Unusual presentation of more common disease/injury

A rare presentation of craniopharyngioma: delayed puberty

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Summary

Craniopharyngiomas are the most frequently encountered suprasellar tumours in children. Owing to the slow growth rate of these tumours, they are often quite large before becoming symptomatic. They are more common among children and older adults (55–74 years). Depending upon the direction of growth and tumour size, craniopharyngiomas can affect the hypothalamus, pituitary stalk, optic nerves and chiasm and carotid arteries. Compression of these neural and vascular structures frequently precipitates endocrine disorders, visual loss and an increased intracranial pressure. Hypopituitarism leading to a delayed puberty is a rare presentation of craniopharyngioma. The diagnosis of craniopharyngioma is usually made with the classic radiological imaging features based on CT and MRI.

BACKGROUND

Craniopharyngiomas are benign and aggressive epithelial neoplasms, constituting 1.2–3% of all intracranial tumours.¹ These are the most common tumours among children involving the hypothalamus and pituitary gland. Endocrine disorders including hypopituitarism may present in 50–60% of patients.²

In the present study, we report a case of craniopharyngioma which presents with delayed puberty and a short stature in a 15-year-old boy.

CASE PRESENTATION

A 15-year-old boy presented with bilateral frontal headache along with a decreased growth of male genitalia and a short stature. The genitalia were masculine having no ambiguity. No mental retardation was found. The patient was 112 cm in height and weighed 44 kg.

INVESTIGATIONS

An initial CT scan of the brain demonstrated a large well-circumscribed tumour measured as 44×52×37 mm on the suprasellar region, but also involving and expanding the pituitary fossa. The important feature of the mass was the presence of multiple calcifications (figure 1). It was difficult to fully assess the soft-tissue component owing to an artefact caused by the calcifications. The age of the child, localisation of the tumour and the imaging characteristics led to a preliminary diagnosis of craniopharyngioma.

The patient also had an MRI of the brain for confirming definitive diagnosis and preoperative planning. The mass was clearly delineated with internal focal low-signal intensity foci on T1W, T2W and GRE sequences in keeping with the calcifications. The soft-tissue component was iso- to low-signal on T1W and high-signal on T2W and FLAIR. Post administration of gadolinium, the soft-tissue component demonstrated heterogeneous enhancement (figure 2A,B). The pituitary gland and hypothalamic structures could not be clearly discerned from the tumour mass.

The laboratory findings showed an inadequate response to the growth hormone stimulation test and >2-fold increase in serum testosterone in response to the hCG stimulation test which is consistent with hypopituitarism.

TREATMENT

Considering all the laboratory, radiological and clinical findings consisted with craniopharyngioma and operative resection was done successfully without any neurological sequelae.

OUTCOME AND FOLLOW-UP

The resected mass was sent for histological examination which confirmed the presence of adamantinomatous craniopharyngioma (WHO Grade 1) (figure 3). The patient

Figure 1 Axial non-enhanced CT scan showed a large heterogeneous mass containing calcifications arising from sellar–suprasellar region with destruction of the sella turcica and pituitary gland.

Figure 2 (A, B) Coronal T2 MR image demonstrates the cystic portion of the tumour and hypointensities of the calcifications in the sellar-suprasellar location of the mass (A); post-gadolinium sagittal T1 MR image demonstrates heterogeneous enhanced mass in the sellar-suprasellar region (B).

was referred to endocrinologists for further evaluation and medical treatment.

DISCUSSION

Craniopharyngioma is considered the commonest intracranial tumour among children and adolescence accounting for 8.3% of intracranial tumours and 14.5% of all supratentorial tumours. These tumours grow as a pseudoencapsulated mass, usually in a suprasellar location and less commonly are intrasellar. Suprasellar tumours are usually both solid and cystic. Although they often destroy the pituitary, they may also grow towards the third ventricle, optic chiasm and tract, pons and thalamus. Through continued growth and pressure, the sella turcica is destroyed, as is the pituitary.³

Craniopharyngiomas are classically divided into two subtypes: adamantinomatous and papillary. The adamantinomatous subtype is most commonly found in the paediatric age group and usually presents as a complex

Figure 3 Photomicrography of adamantinomatous craniopharyngioma showing an epithelial component with a loose fibrous stroma (haematoxylin and eosin, original magnification $\times 125$).

mixed cystic and solid mass which characteristically contains flecks of calcification.⁴

Despite high survival rates (87–95% in recent series), quality of life is frequently impaired in long-term survivors owing to sequelae caused by the anatomical proximity of the tumour to the optic nerve, pituitary gland and hypothalamus, in many cases even entanglement with the hypothalamus.^{5–6} One of the serious quality-of-life complication of craniopharyngioma is a delayed puberty which is a problem that can manifest before and/or after a surgical treatment.⁷ In these patients hormonal studies have revealed the deficiencies of GH, FSH and LH in cases with craniopharyngiomas, which results in growth failure and a delayed puberty or even, sexual retardation.² In our patient hypopituitarism owing to craniopharyngioma was responsible for a decreased growth of male genitalia and a short stature.

Radiological examinations like skull x-ray study and CT scan are the most common radiographic techniques employed in evaluation. Intrasellar craniopharyngioma cases show sinus opacification, tumour expansion, enlarged sella turcica, cystic lesions, fluid level, bony erosions and calcifications.⁸ CT usually reveals the heterogeneous nature of the tumour with its solid and cystic components, calcification, multicysts, lytic lesions and may have irregular enhancement. Multiple calcifications are usually observed within the tumour, especially in younger patients. In adults, craniopharyngiomas are often not calcified. An MRI clearly shows the tumour extension, cystic portions, mixed-intensity signal and/or heterogeneous enhancement.⁸ On an MRI, the cystic component is usually hyperintense on T1W owing to the high protein content of the cysts, while the solid component is usually of a heterogeneous signal on T1W and T2W images. The solid component is enhanced heterogeneously. Gradient echo sequences are useful for the presence of calcification confirmed by demonstrating a 'blooming artefact'. Our case demonstrates typical radiological features of craniopharyngioma.

Treatment of craniopharyngioma is mostly excisional, through a craniotomy or transsphenoid approach. A complete removal of the tumour is preferred.² When vital

structures are involved and there is a significant risk of morbidity and mortality, subtotal removal of the tumour followed by supplemental radiotherapy is the preferred treatment.⁷ Craniopharyngiomas are radiosensitive, and radiotherapy plays a major role in preventing recurrence and improving survival of the patient.⁹

In conclusion, patients who are presenting with signs of hypopituitarism including delayed puberty and short stature, craniopharyngioma should be doubted when diagnosing tumours in the sellar–suprasellar region. The diagnosis of craniopharyngioma is usually made with the classic radiological imaging features.

Learning points

- ▶ Craniopharyngiomas are the commonest intracranial tumours among children and adolescence accounting for 8.3% of intracranial tumours and 14.5% of all supratentorial tumours.
- ▶ Depending upon the direction of growth and size, craniopharyngiomas can affect the pituitary gland and lead to a delayed puberty.
- ▶ The diagnosis of craniopharyngioma is usually made with the classic radiological imaging features based on a CT and magnetic resonance imaging.

Competing interests None.

Patient consent Obtained.

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