Endoscopic management of intracranial arachnoid cysts

Essay

Submitted for Partial Fulfillment of Master Degree in General Surgery

By

Ehab Ahmed Mohamed El Refaee

M.B.B.Ch.

Cairo University

Supervised by

Prof. Dr. Ahmed Sherif Kareem

Professor of General Surgery
Cairo University

Prof. Dr. El Gohary Mohamed El Gohary

Professor of Neurosurgery
Cairo University

Prof. Dr. Khaled Bassiem Aly

Professor of Neurosurgery
Cairo University

Faculty of Medicine

Cairo University

2008

Acknowledgment

First of all I would like to thank GOD for giving me the power to complete this work, may HE be generous on me and give me the knowledge to help others.

I would like to express the sincerest gratitude to **Prof. Dr. Ahmed Sherif Kareem**, Professor of General surgery, Faculty of Medicine, Cairo University for his guidance and continuous support.

Any words won't express my feelings towards **Professor Dr. Ahmed Zohdi Mostafa Zohdi**; to whom I am grateful to be his student.I learnt and still learning treasures every day I am with him;in life, neurosurgery in general, and in neuroendoscopy in particular.

I am also very thankful to **Prof. Dr. El Gohary Mohamed El Gohary,** Professor of Neurosurgery, Faculty of Medicine, Cairo University giving me the privilege to work under his supervision, and for his guidance and continuous support .

I am sincerely grateful to **Prof. Dr. Khaled Bassiem Aly**, Professor of Neurosurgery, Faculty of Medicine, Cairo University. His passion for excellence and his unlimited patience made this work what it is.

I would like also to thank all my colleagues who supported and guided me ;especially to my colleague who helped me in finishing this work.

Last but not least, a special tribute is owed to my family for their never ending patience and support.

List of Abbreviations

AICA	*Anterior inferior cerebellar artery*
ACoA	*Anterior communicating artery*
CPA	*Cerebello-pontine angle*
CT	*Computerized tomography*
CSF	*Cerebrospinal fluid*
ICA	*Internal carotid artey*
ICP	*Intra—cranial pressure*
MRI	*Magnetic resonance imaging*
PICA	*Posterior inferior cerebellar artery*

Contents

List of Tables

List of Figures

Abstract

Arachnoid cysts most probably arise from incomplete separation of the perimedullary mesh (endomeninx) during the early stages of embryogenesis.

Arachnoid cysts present due to compressing adjacent neural structures, increasing the intracranial pressure by their own mass effect and by obstructing CSF flow and causing hydrocephalus.

Endoscopic management of arachnoid cysts is a safe and effective modality provided that each case is approached individually and studied carefully intraoperatively.

Key words: Arachnoid-Endoscope-Intracranial pressure-Hydrocephalus.

Aim Of the work

Although arachnoid cysts are a relatively benign pathological entity treatment in children, or in patients with symptomatic lesions regardless of age can be managed with endoscopically guided cyst wall fenestration into the ventricular system or CSF containing cisterns.

Introduction

Arachnoid cysts are benign developmental cysts, that occur in the cerebrospinal axis in relation to the arachnoid membrane. The cysts generally contain clear, colorless fluid resembling normal CSF. Rarely they contain xanthochromic fluid (***Rengachary and Kennedy., 1996).*** Arachnoid cysts account for about 1% of intracranial space occupying lesion, and occurs in about, 0.5% of autopsies as an incidental finding (***Mastuda, et al., 1982***).

Arachnoid cysts arise during development from splitting of arachnoid membrane (thus they are technically intra-arachnoid cysts). ***(Greenberg 2005).***

The lesions are congenital as they have predominant presentation in the first and second decades of life, rarely to be hereditary and more than one cyst occur in the same patient in fewer than 10% (***Raffel et al., 1988)***.

Two thirds are supra tentorial and one third are infra tentorial (***Robinson, 1971)***. An understanding of the mechanism commonly thought to underlie the development of arachnoid cyst requires a brief review of the embryology of the subarachnoid spaces (***Gandy and Heier, 1987)***.

A complete picture of the morphology of the arachnoid cyst emerges only when such cysts are dissected at autopsies ***(Rengachary and Kennedy, 1996)***.

An analysis of the distribution of arachnoid cysts in the intracranial cavity leads to inevitable conclusion that these cysts occur nearly always in relation to an arachnoid cistern ***(Rengachary and Kennedy, 1996)***.

Arachnoid cysts present due to compressing adjacent neural structures, increasing the intracranial pressure by their own mass effect and by obstructing CSF flow and causing hydrocephalus ***(Pradilla and Jallo,2007)***.

With universal use of C.T and M.R.I as noninvasive neurodiagnostic tests increasing number of these cysts are recognized and treated, concurrent with this is resurgence of interest in the origin, pathogenesis and structures of these lesions ***(Rengachary and Kennedy, 1996)*** .

M.R.I. scanning is the imaging modality of choice for visualizing arachnoid cyst with addition of FLAIR. It can also be seen in C.T. but M.R.I. is useful for demonstration of multiplaner relationship, localizing middle and posterior fossa cysts and differentiating arachnoid cysts from other cystic lesions that may also appear hypodense in the C.T. ***(Osborn,2004)***.

Numerous operative procedures have been recommended, including stereotactic aspiration, cyst excision, endoscopic cyst fenestration, cysto-cisternostomy, ventriculocystostomy, and cysto-peritoneal shunting. However, it remains controversial as to which is the best method. Recently, the introduction of neuroendoscopy has provided neurosurgeons with a new technique for the treatment of arachnoid cysts ***(Choi et al., 1999).***

Because of their benign nature and slow extension, arachnoid cysts may remain asymptomatic or produce only subtle symptoms and signs,

on the other hand, they sometimes give rise to focal neurological deficit, raised intracranial pressure and or epileptic seizures, the question of when this lesion should be operated upon is ,therefore, not always easy to answer, moreover, the choice of the most appropriate surgical approach is still debated ***(Greenberg,2005).***

Historical back ground terminology

Arachnoid cysts seem to be the simplest and most appropriate term. Leptomeningeal cyst, is less specific, because leptomeninges refers to both the arachnoid and the pia mater, and the latter is not involved in the pathogenesis of these cysts. The term subarachnoid cyst is somewhat inaccurate in that, although the cysts encroach on the subarachnoid space, their primary location is within the arachnoid membrane. Intra-arachnoid cyst is perhaps a more accurate term in the light of histologic observation, but it has not gained wide acceptance. When the cyst is in communication with the subarachnoid space, it has been variously named arachnoidocele, arachnoid pouch, arachnoid diverticulum, internal meningocele, etc., but such a plethora of terms may be avoided by the use of the terms communicating and noncommunicating arachnoid cyst. While communication or lack of it with the subarachnoid space may be an important factor in the pathophysiology of arachnoid cysts. The more clinically relevant issue is the degree of compression of adjacent brain parenchyma. In contemporary neurosurgical practice, MRI provides the information necessary to assess the degree of brain compression and mass effect caused by the cyst. As a result, invasive diagnostic studies (e.g., water soluble contrast cisternography) are now infrequently used ***(Rengachary and Kennedy, 1996***).

Historical aspects

Quain is often mistakenly credited with the first description of the disease but neither was he the first to describe the disease nor was the entity that he described an arachnoid cyst ***(Quain., 1855***).

Bright (of "bright's disease" fame) was the first to accurately describe the disease under the title***. "Serous cysts in the arachnoid"*** in his Reports of Medical cases published in 1831. Following this description, the pathogenesis and clinical significance of this entity remained unclear. The numerous terms used over the years to describe it attest to this: chronic cystic arachoniditis, meningitis serosa circumscripta, subdural hygroma, external hydrocephalus, etc. the etiology was variously atritubted to trauma, subarachoid hemorrhage, postmeningitic adhesions, and congenital factors. Confusion surrounded not only the etiology but also the precise anatomic relationship of these cysts to meninges contained series describing these cysts also the fact that clinical example of ependymal, porencephalic, and post infection cysts further attests to the conceptual ambiguity that existed ***(Rengachary and Kennedy, 1996)***.

Robinson, describing a large series of personal cases of middle fossa arachnoid cysts, recognized this entity as a developmental anomaly but postulated (erroneously, in retrospect) that the primary defect is agenesis of the temporal lobe and that there is merely a secondary passive dilation of the subarachnoid space to make up the intracranial volume. He used the term "temporal lobe agenesis syndrome ***(Robinson 1971***).

In a later review, he suggested abandoning the term, recognizing that these cysts indeed represent arachnoid malformations ***(Robinson 1971)*** .

On review of the original description by (***Bright 1831)*** one is struck not only by his lucid description of the lesion but also by his awareness of the fact that these cysts are intra-arachnoid 127 years before Starkman and colleagues came to the same conclusion Even more impressive is the fact the Bright's conclusions were based on observation without the use of a microscope ***(Rengachary and Kennedy, 1996)***.

In 1958, Starkman and colleagues in their classic paper, reported a systematic and critical study of autopsy specimens that included examination of the various parts of such cysts, including the dome, the edges, and the inner wall. They concluded that arachniod cysts evolve from a developmental aberration characterized by splitting and duplication of the arachnoid membrane and that they are truly intra-arachnoid in location ***(Rengachary and Kennedy, 1996)***.

The definition "primary (or true) congenital arachnoid cyst" is applied to collection fluid that develops within the arachnoid membrane because of splitting or duplication of this structure ***(Schachenmayr et al., 1978)***.

The definition is meant to differentiate congenital arachnoid cysts from the "secondary" or "false" arachnoid cysts that have been described in the past with a variety of names, such as "leptomeningeal cysts", "chronic cystic arachnoiditis", and "leptomeningitis chroncial circumscripta adhaesiva seu cystic" ***(Oliver, 1958)***.

These cysts are, in fact, acquired accumulations of cerebrospinal fluid that result from postinflammatory loculation of the subarachnoid space in patients with head injury, intracranial infection, or hemorrhage; their lining membrane is characterized by the presence of inflammatory cells and hemosiderin deposits ***(Winand et al., 1989)***.

Similarly, the presence of glial tissue and epithelial cells differentiates the less common glioependymal cysts, which may also develop (although rarely) within t he subarachnoid space ***(Patrick 1971)***.

As the term cyst defines a closed cavity of abnormal origin, arachnoid cysts should be differentiated from congenital or acquired focal dilations of the subarachnoid space that maintain free communications with the natural pathways of the cerebrospinal fluid circulation, such as dilated cisterns, arachnoid pouches, sacs, and diverticula, as well as the cavities that result from cavitation processes of the brain-namely, the porencephalic cavities and the cavities that occur secondary to vascular infarction ***(Rengachary and Kennedy., 1996)***.

Several aspects of congenital arachnoid cysts have not yet been clarified. First, their differentiation from other intrathecal cystic lesions may be difficult despite the relative ease of recognition presently allowed by the modern diagnostic tools of neuroimaging. Second, the surgical indications as well as the choice of the surgical modalities remain a matter of discussion in many instances, depending on the limited knowledge available on the physiophathogenetic mechanisms and natural history of this specific pathological entity. Further uncertainty is added by thee relative unpredictability of t he surgical outcome in significant proportion of cases ***(Rengachary and Kennedy, 1996)***.

Embryology of meningeal development

The brain and spinal cord are enclosed within membranous structures collectively known as meninges. They are commonly subdivided into pachymeninx (Dura mater) and Leptomeninx (Arachnoidea and pia mater). Anatomical organization and development of these different parts of the meninges show considerably species-dependent differences. Findings in other species cannot be extrapolated to humans without reservation ***(Yasargil, 1986)***.

At Carnegie stage XI (gestational age 23-26; (days) 2.5-4.5mmCR) a single layer of cells is first seen along the lateral aspect of the primitive neural tube. These cells are continuous with and probably derived from the neural crest. This layer participate in the later formation of the intima pia. In stage XII (gestational age 26-30; (days) 3-5mmCR) vascularization begins in tissues around the neural tube and in stage XV (35-38; (days) 7-9mm CR) the neural tube is completely surrounded by developing vessels. At the same time a loose, sparsely cellular area lying between the neural tube, somites and notochord is discernible. This mesoderm derived tissue is called meninx primitive ***(Yasargil, 1986)***.

At stages XVII-XVIII (42-48; (days) 11-17mmCR) vascular channels penetrate the neural tube, carrying cells of the intima pia and meninx primitive as their adventitia. At certain sites the second component of the pia mater, the epipial tissue, forms as a stratified cell layer upon the single cell layer of the intima pia ***(Yasargil, 1986)***.