Rare disease

Pituitary stalk craniopharyngioma

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Summary

Craniopharyngiomas are benign but aggressive neoplasms arising along the craniopharyngeal duct. It is frequently located in the suprasellar region. Primarily pituitary stalk craniopharyngioma is unusual and uncommonly early diagnosed, before it enlarges and extends to supra or parasellar region. This unusual location and the small size pose therapeutic dilemmas, since it has the ability to grow larger. Currently, no consensus exists regarding the optimal management. The authors have recommended complete resection.

BACKGROUND

Craniopharyngiomas are benign but aggressive tumours arising along the path of the craniopharyngeal duct and most frequently located in the sellar or suprasellar region.¹ Controversies still exist over the exact pathogenesis. Evidence supports the theory that embryonic rests of cells from the craniopharyngeal duct are responsible for the development of the tumour. Other theory is the premise that existing cell-rests in the adenohypophysis undergo metaplasia.² Histologically confirmed primary craniopharyngioma at an ectopic location is rare, although cases of epidural and even extracranial neoplasms have been reported in sphenoid bone, ethmoid sinus, the nasopharyngeal region and intracranially in the corpus callosum, third ventricle, or pineal region.^{2–6}

We report a rare and early histologically confirmed case of a primary craniopharyngioma that arose in the pituitary stalk. The clinics, therapeutic choices and the surgical intervention will be discussed along with the disease processes that affect the pituitary stalk.

CASE PRESENTATION

A 42-year-old woman presented with persistent headache, polyuria and polydipsia for 12 months. The headache was tension type and not related to an electrolyte imbalance.

INVESTIGATIONS

MRI revealed a round lesion that adopt a homogenous hypointense signal on T1 sequences and enhanced homogeneously after gadolinium (figure 1A,B) and a hyperintense signal on T2 sequences.

TREATMENT

The patient underwent a right pterional craniotomy. The solid lesion was completely removed and the pituitary stalk was preserved. Histological studies revealed adamantinomatous craniopharyngioma (figure 2).

OUTCOME AND FOLLOW-UP

Follow-up MRI studies revealed no lesion and pituitary stalk preservation; midsagittal T1-weighted MR image obtained without contrast material demonstrated a normal sellar-suprasellar region (figure 3). The patient developed pituitary hypofunction, and postoperative corticosteroids and thyroid hormone replacement was necessary. The diabetes insipidus (DI) did not increase and was managed with intranasal desmopressin. The patient did not develop any new neurological symptoms following surgery and is being followed-up as an outpatient.

DISCUSSION

Craniopharyngiomas account for 1–4% of all intracranial tumours and 20% of the tumours of the sellar and chiasmatic region.^{1–7} These are predominantly suprasellar tumours, although they may be found within the sella turcica. In their usual location they may originate from squamous epithelial remnants of the Rathke pouch and sac. There is a possibility that craniopharyngiomas can originate anywhere along the tract of the obliterated craniopharyngeal duct.^{6–7} Another observation suggests that it is not necessary to postulate an embryonic origin for such cells, because they could appear later in life as a result of either cellular alteration or metaplasia from pituitary cells, which are also of ectodermal origin.^{7–8} The ectopic seeding of craniopharyngiomas is rare, and in most cases may be observed after the surgical removal of a suprasellar tumour.⁹ The occurrence of postoperative ectopic craniopharyngiomas by direct mechanical transplantation or via meningeal seeding is a known entity.^{9–10} Rare cases of epidural and even extracranial craniopharyngioma have been reported in sphenoid bone, ethmoid sinus and the nasopharyngeal region, and intracranially in the corpus callosum, third ventricle or pineal region.^{6–8–9–11–12}

Disease processes that affect the pituitary stalk can be distinct from processes that affect the hypothalamus or

Figure 1 (A) Presentation MRI scan: preoperative coronal T1-weighted MR image demonstrating a hypointense region lesion in the pituitary stalk (arrow). (B) Presentation MRI scan: preoperative sagittal T1-weighted MR image demonstrating an enhancing pituitary stalk and a hypointense region lesion in the pituitary stalk (arrow).

pituitary. Pituitary stalk lesions fall into one of the three categories: congenital-developmental, inflammatory-infectious and neoplastic.¹³ Pituitary stalk lesions are often discovered on MRI either incidentally or carried out to investigate symptoms such as those caused by DI like this patient.¹³ Primary tumours that can involve the pituitary stalk include gliomas such as astrocytomas, ependymomas and pleomorphic xanthoastrocytomas.^{3 5} A newly characterised tumour, the tanycytoma, can also involve the pituitary stalk. Tanycytomas are hypothalamic-suprasellar tumours that are biologically aggressive and have a high

rate of recurrence.^{13 14} Germinomas typically present as a hypothalamic or a pineal mass, however, they can also manifest as an isolated pituitary stalk thickening.¹⁵ Metastases to the infundibulum were the most common neoplastic cause of infundibular lesions in the series of Hamilton *et al.*¹⁴ The most common malignancies that result in pituitary metastases are breast and lung cancers.¹³ Leukaemia (chronic myelogenous leukaemia and acute myelogenous leukaemia), lymphoma and benign tumours like meningioma have been reported to involve the pituitary stalk.^{13 14 16}

Figure 2 Tumour histopathology. Photomicrograph showing light microscopy study of surgical specimen obtained in the pituitary stalk: palisaded columnar cell lining and wet keratin, demonstrating a typical craniopharyngioma of the adamantinomatous type.

Figure 3 Follow-up MRI scan: postoperative midsagittal image obtained demonstrated a normal sellar-suprasellar and pituitary stalk region.

It is generally accepted that two different subtypes of craniopharyngiomas exist. The classic adamantinomatous type appears to be present in more than 95% of paediatric cases, and is found most often in the suprasellar space, where they adhere to the adjacent vessels and indent the floor of the third ventricle.⁴ The papillary squamous variant is almost exclusively seen in adults and represents close to 30% of all the craniopharyngiomas seen in this population. These lesions are more often solid and lack the machine-like

oil seen in the adamantinomatous variant and are less likely to adhere to vessels and invade the brain.⁴

The present study illustrates a very uncommon case of a small and primary pituitary stalk craniopharyngioma. Complaints of headache, polyuria and polydipsia were responsible for the early diagnosis. This scenario poses therapeutic dilemmas; conservative management is a treatment option and a period of observation could be the option since the tumour is not threatening to the visual apparatus; it is a

small and early diagnosed tumour, without visual alteration or any structural compression. Usually, prechiasmatic craniopharyngiomas have the ability to grow to a larger size before being recognised⁴ and we intended that resection could avoid the growth. It is worth preserving the pituitary stalk, even though it is very difficult in surgeries involving large craniopharyngiomas. It may be sacrificed for total removal of the tumour.¹⁷ The resultant hypothalamic pituitary hypofunction may require cortisol and thyroid hormone replacement therapy.¹⁷ Patients may also develop DI and thus require nasal administration of desmopressin.¹⁷

The approach is fundamental to help visualise and manipulate the structures and maintaining the normal anterior pituitary function which is more often than generally supposed if the pituitary gland and stalk are preserved whenever possible.^{12 18 19} The highest percentage of hypothalamopituitary failure is encountered early postsurgery and there is a potential for recovery with time.^{17 19} The patient had already presented signs of stalk dysfunction (DI) owing to the authors decision for surgical intervention. The pituitary stalk was carefully handled during surgery and completely preserved but postoperative hormonal replacement was required. Probably it occurred from the minimal manipulation and the pre-existent dysfunction signalled by the clinical symptoms. There is no clear consensus on the treatment of craniopharyngiomas and aggressive conduct is generally indicated because they can be theoretically curable with complete resection. On the other hand, considering the high-morbidity rate related to radical excision, many authors believe that the best treatment is limited surgery followed by radiotherapy.⁴ Many surgical approaches have been used to remove these tumours, including pterional, subtemporal, interhemispheric, transcortical and transcallosal.¹⁸ Frontobasal interhemispheric approach in patients with craniopharyngiomas is very useful, safe and accurate for removal of craniopharyngiomas extending outside the sellar-suprasellar regions well as their relationship with hypothalamic structures and perforating vessels.¹⁹ The pterional approach, used in this case, provides a short distance to the suprasellar region and can be used for craniopharyngiomas located in the chiasmatic and interpeduncular cisterns.^{11 18 20}

Learning points

- ▶ Primary and small craniopharyngiomas occurring only in pituitary stalk are rare and the early diagnosis is unusual.
- ▶ The pterional approach is a valid choice for the removal of these tumours with preservation of the pituitary stalk, hypothalamic structures and perforating vessels.
- ▶ This unusual tumour location and the small size pose therapeutic dilemmas, since the tumour has the ability to grow to a larger size.
- ▶ In our opinion, the hormonal disturbance would occur in this location despite the size of the lesion. In this context, the authors have intended that resection could avoid the growth and recommend complete resection.

Competing interests None.

Patient consent Obtained.

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