Case report and literature review. Surg Neurol Int. 2013;4(1):80.
[6] Abramson R, Morawetz R, Schlitt M. Multiple complications from an intracranial epidermoid cyst: case report and literature review. Neurosurgery. 1989;24(4):574-78.
[7] Chowdhury F, Haque M, Sarker M. Intracranial epidermoid tumour; microneurosurgical management: An experience of 23 cases. Asian J Neurosurg. 2013;8(1):21.
[8] Murrone D, Romanelli B, Ierardi A. Ruptured intracranial dermoid cyst: A case report. Integrative Molecular Medicine. 2016;3(5):793-95.
[9] Yan W, Xu L, Wu Q, Chen G, Zhang J, Wei S, et al. A case report of spontaneous rupture of intracranial epidermoid cyst with dramatic increase of serum carbohydrate antigen 199: A three-year follow-up study. BMC Neurol. 2015;15(1):198.
[10] Caldarelli M, Massimi L, Kondageski C, Di Rocco C. Intracranial midline dermoid and epidermoid cysts in children. J Neurosurg. 2004;100(5):473-80.
[11] Ghartimagar D, Shrestha M, Ghosh A. Recurrence of ruptured intracranial epidermoid cyst-A rare case report and presentation. Int J Surg Case Rep. 2020;76:310-14.
[12] Mouskou S, Stamati A, Sfakianos G, Malama A, Gavra M, Vartzelis G, et al. Ruptured intracranial dermoid cyst in a child. Cause of atypical neurological symptoms. Interdisciplinary Neurosurgery. 2020;20:100482.

PARTICULARS OF CONTRIBUTORS:
1. Junior Resident, Department of Pathology, Jawaharlal Nehru Medical College, Sawangi, Maharashtra, India.
2. Professor, Department of Pathology, Jawaharlal Nehru Medical College, Sawangi, Maharashtra, India.
3. Professor and Head, Department of Medicine, Jawaharlal Nehru Medical College, Sawangi, Maharashtra, India.
4. Professor and Head, Department of Pathology, Jawaharlal Nehru Medical College, Sawangi, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Anmol Taneja,
122-A, Model Town, Fatehabad, Haryana, India.
E-mail: anmoltaneja1415@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: May 07, 2021
- Manual Googling: Aug 18, 2021
- iThenticate Software: Sep 20, 2021 (20%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **May 06, 2021**
Date of Peer Review: **Jun 29, 2021**
Date of Acceptance: **Aug 19, 2021**
Date of Publishing: **Nov 01, 2021**