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Case report

Combined endoscopic and microsurgical removal of a giant cystic craniopharyngioma in a six-year-old boy

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ABSTRACT

Giant cystic craniopharyngiomas are rare lesions whose clinical and surgical management is extremely challenging, often requiring more than one craniotomy before obtaining a satisfying removal. We report one case of a giant cystic craniopharyngioma completely excised with a two-step combined use of a minimally invasive endoscopic approach followed by a single microsurgical transcranial procedure.

A six-year-old boy presented with symptoms of increased intracranial pressure and posterior fossa involvement. Preliminary imaging revealed a large para- and suprasellar cystic tumor bulging superiorly into the third ventricle, and extending posteriorly from the retroclival region into the posterior fossa to the level of the foramen magnum.

The suprasellar cystic quota was initially approached endoscopically through a right precoronal-transventricular approach and the cyst drained, while the remaining tumor was macroscopically totally removed one week later by a right pterional approach.

A combined technique – endoscopic drainage followed by microsurgical removal – can be a valid alternative for the removal of giant cystic craniopharyngiomas in cases in which the cystic compartment bulges within the ventricular spaces, and may avoid multiple craniotomies.

1. Introduction

Craniopharyngiomas are hystologically benign tumors of maldevelopmental origin arising from epithelial remnants of squamous cells of Rathke's pouch stomodeal epithelium, representing approximately 2–4% of all intracranial tumors and 10% of childhood brain tumors [1]. Craniopharyngiomas involve mainly the intra- and suprasellar regions. These tumors may be predominantly cystic (with no significant solid portion), mixed (cystic and solid components), or predominantly solid. Giant cystic craniopharyngiomas may extend to the posterior fossa, reaching the cerebellopontine angle and/or the foramen magnum [1–5]. The ideal treatment for craniopharyngiomas is total removal, although this should be attempted only if compatible with preservation of neural and endocrine function [6,7]. Transcranial microsurgical approaches are still considered the approaches of choice for the removal of suprasellar and intraventricular tumors even if, in the last years, neuroendoscopy has been increasingly used with a wide range of therapeutic effect ranging from surgical excision of intrasellar or suprasellar craniopharyngiomas, using a pure endoscopic extended transsphenoidal approach, to transventricular marsupialization of predominantly cystic craniopharyngiomas [8].

Giant craniopharyngiomas are difficultly removed in a single stage and are associated with a high rate of surgical mortality and morbidity, as well as a high recurrence rate [6,7,9,10]. Many different approaches have been proposed for giant cystic craniopharyngiomas: pterional approach [3,11,12], staged procedures through pterional and suboccipital approaches [1,5], presigmoid transtentorial approaches [13] and anterior transpetrous transtentorial approach [14]. We present a case of a giant cystic craniopharyngioma with a large extension in the posterior fossa, which was totally removed via a two-step surgical procedure, i.e. first endoscopic and than a standard microsurgical approach.

2. Case report

2.1. History and examination

A six-year-old boy presented with a three-week history of headache associated with nausea and vomiting. On examination, the child showed ataxia, horizontal nystagmus, dysphagia and dysphonia. Fundoscopy revealed bilateral papilledema. Blood hormone profile was normal.

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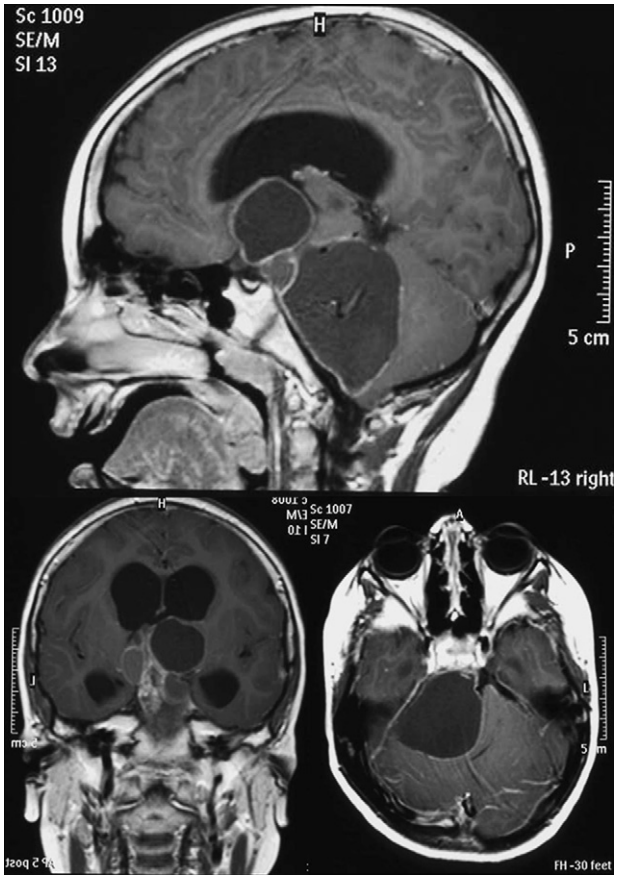

Fig. 1. Preoperative magnetic resonance imaging (MRI) scans showing the presence of a large para- and suprasellar cystic tumor reaching from the retroclival region into the posterior fossa.

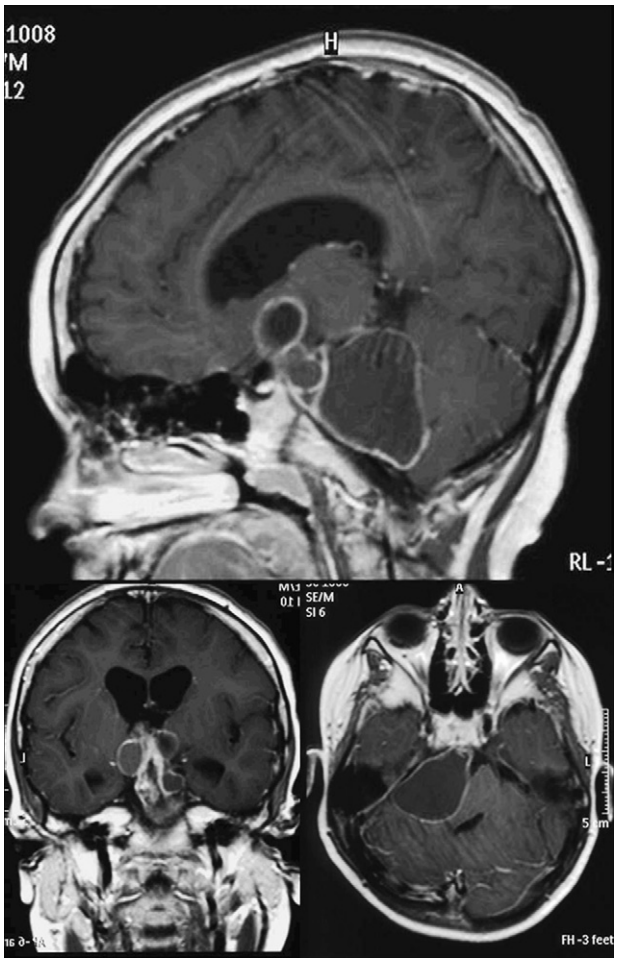

Fig. 2. Magnetic resonance imaging (MRI) scans demonstrating the reduction of the cystic component both in the suprasellar region and in the posterior fossa after the endoscopic procedure.

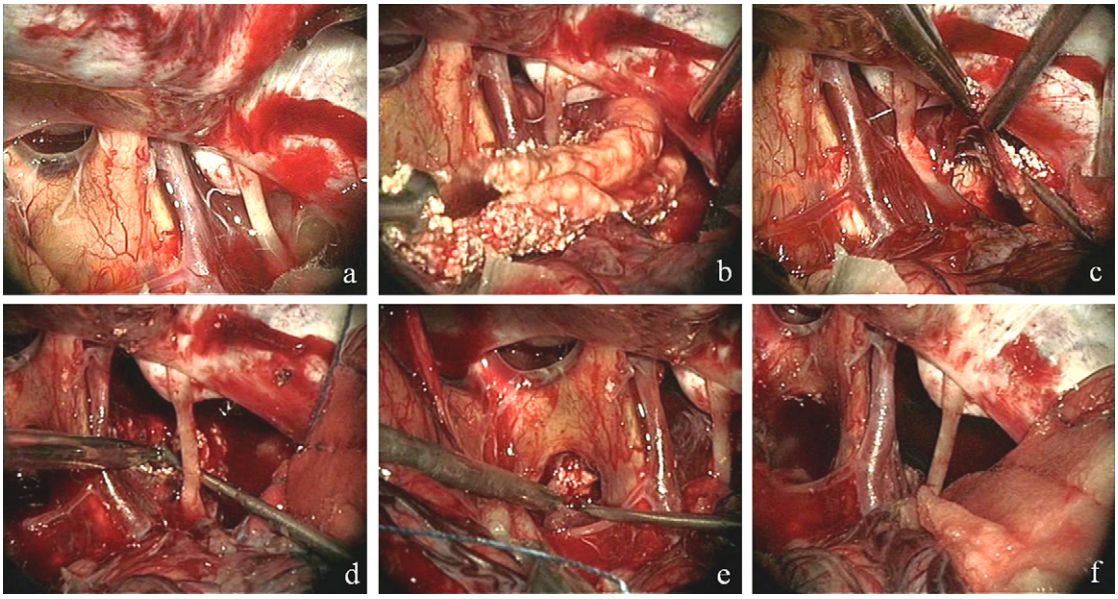

Fig. 3. Intraoperative images. (a) Surgical field before beginning the procedure; (b and c) removal of the portion of the tumor located in the posterior fossa; (d) debulking of the tumor mass between the carotid artery and the oculomotor nerve; (e) intraventricular components removed after opening of the lamina terminalis; (f) surgical field at the end of the procedure.