"Ma, I'm seein' double!"

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Gene, 11, presents with a recurring headache, slightly decreased vision and intermittent exotropia. He has a poor appetite and has not gained any weight in the last six months.

Patient statistics

- Recurrent headache
- Poor appetite
- Slightly decreased vision
- Optic nerve cupping bilaterally
- Otherwise, neurologic exam normal

Medical history

- Unremarkable

What's your diagnosis?

a) Pituitary adenoma
b) Arachnoid cyst
c) Cystic cranial pharyngioma
d) Rathke cleft cyst

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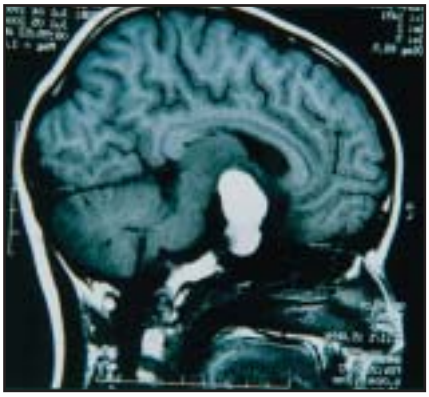

Figure 1. MRI of infused brain.

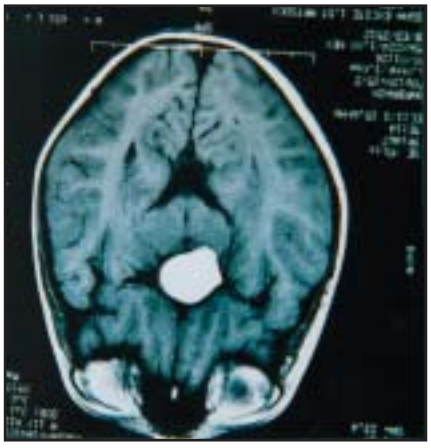

Figure 2. MRI of infused brain.

Clinical investigations

- Magnetic resonance imaging (MRI) of infused brain (Figure 1 and Figure 2); pediatric endocrinologist assessment.
- MRI showed a large cystic mass arising within the pituitary fossa and extending into the suprasellar cistern with mass effect on the right hypothalamus.
- Endocrinologist found a slight deficiency in thyroid and cortisol hormone and Gene has been started on both these medications.

Answer:
Cystic cranial pharyngioma

About cystic cranial pharyngioma

Craniopharyngioma is usually a slow-growing tumour. Symptoms frequently develop insidiously and become obvious after the tumour attains an approximate 3 cm diameter. The time from symptom onset to diagnosis ranges from one to two years.

Symptoms only become obvious after the tumour has a 3 cm diameter.

The most common presenting symptoms are:

- headache—between 55% and 86%,
- endocrine dysfunction—between 66% and 90% and
- visual disturbances—between 37% and 68%.

Three major clinical syndromes have been described that relate to the anatomic location of the craniopharyngioma:

- *prechiasmal localization* typically results in associated findings of optic atrophy (*e.g.*, progressive decline of visual acuity and constriction of visual fields),
- *retrochiasmal location* is commonly associated with hydrocephalus with signs of increased intracranial pressure (*e.g.*, papilledema, horizontal double vision) and
- *intrasellar craniopharyngioma* usually manifests with headache and endocrinopathy.

Both neurologic and general examinations are indicated.

Neurologic examination

- Signs suggestive of increased intracranial pressure, both horizontal double vision (unilateral/bilateral) and papilledema (unilateral/bilateral), should be sought in any patient suspected of having an intracranial mass.
- Visual-field examination may reveal various patterns of visual loss (most frequently bitemporal hemianopsia) suggestive of involvement (*i.e.*, compression) of the optic chiasma and/or tracts; visual fields should be tested further with formal testing.

General examination

- Hypothyroidism: Includes puffiness and non-pitting edema, slow return phase of deep tendon reflexes, long-standing effects on organ systems, hypoventilation and decrease in cardiac output, pericardial and pleural

effusions, constipation, anemia (normochromic and normocytic), decreased mental function and psychiatric changes.

Adrenal insufficiency

- Cortisol deficiency: This results in hypotension, which is often orthostatic. Gastrointestinal symptoms include anorexia, nausea and vomiting; other signs and symptoms include weight loss, hypoglycemia, lethargy, confusion, psychosis and intolerance to stress.

Headache, endocrine dysfunction and visual disturbance are the most common presenting symptoms.

- Aldosterone deficiency: Includes hypovolemia, decreased cardiac output, decreased renal blood flow with azotemia, fatigue, weight loss and cardiac arrhythmias due to hyperkalemia.

The diagnostic evaluation for craniopharyngioma includes precontrast and postcontrast computed tomography scans and magnetic resonance imaging, magnetic resonance angiography, complete endocrinologic and neuro-ophtalmologic evaluation with formal visual-field documentation, as well as neuropsychologic assessment.

Treatment

There are essentially two main management options available for treating craniopharyngioma:

- attempt at gross total resection or
- planned limited surgery followed by radiotherapy.

Other approaches that can be useful in the management of giant craniopharyngioma especially at the time of recurrence include:

- intermittent aspiration by stereotactic puncture or Ommaya reservoir placement,
- intracystic injection of bleomycin and
- internal irradiation with radiosotopes. Dx

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