Intrinsic Epidermoid Cyst of the Brainstem in Children – Case Report and Review

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Case Report

Keywords: Intrinsic Brainstem, Epidermoid cysts, Epidermoid Surgical resection, Pontomedullary, Children

Posted Date: February 9th, 2023

DOI: https://doi.org/10.21203/rs.3.rs-2557297/v1

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Abstract

Purpose

The study aimed to summarize all published cases of intrinsic brainstem epidermoid cysts in a timeline to highlight the specific characteristics and individualize the disease, in addition to discussing the best treatment used.

Methods

The scientific literature on pediatric cases of intrinsic epidermoid cysts of the brainstem was analysed. We present the case of a 1.5-year-old male with incidental presentation, who was treated with gross total resection. We summarize all previously published cases to individualize the disease.

Results

We identified 21 patients, including 10 boys and 11 girls, with a mean age of 4.85 (1–15) years at the time of surgery. The most frequent symptoms were cranial nerve palsy (71.4%), pyramidal tract deficit (57.14%) and headache (52.38%). Among the affected cranial nerves, VII was the most frequent, reported in 10 patients.

Conclusion

Brainstem epidermoid cysts are extremely rare pathologies with relevant age involvement in young children. The treatment objective should be the maximum resection of the

Introduction

The epidermoid cyst (EC) is a rare benign lesion that affects the central nervous system (CNS) with an incidence that varies from 0.2 to 1.8% of all intracranial expansive lesions and constitutes the main diagnosis of non-neoplastic cystic lesions. [1, 2]

The common location of CNS involvement is in the basal cistern, especially at the angle of the cerebellar and parasellar points, and involvement in the brainstem is extremely uncommon. [3–6]

Case

A 1.5-year-old male, previously health, presented an expansive lesion an incidental finding on CT scan after mild traumatic brain injury. Physical examination showed only headache, without neurological deficit. In an investigation with cerebral MRI, an intrinsic expansive lesion was observed in the
pontomedullary transition, the T1W image was heterogeneous, with a hyperintense center and hypointense periphery, on T2W predominantly hyperintense and without contrast-enhancement. The lesion presented hyperintense diffusion, being confirmed as true diffusion in the ADC (apparent diffusion coefficient), in addition, no bleeding was found in the SWI sequence. The lesion, despite being intrinsic, deformed the anterior part of the brainstem and displaced the pons cranially.

The patient was operated on in the prone position with electrophysiological monitoring, submitted to a median suboccipital craniotomy associated with Far lateral extension and resection of the C1 arch, with the aim of exposing the ventral portion of the brainstem. With dissection through the lower cranial nerves, an exophytic pontomedullary lesion was identified anterior to the olive. An incision was made in a region without electrophysiological response, with subsequent dissection and irrigation of the lesion, with the output of viscous content and pearly color being perceived. Capsular portions adhered to the brainstem were not resected.

The material was analyzed for pathology, which confirmed it as stratified squamous epithelium compatible with an EC. The patient evolved stable without neurological deficit in the postoperative period, and a new brain MRI was performed at 6 months of follow-up.

**Discussion**

**Pathogenesis**

The origin is still uncertain, the main agreement in the literature is that the origin of the problem is from a failure of neural tube closure between, in which there is a failure in the disjunction between the differentiated neural ectoderm and the ectoderm. [7, 8]

Theories emerged trying to justify the most distinct presentations reported, such as multiple, intraventricular and intraparenchymal cysts. Kaido et al suggest that according to the gestational age at which the defect occurs and the position of the ectodermal elements in relation to the neural tube, which may be inside, superficial or close to the optic vesicles, presentations can occur both intra-axially and extra-axially. In addition, it proposes that failures that occur earlier in gestational life are associated with intraparenchymal and intraventricular lesions. [5, 7, 9]. Obana et al, in their series, suggest that the cysts become intra-axial from the spontaneous flexion of the metencephalon during embryonic development, which would result in total involvement by the brainstem parenchyma, which may justify the location similar all the cases.[10]

**Histology**

The pathologist Cruveilhier described it for the first time in the literature in 1829, characterizing it as “the most beautiful of all tumors” due to its pearly color.

The EC is formed by keratinized stratified squamous epithelium over a thin layer of fibrous tissue that is in contact with the nervous tissue. The fluid content comes from the breakdown of cells generating the
accumulation of keratin and cholesterol. Its cellular characteristic is the same as that of the epidermis, resulting in slow growth of the lesion over time.[6]

Symptomatology

Teegala et al, showed a significant difference in the time from symptoms to diagnosis, while adults had an average of 42 months, children had 1.5 months of symptoms to make the diagnosis. [11–13]. The main symptoms described are headache, cranial nerve palsy, hemiparesis and ataxia. The exophytic extension commonly obstructs the fourth ventricle, resulting in obstructive hydrocephalus.[11]

Radiological Aspects

Magnetic resonance imaging (MRI) is the main exam for the investigation. Mostly, the EC is perceived as hypointense on T1W MRI and hyperintense on T2W MRI, with little or no enhanced-contrast, demonstrating an intensity slightly higher than that of CSF.[7]

Diffusion and ADC mapping, appears to be the most consistent and useful sequence in the diagnosis of EC, demonstrate true restriction resulting from the hypercellularity of the cyst that develops from multiple cell layers [14, 15]

Five cases, including the one we described, present radiological features of the white EC. Mishra et al, first described the white EC as cyst that presents the reverse signal in the images, in this way the T1W signal would be increased and the T2W signal would be a low signal, in addition, they are not completely suppressed in the FLAIR and has high broadcast signal. The main justification consists of a high protein cystic content due to a proliferative and exudative defense reaction, being noticeable in histology by the cellular increase of polymorphonucleates in its content. [16, 17] In the pediatric population, hypersignal is not rare, but rather a diversity that should be considered in the diagnosis.

Treatment

The therapeutic is the microsurgical evacuation of the cystic contents with the total resection of the capsule, thus avoiding recurrences [7, 18]. Cavalheiro et al, emphasize that treating brainstem lesions is extremely technically difficult, especially in pediatric patients. Planning through diagnostic tests, choosing safe areas of access to the brainstem and the use of appropriate materials make therapy safer and more effective by reducing morbidity and mortality. [19]

The cases need to be individualized to obtain the best treatment. Different authors describe capsule adhesions that made total resection impossible, but 7 cases were described as total resection in a single approach and without prioritizing the neurological deficit, suggesting that we can try total capsule excision.

In conclusion, the treatment objective should be the maximum resection of the lesion through a careful approach and with the appropriate tools for the functional preservation of the patient.
Declarations

Ethics approval and consent to participate: The study was approved by the Research Ethics Committee of the Universidade Federal de São Paulo, São Paulo, Brazil. Informed consent was obtained from the guardians.

Consent for publication: The informed consent was obtained from the guardians for publication.

Availability of data and material: not applicable

Competing interests: no competing interests to declare

Funding: Not applicable


Acknowledgements: Not applicable

References


Tables

Table 1 and 2 are available in the Supplementary Files section.

Figures
Figure 1

Legend not included with this version.
Figure 2

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Figure 3

Legend not included with this version.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.
- IntrinsecEpidermoidCystoftheBrainstem.mov
- TABLE1..tiff
- TABLE2..pdf