




# Papillary craniopharyngioma coexisting with an intratumoral abscess in a pediatric patient: A case report and review of the literature

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## Abstract

Craniopharyngiomas are benign neoplasms with two histological subtypes: adamantinomatous and papillary. Papillary craniopharyngiomas are rare in children, and those with a pituitary abscess within are even rarer. Herein, we present the case of a 14-year-old boy with a papillary craniopharyngioma and a coexisting intratumoral abscess, who was hospitalized for persistent pyrexia, polyuria, and polydipsia. The absence of calcification on computed tomography, high signal intensity inside the tumor on diffusion-weighted imaging, and clinical findings such as fever, a high inflammatory response, and meningitis, as well as short-term morphological changes on imaging, could aid in diagnosis.

## Keywords

Craniopharyngioma, papillary craniopharyngioma, adamantinomatous craniopharyngioma, pituitary abscess, diffusion-weighted imaging

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## Introduction

Craniopharyngiomas are benign neoplasms (World Health Organization [WHO] grade I) that typically arise in the sellar to suprasellar regions.<sup>1</sup> They account for approximately 1–5% of the primary brain tumors in patients of all ages.<sup>1</sup> In Japan, craniopharyngiomas are the fourth most common (10.6%) pediatric brain tumors.<sup>2</sup> They have two histological subtypes<sup>1</sup>: the adamantinomatous subtype accounts for most pediatric cases, while the papillary subtype is diagnosed almost exclusively in adults<sup>3</sup> and is rare in children.<sup>4</sup>

Pituitary abscesses are rare<sup>5</sup> and are either primary (developing *de novo* inside a normal pituitary gland) or secondary (forming within a diseased gland, such as a

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pituitary adenoma and a Rathke's cleft cyst).<sup>6</sup> Secondary pituitary abscesses are hypothesized to be caused by tumors that disrupt the local blood circulation and impair the immune response, which in turn disrupt the control of infection. Furthermore, these tumors may have necrotic areas, which can make them more susceptible to infections.<sup>7</sup> While few adult cases presenting with an abscess inside a craniopharyngioma have been reported,<sup>5,6,8–12</sup> no pediatric case has been reported yet.

Herein, we report a case of a pediatric patient with a histopathologically confirmed papillary craniopharyngioma with a coexisting intratumoral abscess. We demonstrate its rarity through a review of the literature and compare the radiological findings of papillary craniopharyngiomas and pituitary abscesses between children and adults.

### Case report

A 14-year-old boy was referred to our hospital for a 2-month history of polydipsia and polyuria; for 1 month before presentation, he had experienced persistent fever, worsening polyuria, and worsened polydipsia causing nocturnal urination and insomnia. He was diagnosed with diabetes insipidus. The clinical symptom comprised bitemporal hemianopia, with no other obvious neurological abnormalities. While he had a history of treatment for frontal sinusitis, there was no relevant family medical history.

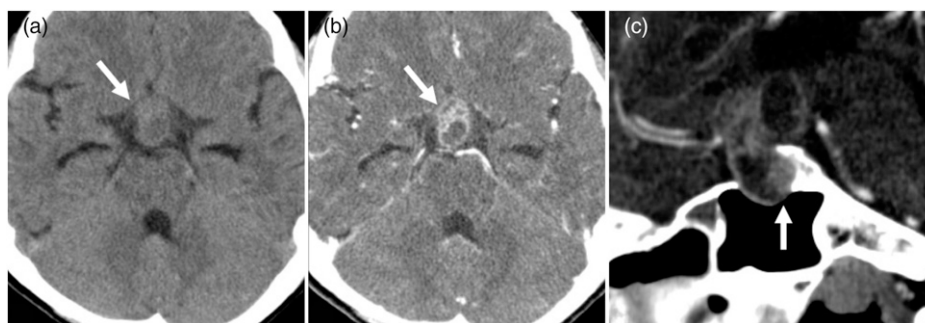
Computed tomography (CT) revealed a  $31 \times 22 \times 20$ -mm heterogeneously enhancing mass extending from the sella turcica to the suprasellar region. The sella turcica was slightly enlarged by the mass, and its floor had thinned. No calcification was observed in the mass (Fig. 1). He had sinusitis in his frontal and maxillary sinuses (not shown), but his sphenoidal sinus was clear (Fig. 1).

Magnetic resonance imaging (MRI) identified a  $29 \times 25 \times 21$ -mm heterogeneously enhancing mass with cystic components mainly in the sellar to suprasellar regions (Fig. 2). T1-weighted imaging (T1WI) and T2-weighted imaging (T2WI) revealed a mass with heterogeneous signals (mixed

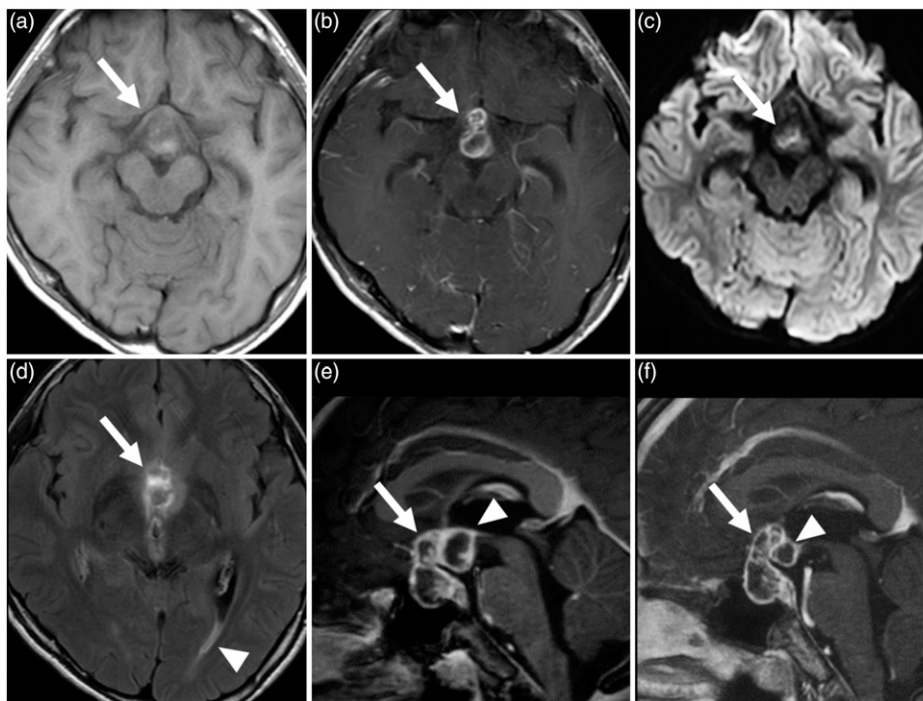
high and low intensity signals). The optic chiasm was squeezed upward and thinned due to the mass. Post-contrast fluid-attenuated inversion recovery images showed wall enhancement in the left lateral ventricle and the third and fourth ventricles, indicating ventriculitis. Hydrocephalus was also observed. Diffusion-weighted imaging (DWI) showed a partially high signal intensity area in the mass. The minimum apparent diffusion coefficient (ADC) in the cystic area was  $0.66 \times 10^{-3} \text{ mm}^2/\text{s}$ . Follow-up MRI revealed an obvious increase in size after 2 weeks. Germinoma, Rathke's cleft cyst, pituitary adenoma, pituitary xanthogranuloma, and Langerhans cell histiocytosis were considered as the differential diagnoses. However, because this case was of a tumor with a cystic and solid component, craniopharyngioma was suspected from the high frequency. Based on the restricted diffusion and short-term increase in size, we diagnosed the patient with an abscess within the craniopharyngioma. However, due to the patient's young age, we expected an adamantinomatous type and not a papillary type.

An emergency surgery was performed via the trans-sphenoidal approach for the parasellar tumor with an abscess. The dura mater of the sella turcica was incised, and pus was drained from the tumor. A papillary solid component was observed in the deep part; this was biopsied, and histopathological analysis confirmed the diagnosis of a papillary craniopharyngioma. Due to the presence of meningitis and ventriculitis, the initial surgery was limited to abscess drainage and biopsy to avoid surgical adhesions and cerebrospinal fluid leakage. After the inflammation was improved with antibiotic treatment for 1 month, the residual tumor was totally resected during a second surgery.

Histopathological findings revealed a squamous epithelium abutting a fibrocollagenous stroma arranged in a sheet pattern (Fig. 3). Neither wet keratin nor calcification was observed. Immunohistochemistry revealed positive staining for the B-Raf proto-oncogene, serine/threonine kinase (*BRAF*) V600E, but not for nuclear translocation of  $\beta$ -catenin. Immunopositivity for Ki-67 in the squamous



**Fig. 1.** Non-contrast axial CT reveals the cystic mass in the suprasellar region. There is no calcification in the tumor (a; arrow). After contrast administration, an irregular ring-like enhancement is seen (b; arrow). Contrast-enhanced sagittal CT reveals that the sella turcica is slightly enlarged by the mass and its floor is thinned (c; arrow). CT: computed tomography.



**Fig. 2.** On non-contrast axial T1WI, the mass shows a heterogeneous signal intensity. The optic chiasm is squeezed upward and thinned due to the effect of the mass (a; arrow). On contrast-enhanced axial T1WI, the mass shows an irregular ring-like enhancement (b; arrow). On DWI with  $b = 1000 \text{ s/mm}^2$ , the mass shows a high signal intensity (c; arrow), and the ADC value of this area is  $0.66 \times 10^{-3} \text{ mm}^2/\text{s}$  (image not shown). Contrast-enhanced FLAIR image shows wall enhancement of the 3rd (d; arrow) and left lateral ventricles (d; arrowhead), indicating ventriculitis. Contrast-enhanced sagittal T1WI reveals a heterogeneous enhancing mass with cystic components mainly in the sellar to suprasellar regions (e and f; arrows). A dorsal cystic component becomes more enlarged (e; arrowhead) as compared to 2 weeks earlier (f; arrowhead). ADC: apparent diffusion coefficient; DWI: diffusion-weighted imaging; FLAIR: fluid-attenuated inversion recovery; T1WI: T1-weighted imaging.

cells was largely restricted to the basal layer. No obvious abscess was noted due to the abscess drainage and antibiotic treatment. These features were compatible with the diagnosis of a papillary craniopharyngioma.

No apparent tumor recurrence or central nervous system infection was observed during follow-up MRI after 1 year. Hormone replacement therapy was administered due to hypopituitarism.

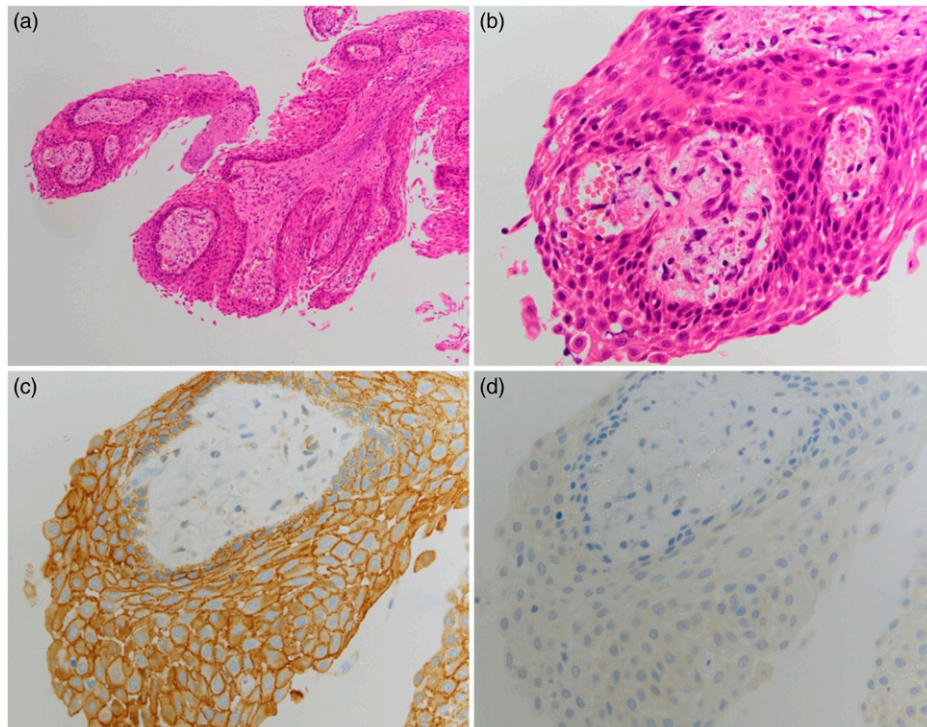
## Discussion

Papillary craniopharyngiomas are associated with the *BRAF* V600E mutation, which may cause tumorigenesis; however, there is no explanation on why this mutation is mainly detected in adult-onset papillary craniopharyngiomas.<sup>13</sup> In adults, these craniopharyngiomas usually occur in the upper saddle area and present as solid or predominantly solid augmented masses without calcification.<sup>13</sup> Conversely, papillary craniopharyngiomas are extremely rare in children.<sup>4</sup> To the best of our knowledge, this is the first reported pediatric case of a papillary craniopharyngioma with a coexisting intratumoral abscess. In this case, the craniopharyngioma

was a heterogeneous augmented mass containing a cystic component without calcification, mainly extending from the saddle to the upper saddle area. Previous studies revealed only one pediatric case (2%) among 48 cases<sup>3</sup> and only two pediatric cases (1.1%) among 189 cases of craniopharyngiomas.<sup>14</sup> Our literature search identified 24 pediatric cases of papillary craniopharyngiomas (ages: 3–14 years), with none presenting with coexisting intratumoral abscesses<sup>3,4,14–18</sup> (Table 1); there was a mild predilection for boys (3/5, 60%).

The association of an abscess with a craniopharyngioma is extremely rare, and only seven cases have been described previously (all adults, age: 26–46 years; Table 2).<sup>5,6,8–12</sup> Thus, our patient is the first reported pediatric case presenting with an abscess inside the craniopharyngioma.

Because papillary craniopharyngiomas are rare in children, we will discuss the imaging findings of craniopharyngiomas in adults. An analysis of 32 reported cases of papillary craniopharyngiomas<sup>3,19</sup> revealed that 29 (91%), 2 (6%), and 1 (3%) tumors were located in the suprasellar region, intrasellar to suprasellar region, and third ventricle, respectively. Around 41%, 41%, and 18% of the tumors



**Fig. 3.** H&E staining section shows a squamous epithelium abutting on a fibrocollagenous stroma arranged in a sheet pattern. Neither wet keratin nor calcification is present (a, b). On immunohistochemistry,  $\beta$ -catenin (c) is negative for nuclear translocation, but *BRAF* V600E is positive (d). No obvious abscess is noted because the specimen has been collected after abscess drainage and antibacterial treatment. *BRAF*: B-Raf proto-oncogene, serine/threonine kinase; H&E: hematoxylin and eosin.

**Table I.** Previous cases of pediatric papillary craniopharyngioma published in the literature.

Report (yr) [ref]	Number of cases	Age (yr)/sex (M/F)	Molecular confirmation	CT findings (shape/Cal)	MR findings (T1WI/T2WI/DWI/CE)
Present case (2021)	1	14/M	<i>BRAF</i> V600E	Cystic and solid/no cal	Low/high/high/enhancing
Borrill et al. (2019) <sup>4</sup>	1	4/F	<i>BRAF</i> V600E	Cystic and solid/no cal	Low/high/NA/enhancing
Schlaffer et al. (2018) <sup>13</sup>	1	6/NR	<i>BRAF</i> V600E	NA	NA
Tariq et al. (2017) <sup>14</sup>	2	3/M	NA	NA	NA
Cheng et al. (2016) <sup>15</sup>	16	NA/NA	NA	NA	NA
Brastianos et al. (2014) <sup>16</sup>	1	9/F	NA	NA	NA
Zhang et al. (2002) <sup>12</sup>	2	NA/NA	NA	NA	NA
Crotty et al. (1995) <sup>3</sup>	1	10/M	NA	NA	NA

*BRAF*: B-Raf proto-oncogene, serine/threonine kinase; Cal: calcification; CE: contrast enhancement; DWI: diffusion-weighted imaging; F: female; M: male; NA: not available; ref: reference; T1WI: T1-weighted imaging; T2WI: T2-weighted imaging; yr: year.

were solid, solid and cystic, and cystic, respectively. On non-contrast (NC) T1WI, the solid tumor parts showed iso- or slightly low signal intensity relative to that of the gray matter. On T2WI, the solid tumor parts showed an inhomogeneous, but high signal intensity. The signal intensity of the rare cystic tumor parts was mostly low on NC-T1WI and high on T2WI. A thin peripheral contrast-enhancing rim was visible on contrast-enhanced (CE) T1WI. Calcifications

are the key imaging findings because no<sup>3</sup> or rare (<1%)<sup>19</sup> calcifications are seen in papillary craniopharyngiomas; conversely, the typical adamantinomatous craniopharyngiomas can be summarized by the “90% rule”: approximately 90% of the tumors are predominantly cystic, show more or less prominent calcifications, and show enhancement in the cyst walls.<sup>13</sup> In summary, a papillary craniopharyngioma is typically located in the suprasellar region and

**Table 2.** Previously published cases of an abscess inside a craniopharyngioma.

Report (yr) [ref]	Age (yr)/sex (M/F)	Culture	CT findings (shape/Cal)	MR findings (T1WI/T2WI/DWI/CE)
Present case (2021)	14/M	Not identified	Cystic and solid/no cal	Low/high/high/enhancing
Bhaisora et al. (2018) <sup>5</sup>	38/F	Methicillin-sensitive coagulase-negative <i>Staphylococcus aureus</i>	Cystic/NA	High/Low/NA/enhancing
Qi et al. (2009) <sup>6</sup>	26/M	<i>Staphylococcus aureus</i>	Cystic/NA	Low/NA/NA/enhancing
Shanley et al. (1994) <sup>7</sup>	28/F	<i>Salmonella typhi</i>	Cystic and solid/no cal	Low/NA/NA/enhancing
Arseni et al. (1975) <sup>8</sup>	31/M	<i>Escherichia coli</i>	NA	NA
Obrador et al. (1972) <sup>9</sup>	34/M	Not identified	NA	NA
Montrieu et al. (1965) <sup>10</sup>	45/M	Not identified	NA	NA
Riser et al. (1956) <sup>11</sup>	46/F	Not identified	NA	NA

Cal: calcification; CE: contrast enhancement; DWI: diffusion-weighted imaging; F: female; M: male; NA: not available; ref: reference; T1WI: T1-weighted imaging; T2WI: T2-weighted imaging; yr: year.

usually presents as a solid or predominantly solid enhancing mass without calcification in adult patients. In our patient's case, there was no calcification in the tumor; however, the tumor components showed an irregular enhancing cystic mass. The reason for this finding was thought to be the effect of the intratumoral abscess.

Pituitary abscesses are classified as either primary (or *de novo*) and secondary (arising within preexisting pituitary lesions such as pituitary adenomas and craniopharyngiomas).<sup>20</sup> Secondary pituitary abscesses are hypothesized to be caused by tumors that disrupt the local blood circulation and impair the immune response, which in turn, disrupt the control of infection.<sup>21</sup> Furthermore, these tumors may have areas of necrosis, which can make them more susceptible to infection.<sup>21</sup> A retrospective analysis of the MRI features of 51 cases with pituitary abscesses by Wang et al.<sup>22</sup> revealed specific radiological features on MRI, including iso- or low signal intensity on T1WI (58.8%), iso- or high signal intensity on T2WI (76.5%), and the disappearance of the posterior pituitary bright spot in most cases; these findings are consistent with those of the present case. Furthermore, they observed rim or rim-like enhancement after contrast material injection in 82.4% of the patients; half of these showed typical rim enhancement, while the remaining half showed atypical rim enhancement with special signs, such as an enhancing, thick abscess wall and high intensity flocculent or cotton-like foci within the internal low signal intensity region. Other features described are evidence of pituitary stalk thickening and leptomeningeal enhancement.<sup>23</sup> Our patient showed a thick and irregular rim-like enhancement in the periphery of the lesion with ventricular wall enhancement on CE-T1WI. The pituitary stalk and pituitary gland could not be visualized.

DWI is widely used to differentiate cerebral abscesses from other necrotic masses.<sup>24</sup> Brain abscesses typically give a high intensity signal on DWI, with a decreased ADC value in the central region. However, the high signal intensity on

DWI is not specific to pituitary abscesses, because pituitary apoplexy can also exhibit a high signal intensity on DWI, but with normal to high ADC values due to the T2 shine-through effect.<sup>25</sup> The accuracy of DWI in pituitary abscess identification remains controversial; however, clinical findings such as fever, a high inflammatory response, and meningitis, as well as short-term morphological changes on imaging, may help in accurate diagnosis.

In conclusion, we have reported a case of a teenager with a papillary craniopharyngioma coexisting with an intratumoral abscess. Papillary craniopharyngioma is located in the suprasellar region and is usually a solid or predominantly solid enhancing mass without calcification. Pituitary abscesses show high signal intensity on DWI, with a decreased ADC value in the central region, as well as rim-like enhancement. Clinical findings (fever, high inflammatory response, and meningitis) as well as short-term morphological changes on imaging could also be key to an accurate diagnosis.

#### Author contributions

Katsuhiro Takagi, Kazufumi Kikuchi, Akio Hiwatashi, Osamu Togao, and Kousei Ishigami drafted the manuscript and designed the figures. Yuhei Sangatsuda, Daisuke Kuga, and Masahiro Mizoguchi edited the surgical part. Hidetaka Yamamoto and Toru Iwaki edited the pathological part. All authors discussed the results and contributed to the final manuscript.

#### Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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### References

- Louis DN, Ohgaki H, Wiestler OD, et al. WHO classification of tumours of the central nervous system. 4th ed. Lyon: IARC Press, 2016.
- Brain Tumor Registry of Japan (2005-2008). *Neurol Med Chir (Tokyo)* 2017;57:9–102.
- Crotty TB, Scheithauer BW, Young WF Jr, et al. Papillary craniopharyngioma: a clinicopathological study of 48 cases. *J Neurosurg* 1995;83:206–214.
- Borrill R, Cheesman E, Stivaros S, et al. Papillary craniopharyngioma in a 4-year-old girl with BRAF V600E mutation: a case report and review of the literature. *Childs Nerv Syst* 2019;35:169–173.
- Bhaisora KS, Prasad SN, Das KK, et al. Abscess inside craniopharyngioma: diagnostic and management implications. *BMJ Case Rep* 2018;2018:bcr201722304.
- Qi S, Peng J, Pan J, et al. Secondary abscess arising in a craniopharyngioma. *J Clin Neurosci* 2009;16:1667–1669.
- Hanel RA, Koerbel A, Prevedello DM-S, et al. Primary pituitary abscess: case report. *Arq Neuropsiquiatr* 2002;60:861–865.
- Shanley DJ, Holmes SM. Salmonella typhi abscess in a craniopharyngioma: CT and MRI. *Neuroradiology* 1994;36:35–36.
- Arseni C, Dănăilă L, Carp N, et al. Intrasellar abscess. *Neurochirurgia (Stuttg)* 1975;18:207–213.
- Obrador S, Blazquez MG. Pituitary abscess in a craniopharyngioma. Case report. *J Neurosurg* 1972;36:785–789.
- Montrieux B, Janny P, Pignide L, et al. Observations on hypophyseal abscesses. *Neurochirurgie* 1965;11:366–371.
- Riser M, Lazorthes G, Anduzecher H. Hypophysial abscesses. *Rev Otonéuroophthalmol* 1956;28:494–496.
- Müller HL. The diagnosis and treatment of craniopharyngioma. *Neuroendocrinology* 2020;110:753–766.
- Zhang Y-Q, Wang C-C, Ma Z-Y. Pediatric craniopharyngiomas: clinicomorphological study of 189 cases. *Pediatr Neurosurg* 2002;36:80–84.
- Schlaffner S-M, Buchfelder M, Stoehr R, et al. Rathke's cleft cyst as origin of a pediatric papillary craniopharyngioma. *Front Genet* 2018;9:49.
- Tariq MU, Din NU, Ahmad Z, et al. Papillary craniopharyngioma: a clinicopathologic study of a rare entity from a major tertiary care center in Pakistan. *Neurol India* 2017;65:570–576.
- Cheng J, Shao Q, Pan Z, et al. Analysis and long-term follow-up of the surgical treatment of children with craniopharyngioma. *J Craniofac Surg* 2016;27:e763–e766.
- Brastianos PK, Taylor-Weiner A, Manley PE, et al. Exome sequencing identifies BRAF mutations in papillary craniopharyngiomas. *Nat Genet* 2014;46:161–165.
- Sartoretti-Schefer S, Wichmann W, Aguzzi A, et al. MR differentiation of adamantinous and squamous-papillary craniopharyngiomas. *AJNR Am J Neuroradiol* 1997;18:77–87.
- Wolansky LJ, Gallagher JD, Heary RF, et al. MRI of pituitary abscess: two cases and review of the literature. *Neuroradiology* 1997;39:499–503.
- Agyei JO, Lipinski LJ, Leonardo J. Case report of a primary pituitary abscess and systematic literature review of pituitary abscess with a focus on patient outcomes. *World Neurosurg* 2017;101:76–92.
- Wang Z, Gao L, Zhou X, et al. Magnetic resonance imaging characteristics of pituitary abscess: a review of 51 cases. *World Neurosurg* 2018;114:e900–e912.
- Kuge A, Sato S, Takemura S, et al. Abscess formation associated with pituitary adenoma: a case report: changes in the MRI appearance of pituitary adenoma before and after abscess formation. *Surg Neurol Int* 2011;2:3.
- Xu X-X, Li B, Yang H-F, et al. Can diffusion-weighted imaging be used to differentiate brain abscess from other ring-enhancing brain lesions? A meta-analysis. *Clin Radiol* 2014;69:909–915.
- Burdette JH, Elster AD, Ricci PE. Acute cerebral infarction: quantification of spin-density and T2 shine-through phenomena on diffusion-weighted MR images. *Radiology* 1999;212:333–339.