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CASE REPORT

# A middle cerebral artery ischemic stroke occurring in a child with a large prolactinoma

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**Abstract**
Pituitary adenomas are rare in children, and often present with symptoms of headache, nausea or emesis, visual disturbance, or hormonal hypersecretion. With large tumors, mass effect from the lesion can lead to severe endocrinopathy and compression of intracranial neurovascular structures. In this case report, we describe an unusual presentation of an ischemic stroke in the territory of the right middle cerebral artery resulting from a prolactin-secreting macroadenoma. The patient's primary symptoms were headache, left facial droop, and left hemibody weakness. She was successfully managed with cabergoline, a dopamine agonist, with a reduction in the size of the tumor and normalization of serum prolactin levels. She remained clinically stable throughout her hospitalization, and was safely discharged without surgical intervention. In her recent 2-year follow-up, her tumor and prolactin levels were stable and she had dramatic improvements in her left-sided muscle strength.

**Keywords** Pituitary adenoma · MCA · Infarct

## Introduction

In general, sellar and suprasellar tumors in children are uncommon with craniopharyngiomas being the most common type arising in this location. Pituitary adenomas are the next most common, but overall are rare [1, 2]. Intrasellar tumors typically present with slow onset of symptoms, either from endocrine deficiency or mass effect leading to compression of the optic pathways. Visual disturbance is the most common presenting symptom, followed by non-specific symptoms such as headache, nausea, or vomiting. With pituitary apoplexy, there is usually a sudden onset of severe headache, oculomotor palsy, and visual loss [3].

Ischemic stroke is an exceptionally rare presentation for intrasellar tumors, and most reported cases within the literature have occurred in the context of apoplexy, as intratumoral hemorrhage can lead to rapid enlargement of the primary mass with subsequent compression of intracranial vessels. Mass effect from the lesion can also induce cerebral vasospasm, which can increase stroke risk [3].

Fewer than thirty cases of ischemic stroke caused by pituitary tumors have been reported; among these, only two have been described in children [4]. One patient was treated with prompt surgical intervention to relieve mechanical compression, while the other was managed medically as the clinicians hypothesized that cerebral vasospasm was the etiology of the infarct [4, 5]. We present a case of a teenage girl who presented with acute onset of left-sided weakness secondary to an ischemic stroke in the territory of the right middle cerebral artery (MCA) presumed to be caused by direct compression from a macroprolactinoma; of note, she did not have pituitary apoplexy. Because of her stable clinical status, the patient was treated with a dopamine agonist. At time of last follow-up, her serum prolactin levels had normalized with a corresponding dramatic reduction in the size of the tumor.

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## Case report

### Clinical findings

Patient IL is a 17-year-old female who presented to the emergency room with a 2-week history of left upper extremity weakness and left facial droop. Prior to these symptoms, she reported a 1-year history of nocturnal headaches, occasionally associated with emesis. She had previously been diagnosed with migraines, and her headaches had been presumptively treated with naproxen. An initial head CT scan revealed a large 4.5 × 4.5 cm, multi-lobulated mass in the sellar and right suprasellar region causing significant mass effect on the brainstem and right temporal lobe. She also reported amenorrhea for the past 7 years.

The patient's neurologic exam was notable for a left facial and left hemi-body weakness. She had grade 1/5 muscle strength in the left upper extremity and grade 4/5 strength in the left lower extremity. A Horner's syndrome was present on the left side, but her mental status and remainder of cranial nerve function were normal. An ophthalmologic evaluation demonstrated 20/40 visual acuity in the right eye and 20/50 visual acuity in the left eye, but no evidence of papilledema or increased intraocular pressure. On confrontational visual fields, she had a constricted OU although this was limited due to poor cooperation.

### Laboratory results

An endocrinologic evaluation demonstrated a serum prolactin of > 4700.0 ng/mL, and LH and FSH of < 0.2 mIU/mL and 0.5 mIU/mL, respectively. Her serum sodium, morning serum cortisol, serum T3/T4, and serum TSH levels were all within normal ranges. Her low LH and FSH levels were felt to reflect secondary amenorrhea from prolactinoma-induced hypo-gonadotrophic hypogonadism.

### Imaging studies

An MRI of the brain demonstrated a solid, 4.5 cm lobulated tumor with cystic components within the sellar and suprasellar region, causing upward displacement of the optic chiasm and direct mass effect on the right optic nerve (Fig. 1 a and b). There was no evidence of acute hemorrhage within the pituitary gland or stalk. Diffusion-weighted imaging showed an acute ischemic stroke in the territory of the right MCA (Fig. 1c). More detailed vessel imaging revealed a stenosis of the cavernous portion of the right internal carotid artery and right middle cerebral artery (Fig. 1d). The tumor also resulted in compression of the left supra-clinoid internal carotid artery as well as the right posterior cerebral, superior cerebellar, and basilar arteries. No other intracranial lesions were noted. An MRI of the spine showed no other lesions or masses.

### Management and outcome

The patient was admitted to the intensive care unit for monitoring and started on oral aspirin. Based upon the location of the mass and the elevated serum prolactin levels, a presumptive diagnosis of a pituitary macroprolactinoma was made, and because of her stable clinical condition (e.g., normal mentation, no bradycardia, or hypertension), she was started on oral cabergoline (0.25 mg twice weekly). Surgery was not selected as an initial treatment because of her stable neurologic status and lack of symptoms suggesting raised intracranial pressure. After 3 months of oral cabergoline, her serum prolactin levels normalized to 19.1 ng/ml. She continued to have a persistent mild left facial weakness and left upper extremity weakness, but had experienced significant improvement. An MRI scan performed at that time showed a dramatic reduction in the size of the tumor and improvement of the mass effect (Fig. 2 a and b). After 6 months, she continued to demonstrate improvements in her strength, and the serum prolactin level was 23.9. The tumor was stable in size on subsequent MR imaging. The dose of the oral cabergoline was increased and she was started on estrogen supplementation. At 9-month follow-up, her serum prolactin level was 13.1, and the size of the residual tumor was unchanged. Two years following her initial presentation, she was doing well from a clinical perspective with 4+/5 grade left-sided strength without evidence of tumor progression. Of note, the patient's time-of-flight MRA scans have shown persistent flow limitation in the right internal cavernous and supraclinoid internal carotid arteries (Fig. 2c).

## Discussion

This is one of the few reports of an acute ischemic stroke occurring secondary to a large pituitary adenoma in a child. Pituitary tumors causing ischemic strokes in a large vascular territory are very rare [5–9], and the majority of reported cases have occurred in the setting of apoplexy. In a recent literature review, Banerjee et al. reported a total of 25 cases of cerebral infarction from pituitary apoplexy. Among these, five patients had bilateral cerebral infarction, predominantly from invasion into the cavernous sinus and compression of the internal carotid arteries. The overall mortality rate was high (36% of patients) with a higher rate (60%) among patients with bilateral infarcts. The majority of patients (80%) in this series also underwent some form of operative intervention [8]. In another review, Rey-Dios and colleagues presented four adult patients with pituitary lesions who had cerebral infarction in the absence of apoplexy. Three of these patients were treated with surgery, and all had non-secretory macroadenomas. Only half of patients had symptomatic improvement [10].

Given the rarity of this phenomenon, optimal decision-making for these patients is a challenge. In this particular

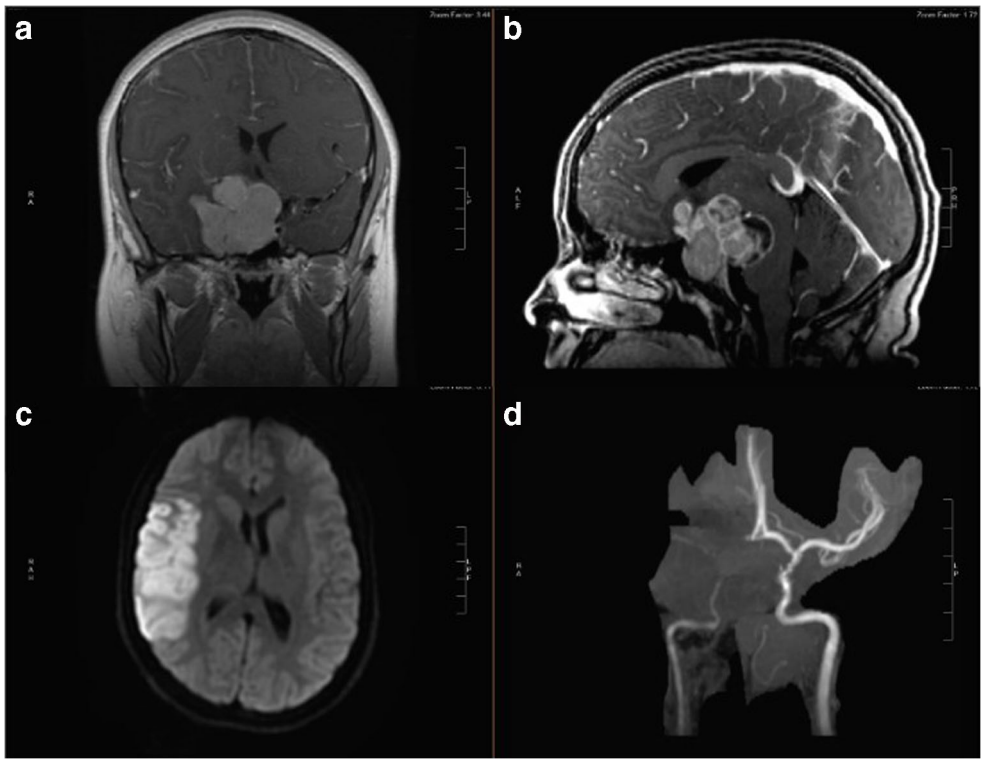


**Fig. 1** MRI with contrast demonstrating pituitary macroadenoma. Representative images of the patient IL's macroprolactinoma. Note the extent of tumor volume at time of presentation (**a** and **b**), with mass effect and upward displacement of the optic chiasm, compression of the brainstem, and extension deep within the right sylvian fissure. No hydrocephalus was seen. The resultant mass effect led to infarction of the right middle cerebral artery (MCA) as seen on diffusion-weighted imaging (**c**), and can be further appreciated on MR angiogram demonstrating near-complete occlusion of the right MCA (**d**)

case, medical management was chosen for several reasons. First, for most prolactinomas, cabergoline is effective and is generally considered to be the first-line treatment [11]. Breil et al. reported their experience with 11 pediatric prolactinomas treated with cabergoline, and achieved a mean tumor volume reduction of 80% [12]. Second, despite our patient's 2-week history of left hemiparesis, she was clinically stable at presentation and throughout her hospitalization. The ischemic stroke had likely occurred 2 weeks prior to presentation, and there were no signs of progression. Her mentation and level of consciousness remained normal; there were no signs or symptoms of increased intracranial pressure. We did anticipate that surgical treatment would be required (a) if the serum prolactin level failed to normalize, (b) if she demonstrated any signs of further clinical deterioration, or (c) if oral cabergoline did not result in a reduction of tumor size. Also, if there was an extension of the size of her stroke, we considered the possibility of decompressive hemicraniectomy.

Although medical management is often successful for the treatment of prolactinomas, the precise indications for

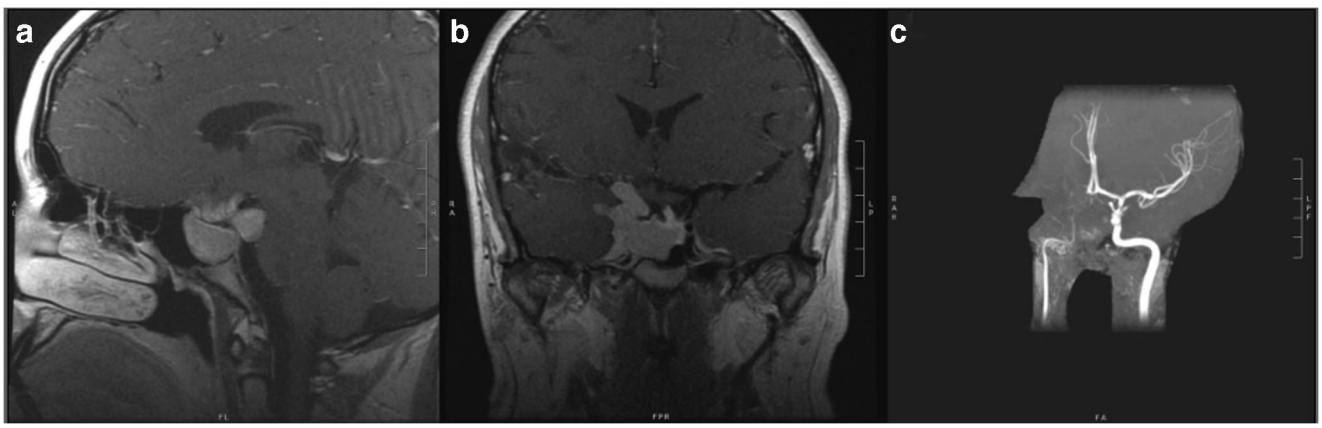


**Fig. 2** Post-treatment reduction of macroprolactinoma. The patient was managed medically on cabergoline. At time of 3-month follow-up, her tumor had dramatically reduced in size (**a** and **b**), and her hemiparesis had slowly demonstrated improvement with aggressive physical therapy. Her prolactin levels had also successfully normalized to 19.1. Time-of-flight MRA (**c**), however, showed persistent flow limitation in the right internal carotid artery

surgery have not been clearly delineated. Hoffman et al. analyzed 27 patients with pediatric pituitary prolactinoma, of which seven patients underwent tumor resection. The indications for surgery among their cohort included impending visual loss or failure of medication to reduce tumor volume [13]. Kim and colleagues followed a similar approach in 7 patients with pediatric prolactinoma as well [14]. Banerjee et al. found that, even in the setting of infarction, declining or severe visual field deficit was the most important factor for deciding when to proceed with surgical resection [8]. Pozzati et al. presented a case of a 15-year-old boy with a right parietal infarct from bilateral ICA occlusion who presented with altered mental status and left facial weakness without visual loss. With conservative management, the patient improved without requiring surgery [5]. Of note, our patient presented with a contralateral post-ganglionic Horner's syndrome in the setting of a predominantly right-sided lesion, which was likely secondary to some compression of the left carotid sinus given the sheer size of the tumor. However, she did not present with subjective visual deficits, and perhaps, this contributed to her favorable outcome.

## Conclusion

In summary, this case report describes the unusual presentation of a large pituitary prolactinoma causing an ischemic stroke in the distribution of the middle cerebral artery. Oral cabergoline is effective in this setting in achieving rapid reduction in tumor size. Surgical intervention should be considered if the patient experiences a clinical decline or if tumor does not respond to therapy.

### Compliance with ethical standards

**Conflict of interest** The authors have no conflicts of interest to report.

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