



CLINIC, DIAGNOSIS AND SURGICAL TREATMENT OF EPIDERMOID CYSTS OF THE BRAIN (LITERATURE REVIEW)

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<https://www.doi.org/10.5281/zenodo.7855021>

ARTICLE INFO

Received: 13th April 2023

Accepted: 21th April 2023

Online: 22th April 2023

KEY WORDS

Epidermoid, epidermoid cyst, surgical treatment.

ABSTRACT

The work presents the results of surgical treatment of epidermoid cysts of the brain. Currently, various types of surgical techniques for this pathology are being continuously improved, but there are no studies with a high level of evidence base on the significant advantages of one or another surgical treatment tactic.

Introduction

Intracranial epidermoid cysts, resulting from a violation of the neural tube anlage at 3-5 weeks of fetal development, are rare heterotopic, dysontogenetic, benign formations. According to world data, the incidence of intracranial epidermoid cysts is 0.2-1.8% among all primary brain tumors [2,3]. Epidermoids do not have a typical localization, occurring both in the medulla and in the bones of the vault and base of the skull, being located both intradurally and extradurally. Epidermoids grow slowly and develop mainly in middle-aged and elderly people. Aseptic inflammation often develops around the epidermoid, and when ruptured, the contents (keratin) break through into the subarachnoid space, into the cerebrospinal fluid spaces, causing aseptic meningitis [1].

The only effective treatment for epidermoid cysts is surgical removal. At the same time, the invasive nature of growth and other features of the biological behavior of these neoplasms determine a rather high risk of cranial nerve dysfunction and the development of aseptic meningitis in the postoperative period. First of all, this is true for epidermoid cysts localized in the ponto-cerebellar angle and in the middle cranial fossa [4]. The high recurrence rate of epidermoid cysts, which, according to various sources, is 10-24%, leads to the need for repeated operations, which significantly increases the risk of neurological complications. The results of treatment of epidermoid cysts should be considered not entirely satisfactory due to the high incidence of cranial nerve dysfunction and aseptic meningitis after surgery. The causes of relapses and complications of surgical treatment of epidermoid cysts are not completely clear [5,6].

Epidemiology and topographic - anatomical features

According to world data, the incidence of intracranial epidermoid cysts is 0.2-1.8% among all primary brain tumors. Epidermoids do not have a typical localization, occurring both in the medulla and in the bones of the vault and base of the skull, being located both intradurally and extradurally [8]. The most common location for epidermoid cysts is the



ponto-cerebellar angle (40 – 50%), IV ventricle (17%), parasellar region and middle cranial fossa (10 – 15%), large hemispheres of the brain (rare 1,5%) [9]. Epidermoid cysts are characterized by slow growth within the subarachnoid space. The growth of these formations is characterized by the shape of the filled cavity and often compresses the corresponding vessels and nerves. For this reason, there is a slow progress in the development of clinical symptoms and an increased intraoperative risk of damage to the corresponding structures of the brain [9,10,11].

Pathological features

Epidermoid cysts are externally covered by a thin capsule of keratinized stratified squamous epithelium, which grows as a result of desquamation of epithelium, later breaking down into keratin and cholesterol. They are named “pearly tumors” for their white and glittering aspect. Inside the capsule they are filled with a soft, waxy, white material formed by the desquamation and deterioration of keratin from epithelial internal layer of the cysts [12].

Clinical presentation

The symptomatology observed in this pathology may vary depending on the location and size of the epidermoid itself. Clinical symptoms are due to the fact that epidermoids spread along the cisterns of the base of the brain, involving cranial nerves and vascular structures in the process [13]. epidermoid cysts are usually discovered in adults, commonly during the 5th decade (mean age of 55 years). Very few occur in childhood, because of their very linear and slow growth. Epidermoid tumors symptomatology reflect the location of the tumor: the most frequently involved cranial nerves are the V, VII, VIII and IX. Clinical neurological features can be caused by cranial nerves irritation (resulting in trigeminal or glossopharyngeal neuralgia or hemifacial spasm) or compression (with functional deficit). Permanent cranial nerve paralysis usually occurs only after the adhesion of the tumor to the nerve, causing ischemia. Other symptoms are usually attributed by compression of anatomical structures close to the neoformation. Patients often report headache and seizures at clinical presentation, which in turn is due not to intracranial hypertension but to irritation of the meninges by epidermoid metabolites [14].

Imaging

Diagnosis of epidermoid is difficult due to their slow growth and absence of clinical symptoms for a long time. When clinical signs appear, the epidermoids reach a significant size. CT and MRI are fundamental diagnostic tools to gain useful information for surgical programming [15]. As additional methods of examination for the diagnosis of epidermoid cysts of the brain, neurological examination, examination of the fundus, EEG are used. In some cases, for differential diagnosis, such research methods as MSCT - angiography of cerebral vessels or selective cerebral angiography are used.

Nonenhanced CT scans show a well-defined, lobulated, low density mass insinuating around the cisterns, sometimes with deep invagination into the adjacent brain parenchyma. Most epidermoid cysts are non enhancing, but may show calcification or hemorrhage [17]. Moreover, CT scan allows to study diploe erosion: it's unusual to observe both diploic tables involved (usually just the inner table is) but in giant epidermoid cysts a massive diploic erosion can be observed [14].



MRI allows to better characterize the lesion, showing low-intensity signal on T1-weighted images, high-intensity on T2-weighted images, FLAIR heterogeneous signal (due to cholesterol and cellular debris) with moderate peripheric enhancement and diffuse hypersignal. Epidermoid cysts show restricted diffusion with higher signal intensity than that of cerebrospinal fluid (CSF) on diffusion-weighted imaging [12]. According to the world literature, there are distinctive signs of epidermoid cysts on MRI: 1) with large volumes of formation, an insignificant “mass effect” is noted; 2) uneven, bumpy (serrated) contour of the formation; 3) absence of perifocal edema; 4) unexpressed hydrocephalus even with large volumes of education; 5) there is no signal change after contrasting [8].

Differential diagnosis

Epidermoid cysts with localization in the cerebellopontine angle are mainly differentiated from other tumors of the same localization, such as caudal nerve neuromas, dermoid cysts, arachnoid cysts, meningiomas, and cancer metastases. Differential diagnosis of epidermoid cysts located in the chiasm-sellar region is carried out with pituitary macroadenomas, meningiomas, craniopharyngiomas and aneurysms of the anterior cerebral artery. With the localization of epidermoid cysts in the ventricles of the brain, differential diagnosis is carried out with dermoid cysts, colloid cysts and choroid plexus meningiomas [8]. In very rare cases, epidermoid bones are found, in this case, it is also necessary to carry out differential diagnosis [16].

Surgical treatment

The only effective treatment for epidermoid cysts is surgical removal. At the same time, the invasive nature of growth and other features of the biological behavior of these tumors determine a rather high risk of cranial nerve dysfunction and the development of aseptic meningitis in the postoperative period. Radiotherapy and chemotherapeutic methods of treatment are ineffective for epidermal cysts of the brain.

True epidermoid cysts are located intradurally and present difficulties for radical removal. Some authors proposed to divide the removal of epidermoid cysts of cerebellopontine angle into the following stages:

Stage 1 - opening the arachnoid membrane, examining the roots of the caudal groups of nerves and anterior inferior cerebellar artery;

Stage 2 - intracapsular removal of the tumor;

Stage 3 - visualization of Dandy's vein and trigeminal nerve roots;

Stage 4 - separation of the epidermoid cyst capsule from the brain stem.

According to some authors, incomplete removal of the epidermoid cyst while maintaining anatomical integrity is considered a better option than the risk of surgical catastrophe. Of particular difficulty for surgical removal are those epidermoid cysts that involve the caudal group of nerves from two sides. Damage of these nerves, even temporary, entails such a formidable complication as bulbar syndrome [8].

Complications of surgical removal

Supporters of the total removal of epidermoid cysts note that the radical removal not only prevents continued growth, but also prevents the occurrence of aseptic meningitis. Aseptic meningitis (AM) is a complication specific to these neoplasms that occurs, according to the literature, in 5–40% of cases. Some authors believe that such a complication as aseptic



meningitis occurs due to the ingress of a cholesterol crystal into the cerebrospinal fluid system, which is most often observed with incomplete resection of the epidermoid cyst [8]. Often, people who have had aseptic meningitis develop aresorptive hydrocephalus due to adhesive arachnoiditis, which leads to fibrosis and marked thickening of the meninges [7].

Actions that reduce the probability of developing aseptic meningitis in the postoperative period include: total removal of the epidermoid cyst along with the capsule, avoidance of epidermoid masses getting into the cerebrospinal fluid [7].

Conclusion

Despite the studied aspects and available data on the features of the clinical picture, diagnostic methods and approaches to the surgical treatment of epidermoid cysts of the brain in the world literature, this topic still remains relevant. In our work, we intend to study the features of the above issues in more depth, and draw our own conclusions.

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