

ROMANIAN  
NEUROSURGERY

Vol. XXXIV | No. 3      September 2020

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# Craniopharyngioma and arteriovenous malformation operated using the same craniotomy. An unusual case

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## ABSTRACT

Craniopharyngiomas (CPs) are rare benign epithelial tumours. Brain arteriovenous malformations (AVMs) are also rare lesions occurring in young adults. The appearance of both these lesions in the same patient is rare. A 42-year-old patient presented with headaches for 3 months and a progressive decrease in his visual acuity. Bitemporal hemianopsia was detected in the visual field. Magnetic resonance imaging (MRI) revealed a tumour with cystic and solid components located in the suprasellar region and AVM in the right temporal lobe. AVM (Martin-Spetzler grade III) was visualised using digital subtract angiography (DSA), which was fed from the right middle cerebral artery and drained through the sigmoid sinus via the inferior petrosal sinus. The patient was operated with enlarged right frontotemporal craniotomy. AVM nidus was totally removed at the first operation. Embolisation was not preferred before the AVM surgery. After 3 days, sylvian dissection was performed using the same craniotomy. The tumour was completely removed via the carotid cistern by making sharp dissection from the infundibulum. Post-operatively, the patient showed normal neurological examination and significant improvement in his visual field examination. There was no residual/recurrent tumour or AVM on contrast-enhanced MRI and DSA at post-operative 6 months. Histopathological examination revealed AVM in the first operative material and papillary-type CP in the second. The coexistence of these two rare pathologies has previously been reported in only one patient. This is the first case of surgical resection of CP and AVM using the same craniotomy.

## INTRODUCTION

Craniopharyngiomas (CPs) are rare benign epithelial tumours with slow growth. They account for 2%–5% of intracranial tumour primers, with an incidence of 0.5–2.0 billion per year.<sup>7</sup> The bimodal age distribution peaks between 0 and 19 and 40 and 79 years of age.<sup>20</sup> While complete improvement can be achieved with radical resection of CP, high recurrence rates have been reported for partial resection.<sup>1</sup>

Brain arteriovenous malformations (AVMs) are also rare lesions that usually occur in young adults. The prevalence of AVMs in the population is 10–18 per 100,000 adults.<sup>11</sup> Morphologically, in AVM, a vascular mass can be observed, with direct blood flow between the arterial and venous

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**Keywords**  
arteriovenous malformation,  
AVM,  
craniopharyngioma

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ISSN online 2344-4959  
© Romanian Society of  
Neurosurgery



First published  
September 2020 by  
London Academic Publishing  
[www.lapub.co.uk](http://www.lapub.co.uk)

circulation without a true capillary bed. Generally, the high current feeder consists of the arteries, nidus and drainage veins.<sup>3</sup>

The coexistence of these two rare pathologies has previously been detected using radiology in only one patient. However, we presented the first case of simultaneous occurrence of CP and AVM in the same patient who was then treated with surgical resection.

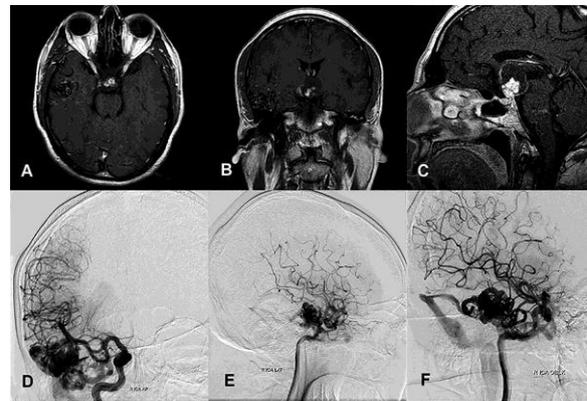
#### CASE REPORT

A 42-year-old patient was referred to our clinic with persistent headache for 3 months and a time-lapse decrease in visual acuity. On magnetic resonance imaging (MRI), a 32 × 26 × 24-mm tumour with cystic and solid components extending from the supracellular region to the third ventricle was detected. At the same time, AVM of size 36 × 28 × 24 mm was observed in the right temporal lobe. AVM (Martin-Spetzler grade III) was seen in the patient's brain digital subtract angiography (DSA), which was fed from the right mild cerebral artery (MCA), with no aneurysm in the nidus, and sigmoid sinus was drained via the inferior petrosal sinus (Figure 1). Bitemporal hemianopsia was detected in the visual field of the patient. The patient then underwent an enlarged right frontotemporal craniotomy. The AVM nidus on the temporal basal was totally removed. Embolisation was not preferred before the surgery. As the duration of the first operation was relatively long, it was decided to perform CP surgery in the next session. After 3 days, sylvian dissection was performed using the same craniotomy. The cyst was aspirated by passing through the carotid cistern. The tumour was totally removed by making a sharp dissection from the infundibulum. After the surgeries, the patient was placed in intensive care for 1 day. His general condition was good, and his neurological examination was normal. However, diabetes insipidus developed after CP surgery. Nasal desmopressin therapy was therefore started but was discontinued after 1 month. The patient showed normal levels of pituitary hormones and no hypothalamic obesity. An improvement was noted in his visual field examination. There was no evidence of residual/recurrent tumour or AVM on contrast-enhanced MRI and DSA during the early post-operative period and at 6-month follow-up (Figure 2).

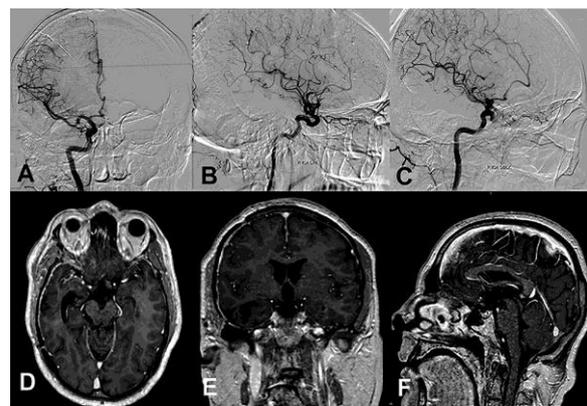
#### Histopathological examination

The resected tissue was fixed in 10% formalin, embedded in paraffin and stained with

haematoxylin-eosin (H&E). In the first surgical material, vessels of different sizes and a lesion composed of the gliotic brain tissue was detected. Some of the existing vessels consisted of small-sized, thick-walled arteries, whereas others included larger lumens surrounded by hyalinised veins. These findings were compatible with those of AVM. The second operative material showed a well-differentiated, non-keratinised squamous epithelium papillary-type CP (pCP) around a partially oedematous fibrovascular stroma. Calcification was not observed (Figure 3).

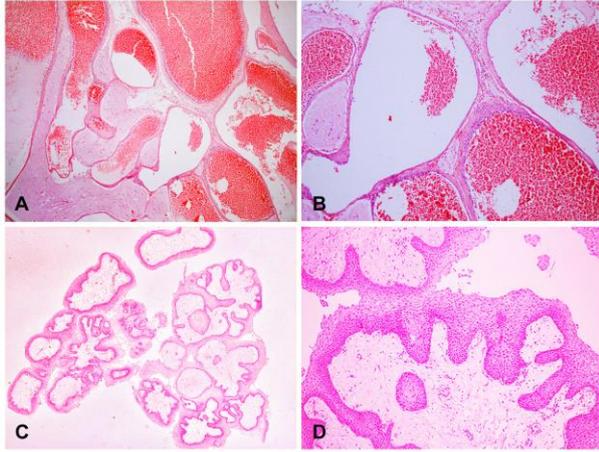


**Figure 1.** (a) Axial, (b) coronal contrast T1 and (c) sagittal T2 MRI sections showing cystic tumour in the midline, extending to the third ventricle, and a right temporal AVM. (d) A-P, (e) lateral (f) and oblique right internal carotid artery DSA images showing AVM from the right MCA, with no aneurysm in the nidus, and drainage of the sigmoid sinus via the inferior petrosal sinus. AVM, arteriovenous malformation; DSA, digital subtract angiography; MCA, mild cerebral artery; MRI, magnetic resonance imaging.



**Figure 2.** Post-operative (a) A-P, (b) lateral (c) and oblique right internal carotid DSA images. No AVM can be seen. Post-operative (a) axial, (b) coronal (c) and sagittal contrast T1 MR images showing total resection of both the tumour and AVM.

AVM, arteriovenous malformation; DSA, digital subtract angiography.



**Figure 3.** (a) Malformed, randomly distributed vessels, both in the veins and arteries, separated by variable amounts of intervening brain parenchyma (H&E,  $\times 40$ ). (b) H&E,  $\times 100$ . and (c) papillary, cauliflower-like appearance, with surface epithelium covering the fibrovascular cores (H&E,  $\times 20$ ). (d) Non-keratinising, well-differentiated squamous epithelium surrounding the fibrovascular stromal cores (H&E,  $\times 100$ ). H&E, haematoxylin and eosin staining.

## DISCUSSION

Vascular malformations of the abnormal arteries and veins are usually congenital. They can appear at any age but are often diagnosed between 20 and 40 years of age. The clinical symptoms of AVM depend on its location. The most common symptoms include headache and seizures, but at least 15% of patients are asymptomatic. Every year, more than half of AVM cases are referenced with intracranial haemorrhages, constituting to approximately 2% of the total haemorrhagic stroke cases.<sup>2,6,8</sup> The methods used for the specific treatment of AVM include endovascular embolisation and surgery or gamma knife treatment.<sup>4</sup> In this study, we preferred to employ only surgical resection as the treatment approach.

It is assumed that CPs are histologically of two subtypes: adamantinomatous (aCP) and papillary (pCP). aCP is the more common type and is characterised by necrotic debris, cystic and/or solid components, fibrous tissues and calcification (more frequent in children). Infiltration is also common in the neighbouring brain tissue. pCP mostly occurs in adults, with 14%–50% of adult cases and only 2% of paediatric cases. pCP is characterised by the

presence of a solid component or a combination of solid and cystic components. The cystic content is usually yellow and viscous, with rare calcification. pCP is generally well-defined, and infiltration into the neoplastic epithelium of the neighbouring brain tissue is minimal or even absent.<sup>10, 13</sup> The patient's pathological findings indicated pCP, and its dissection from the surrounding tissues was easy during the surgery.

The clinical symptoms of CPs are related to hypothalamic/pituitary deficiencies, visual impairment and increased intracranial pressure. The preferred treatment is complete resection which is carefully performed so as to preserve the optic and hypothalamic functions.<sup>15</sup> Post-operatively, in our patient, visual impairment decreased and hypothalamic dysfunction was not observed. In their meta-analysis, Dandurand et al.<sup>7</sup> compared the gross total resection of CPs with subtotal resection followed by adjuvant radiotherapy and found that the lowest recurrence rate was achieved with gross total resection. In our patient, gross total resection of CP was performed, and no recurrence was observed at the first 6-month follow-up.

Endoscopic trans-sphenoidal approach is the most preferred method for CP surgeries, which involves a pterional approach.<sup>18</sup> We prefer endoscopic trans-sphenoidal approach only if the patient is a candidate for CP surgery. However, an extended pterional approach was preferred in this case, considering that both the lesions could be resected using a single craniotomy flap.

Whether there exists any association between the formation of vascular malformations and development of intracranial meningiomas needs to be discussed. Rare cases of vascular malformations with primary brain tumours have been previously reported. In the literature, AVM has been reported mostly in association with oligodendrogliomas and astrocytomas.<sup>12,17,19</sup> Pleomorphic xanthoastrocytoma<sup>16</sup>, meningioma<sup>9</sup> and ganglioneuroma<sup>5</sup> have also been reported with AVM. Mori et al.<sup>14</sup> were the first to report AVM and CP in patients, which were detected radiologically. However, these patients had only CP resections and were following AVM. Our patient is the first case in whom both the lesions were completely excised.

The occurrence of primary brain tumour in association with cerebral AVM has been reported.

However, the simultaneous occurrence of CP and AVM in a patient is extremely rare.

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