Neurochirurgie xxx (2014) xxx–xxx

Disponible en ligne sur
ScienceDirect
www.sciencedirect.com

Elsevier Masson France
EM|consulte
www.em-consulte.com

Report 2013: Tumors of the pineal region

Update on the management of pineal cysts: Case series and a review of the literature

Le point sur la prise en charge des kystes pinéaux : série de 26 cas et revue de la littérature

M. Berhouma [a,*], H. Ni [a], V. Delabar [a], N. Tahhan [a], S. Memou Salem [a], C. Mottolese [a], B. Vallee [a,b]

[a] *Department of Neurosurgery B, Pierre-Wertheimer Neurological and Neurosurgical Hospital, hospices civils de Lyon, 59, boulevard Pinel, 69394 Lyon cedex 03, France*
[b] *Research and Education Unit of Medicine, Claude-Bernard University Lyon 1, 59, boulevard Pinel, 69500 Bron, France*

ARTICLE INFO

Article history:
Received 26 March 2013
Accepted 30 August 2013
Available online xxx

Keywords:
Pineal cyst
Pineal gland
MRI
Endoscopy
Natural history
Diagnostic and operative techniques

ABSTRACT

Objective. – The natural history of pineal cysts still remains unclear. Incidental pineal cysts have become more common which raises the question of their management. Symptomatic pineal cysts may require a surgical solution but therapeutic indications have not yet been clearly established.
Method. – From 1986 to 2012, 26 patients with pineal cysts were identified. Their medical records were retrospectively assessed focusing on the initial symptoms, imaging characteristics of the cyst, management strategy, operative technique and their complications, as well as the latest follow-up. A systematic review of the literature is also presented.
Results. – Twenty-six patients with pineal cysts were identified. The mean age was 23.5 years ranging from 7 to 49 years. Symptoms included intracranial hypertension with obstructive hydrocephalus in 18 cases and oculomotor anomalies in 12 cases. Two adult cases presented with non-specific headaches and did not require surgery. Twenty patients were operated via a suboccipital transtentorial approach with total removal of the cyst in 70% of the cases, while the remaining 4 cases were treated with an intraventricular endoscopic marsupialization associating a third ventriculostomy. Four patients required a preoperative ventriculo-peritoneal shunt due to life-threatening obstructive hydrocephalus. Overall, peri-operative mortality was nil. In the two non-operated patients, the cyst remained stable and no recurrences were observed in all operated patients with a mean follow-up of 144 months.
Conclusion. – In the majority of incidental pineal cysts, a clinical and imaging follow-up is sufficient but occasionally not required especially in adults as very rare cases of increase in size have been reported.

Mots clés :
Kyste pinéal
Glande pinéale
IRM
Endoscopie
Histoire naturelle
Diagnostic et techniques opératoires

RÉSUMÉ

Objectif. – L'histoire naturelle des kystes pinéaux est insuffisamment connue. La fréquence croissante des incidentalomes pose la question de leur traitement. D'un autre côté, les indications thérapeutiques des kystes symptomatiques ne sont pas standardisées.
Méthodes. – Entre 1986 et 2012, 26 patients porteurs de kystes pinéaux ont été identifiés. Leurs caractéristiques médicales ont été relevées : symptômes révélateurs, données d'imagerie, traitement, technique chirurgicale et suivi. Une revue exhaustive de la littérature est proposée.
Résultats. – L'âge moyen à la prise en charge était de 23,5 ans (7–49 ans). L'hypertension intracrânienne par hydrocéphalie obstructive constituait le tableau clinique révélateur dans 18 cas et 12 patients présentaient des troubles oculomoteurs. Deux patients adultes avec des céphalées aspécifiques n'ont pas été opérés. Vingt-deux patients furent opérés par une voie sous-occipitale trans-tentorielle avec résection complète du kyste dans 70% des cas, alors que les 4 cas restant ont bénéficié d'une marsupialisation endoscopique avec ventriculocisternostomie endoscopique. Quatre patients ont nécessité une dérivation

* Corresponding author.
E-mail addresses: berhouma.moncef@gmail.com, berhouma.moncef@laposte.net (M. Berhouma).

0028-3770/$ – see front matter © 2014 Published by Elsevier Masson SAS.
http://dx.doi.org/10.1016/j.neuchi.2013.08.010

ventriculopéritonéale préopératoire pour une hydrocéphalie aiguë préopératoire. La mortalité périopératoire est nulle. Le kyste est resté stable chez les deux patients non opérés et aucune récidive n'a été notée au terme des 144 mois de suivi.
Conclusion. – Dans la majorité des cas, une simple surveillance clinique et IRM est suffisante dans le suivi des kystes pinéaux bénins. Seuls de rares cas d'augmentation de taille du kyste ont été rapportés à l'âge adulte.

1. Introduction

Pineal cysts are benign lesions, usually asymptomatic, and discovered as incidentalomas. The prevalence of asymptomatic pineal cysts have increased since the widespread use of magnetic resonance imaging (MRI). Previous studies regarding the prevalence of asymptomatic pineal cysts have reported occurrence ranging from 1 to 10% [1–12]. As the natural history of asymptomatic pineal cysts is not well known, their management strategy is still a matter of debate, ranging from the absence of any follow-up, i.e. either clinical examination or imaging, or regular follow-up and surgical management. In contrast to these incidentalomas, the management of symptomatic pineal cysts is more defined often requiring a surgical approach including endoscopic intraventricular marsupialization, microsurgical resection and sometimes stereotactic aspiration. Based on the description of our case series and a case illustration of a symptomatic pineal cyst treated successfully by a routine endoscopic intraventricular approach, we provide a systematic review of the relevant literature for both asymptomatic and symptomatic pineal cysts.

2. Methods

From 1986 to 2012, 26 patients with pineal cysts admitted to the Department of Neurosurgery of Pierre-Wertheimer University Hospital were identified. All medical records were retrospectively assessed focusing on the initial symptoms, imaging characteristics of the cyst, management strategy, surgical techniques as well as any complications and the most recent follow-up. An exhaustive and systematic review of the literature is presented based on a range of dedicated databases (PubMed, Google Scholar, ScienceDirect and Scopus) with the keywords "pineal", "cyst", "natural history", "endoscopy", "pineal tumours", "pineal apoplexy", "MRI" and "conservative management".

3. Results

Twenty-six patients with pineal cysts were identified. The mean age was 23.5 years (range from 7 to 49 years). There were 16 women and 10 men (sex ratio: 1:6). Symptoms included intracranial hypertension with obstructive hydrocephalus in 18 cases and oculomotor anomalies in 12 cases. Two adult patients presented with non-specific headaches and they did not undergo surgery. Also, Parinaud's syndrome was found in two patients. Twenty patients were operated via a suboccipital transtentorial approach with total removal of the cyst in 70% of cases, while the remaining 4 cases were treated with an intraventricular endoscopic marsupialization associating third ventriculostomy. Among the 20 patients who underwent a craniotomy, 4 required a preoperative ventriculo-peritoneal shunt due to a life-threatening obstructive hydrocephalus. Overall, peri-operative mortality was nil. Postoperative complications included two occipital pseudo-meningoceles requiring depletive spinal taps and four transient hemianopsia. In the two non-operated patients, the cyst remained stable and no recurrences were observed in all operated patients with a mean follow-up of 144 months.

4. Case illustration

A 49-year-old woman with no previous medical history was admitted in emergency due to the rapid onset of a cognitive decline associated with headaches, vomiting and gait disturbances. The patient was conscious but displayed anterograde amnesia and confusion. Her gait was unstable with ataxia and fundoscopy revealed a bilateral papilledema. No oculomotor abnormalities or nystagmus were observed.

Computed tomography scan and MRI findings revealed a 2 cm diameter pineal cyst with a thin enhancing wall and a fluid-fluid level visible on T2 axial sequences suggesting a pineal apoplexy. The cyst was isointense in T1 and hyperintense in T2 sequences (Fig. 1A–C). There was a major hydrocephalus with trans-ependymal resorption secondary to aqueductal stenosis.

The patient underwent complete endoscopic treatment under general anesthesia in a neutral supine position, the 30° lens endoscope was inserted into the right frontal horn with a skull entry point at the junction of the right pupillary line and a para-coronal line 2 cm anterior to the coronal suture. The right Monro foramen was entered and the floor of the third ventricle identified with the infundibular recess ahead and the mammillary bodies posteriorly. The first stage consisted of a third ventriculostomy. Then the endoscope was directed towards the posterior part of the third ventricle to identify the anterior part of the pineal cyst, which was translucent and vascularized. The cyst wall was therefore coagulated with a bipolar cautery and opened with scissors allowing the issue of a yellow reddish content spreading into the third ventricle. A histological specimen was obtained with the forceps and confirmed the diagnosis. The remainder of the cyst wall was adherent to the ependyma and largely coagulated for retraction until the aqueduct was identified. The ventricle was finally washed with warm saline solution and the endoscope removed.

The postoperative course was uneventful and the patient was asymptomatic at one month. The MRI follow-up at 6 months and one year showed the infracentimetric remnant of the cyst (Fig. 2A–C), a patent Sylvius aqueduct with no hydrocephalus and a functional third ventriculostomy (Fig. 3).

5. Discussion

5.1. Epidemiology and natural history

Pineal cysts can be observed at all ages. Their prevalence ranges from 1 to 10% [1,2,11,13–15]. In the largest series, this prevalence was closer to 1% than 10% [1]. The prevalence could increase in the near future due to the widespread use of MRI, and, as mentioned in a reported autopsy series, this prevalence could reach up to 40% [11,16,17]. Al-Holou et al. recently reported the age-related prevalence of pineal cysts both in children and adults [1,2]. In their adult series, they found a prevalence of 1% of pineal cysts out of 48,417 consecutive adult patients (478 pineal cysts). They also observed a peak prevalence at the ages of 19–30 years (2% of prevalence). These authors also showed that the prevalence significantly decreased with advancing age. Finally, they concluded that both younger age and female gender were independent factors of increased

Please cite this article in press as: Berhouma M, et al. Update on the management of pineal cysts: Case series and a review of the literature. Neurochirurgie (2014), http://dx.doi.org/10.1016/j.neuchi.2013.08.010