A 55-Year-Old Woman Presenting with a Second Diagnosis of Craniopharyngioma Following Diagnosis and Successful Treatment of Craniopharyngioma as a 5-Year-Old Child

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Patient: Female, 5-year-old

Final Diagnosis: Pituitary tumor

Symptoms: Headache

Clinical Procedure: Surgery

Specialty: Endocrinology and Metabolic

Objective: Unusual clinical course

Background: Craniopharyngioma is a rare, partly cystic embryonic malformation of the sellar and parasellar region and is usually benign. This report is of a 55-year-old woman presenting with a second diagnosis of craniopharyngioma following diagnosis and successful treatment of craniopharyngioma as a 5-year-old child.

Case report: Our patient was diagnosed with craniopharyngioma at age 5 when she presented with headaches accompanied by nausea and vomiting, decreased visual acuity, polyuria, and polydipsia for 6 months. She was found to have diplopia and grade II papilledema. A skull X-ray showed separation of the sutures and a calcified mass in the suprasellar region. A pneumoencephalogram showed extension of the tumor into the third ventricle. Surgery was performed via transcallosal approach followed by radiotherapy at 5000 rays. She was followed up clinically and radiologically and had been disease-free until age 55, when she presented with headache and facial numbness. On examination, she had right-eye Horner syndrome, decreased sensation in the right side of the face, diplopia, and grade 2 facial palsy. An MRI revealed interval significant recurrence of the craniopharyngioma at the sellar/suprasellar mass with extension to the right Meckel's cave and the right posterior fossa. On April 6, 2023, she underwent surgical resection through a right-sided craniotomy and Kawase approach. This was followed by CyberKnife radiation therapy.

Conclusions: This report has presented a rare recurrence of craniopharyngioma with a 50-year interval and has highlighted the challenges in the diagnosis and the multidisciplinary approach to patient diagnosis and management.

Keywords: Craniopharyngioma • Adult Craniopharyngioma • Radiotherapy • Adamantinomatous Craniopharyngioma

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Introduction

Craniopharyngiomas (CPs) are rare, cystic, calcified, and slowly growing tumors [1]. They are benign (WHO grade 1) neoplasms that arise in the sellar/suprasellar region [1] but are considered clinically aggressive given their tendency to recur and to involve neighboring structures such as the pituitary gland, hypothalamus, optic chiasm, cranial nerves, and third ventricle [2]. Two types of craniopharyngioma – the more common adamantinomatous and the papillary types – have been recognized, but transitional or mixed types do occur [3-7]. Adamantinomatous craniopharyngioma shows irregular, infiltrative borders, complex architecture, wet keratin, calcifications, peripheral pali-sading, xanthogranulomatous reaction, and loose stellate reticulum [4,6], while in papillary craniopharyngioma there are papillary configuration of the tumor tissue with cauliflower-like morphology and fibrovascular cores [4], as well as solid sheets of well-differentiated nonkeratinizing squamous epithelium [4,6]. The differential diagnoses includes Rathke’s cleft cyst with squamous metaplasia, dermoid cyst, epidermoid cyst, and other sellar/suprasellar tumors [4]. They are believed to arise from neoplastic transformation of the ectoderm-derived epithelial cell remnants of Rathke’s pouch and craniopharyngeal duct [3-5]. While the adamantinomatous type is seen more frequently in children, the papillary type is more common in adults [2]. Clinical presentation is typically related to compressive symptoms in the form of headache, visual disturbance, and hormonal deficiency [2]. The treatment is surgical resection with or without radiation therapy [4]. The incidence of CP ranges from 0.5 to 2 cases per million people each year [2]. It has a very high recurrence rate of approximately 50%, which necessitates long-term follow-up [2]. Although ectopic recurrence is rare, it can lead to serious complications such as disequilibrium, hearing loss, and personality changes [8]. Previous case reports and case series have documented the possibility of recurrence, whether in the original or in ectopic sites [9,10]. This report is of a 55-year-old woman presenting with a second diagnosis of craniopharyngioma following diagnosis and successful treatment of craniopharyngioma as a 5-year-old child.

Case Report

Our patient was known to have a craniopharyngioma that had been treated surgically, followed by radiation therapy in 1972. At that time, the patient was 5 years old; she presented with severe headaches accompanied by nausea and vomiting, decreased visual acuity, polyurea, and polydipsia for 6 months. She was found to have diplopia and grade II papilledema. An X-ray of the skull showed separation of the sutures and a calcified mass in the suprasellar region. She underwent insertion of a bilateral ventriculoperitoneal shunt in August 1972 at Scripps Memorial Hospital, La Jolla, California. The ventriculogram revealed a total block of the ventricular system at the

Figure 1. Time course of craniopharyngioma with routine MRI. (A) Follow-up MRI in 2005. (B) Sagittal post-contrast MRI images of the sella showing recurrence of craniopharyngioma in the sella/suprasellar region with predominant right suprasellar extension. (March 2023). (C) Sagittal, T1 post-contrast MRI of the sella, showing post-operative resection of the sella and parasellar region with decompression of the optic chiasm (June 2023).
foramen of Monro, but with communication between lateral ventricles. Furthermore, a pneumoencephalogram performed in the same sitting with ventriculoperitoneal (VPS) revision at University Hospital, San Diego, California, on September 6, 1972, revealed an extension of the tumor into the anterior portion of the third ventricle. On September 18, 1972, she underwent surgical debulking of the craniopharyngioma via a transcallosal approach. The pathology report showed adamantinomatous craniopharyngioma (but pathology slides were not available). A post-operative pneumoencephalogram showed a reduction of the tumor size, and suprasellar calcification appeared the same as in the pre-operative study, with no evidence of hydrocephalus. The surgical treatment was followed by the 3-field technique of radiation therapy using Cobalt-60, with a course of 5000 rays to the tumor bed.

Since then, she has had regular follow-up visits at King Khalid University Hospital, Riyadh, with basic endocrine investigation and neuroimaging over the years. Her follow-up serial MRI scans did not reveal any tumor recurrence (Figure 1). In March 2023, she had a headache and facial numbness that lasted 2 months. Shortly thereafter, she developed an abnormal gaze, with visual disturbances in her right eye.

The physical examination was normal other than unilateral right-eye temporal hemianopia, right-eye Horner syndrome, decreased sensation on the right side of the face, mainly at the distribution of V2-V3, diplopia, and grade 2 facial palsy. New MRI scans of the brain revealed the remarkable interval progression of a sellar/suprasellar mass, predominantly right suprasellar extension, posterior extension to the posterior fossa, preoptic cistern, and right CPA angle, severely indenting the ventral pons on the right side, and lateral extension to the right Meckel’s cave and medial temporal region. The lesion measured 39×19×37 mm in its maximum anteroposterior (AP), transverse (TR), and craniocaudal (CC) diameters (Figure 1).

On April 6, 2023, she underwent uneventful surgical resection through the right-sided temporal craniotomy and Kawase approach (Figure 2). Near-total resection was achieved for the soft, firm, and fibrotic parts in the Meckel’s cave, posterior fossa, and suprasellar regions, with decompression of the optic chiasm and trigeminal nerve; the residual part was a calcified portion in the suprasellar area (Figure 1). A fat graft for reconstruction was taken from the right thigh. Pathologic evaluation of the tissue specimen from the recurrent adult tumor revealed an adamantinomatous CP (ACP), benign (WHO grade 1), with degenerative changes, including fibrosis and keratin. In the second figure, the classical cords, lobules, and trabeculae of well-differentiated squamous epithelium were bordered by palisading columnar epithelium. These peripheral cells surrounded looser, plumper cells called “stellate reticulum” (Figure 3). There were no signs to suggest that the recurrent tumor was that of the adult papillary craniopharyngioma or Rathke’s cleft cyst. The pathology slides from the first surgery were not available for re-examination.
Surgery was followed by CyberKnife radiation therapy (dose 5000 CGY/25FX), and the treatment was completed in August 2023. During the following months, the patient developed absence seizures and some impairment of cognitive function.

**Discussion**

This case report describes a rare remote recurrence of craniopharyngioma after surgical resection and radiotherapy, emphasizing the need for lifelong follow-up of these patients.

CP constitutes approximately 2.5% to 3% of all brain tumors [11]. Embryologically, it is raised from the remnants of Rathke’s pouch. It displays a bimodal age distribution (5-15 years and 45-60 years), with 2 histological subtypes: ACPs are the most common type in the pediatric age group [11,12], while papillary CPs (PCPs) are more frequent in adults but are less frequent overall [12]. CP is a benign tumor with vague behavior because of its propensity to invade nearby structures and to recur even after total resection. Despite its high survival rate, which can range from 65% to 100%, CP has a high risk of recurrence of about 20% to 40%, which necessitates long-term follow-up [13]. Recurrences commonly occur at the original site [2,13]. Ectopic recurrence has been reported, although it is highly uncommon, through direct implantation of the tumor cells along the surgical path during the procedure [8,13,14]. Another possible mechanism is the seeding of the tumor cells through the subarachnoid spaces or spillage into cerebrospinal fluid [8,15-17].

The long-term prognosis of patients is nevertheless significantly affected by the recurrence of CP. Mortini et al found a recurrence rate of 24.5% on follow-up of 106 patients over 83 months [18]. The recurrence rate was highest in the first 3 years after surgery, after which it plateaued [11,19]. However, recurrences can occur decades later, after a time of slumber, and are well recognized. Our patient had a remote recurrence of CP in the sellar/suprasellar with new extension to the posterior fossa through the Meckel’s cave region 50 years after surgical resection and RT. In the literature there are few case reports of late-onset recurrence of CP. The case with the longest time frame, to our knowledge, was a recurrence after 30 years that showed up as an “ectopic” CP in the Sylvian fissure after the original tumor was resected [16]. Our patient’s clinical presentation was similar to previously reported cases in which headache and visual disturbances were prominent [11,19]. Postoperatively, she developed diabetes insipidus and, over time, some functional disability, as reported previously [19-21].

Several risk factors have been associated with increasing the risk of CP recurrence. Studies have found that after gross total resection (GTR), the likelihood of a tumor coming back has been estimated at between 0% and 26% [11,19-23]. This is much lower than in patients with a subtotal or partial tumor excision, where 25% to 100% of individuals experience tumor recurrence [11,19-25]. Adjuvant RT dramatically increases tumor control rates, with 10-year follow-up recurrence rates ranging from 10% to 63% [11,22,25].

Patients who undergo subtotal resection without further radiation therapy run a higher risk of relapsing [11,19,20,22]. Gender, tumor size, location, and histological subtype have conflicting prognostic value; nevertheless, in general, these characteristics...
do not appear to increase the likelihood of recurrence [11,25]. However, recurrence was more frequent in patients with early-onset disease who were younger than 10 years old [26].

Treatment of recurring diseases results in higher morbidity and mortality than do initial treatments [25,26]. Because of the scars and adhesions from the first operation, the surgical success rates for recurring disease are much lower than for primary surgery [4,11,25]. Only when the patient exhibits indicators such as compressive symptoms or a midline shift should surgery be done [11,25,26]. Reirradiation is an alternative to surgery for selected patients with recurrent, refractory craniopharyngiomas, and it appears that stereotactic radiotherapy can be successful for controlling recurrent tumors after surgery, especially if the tumor is too close to the optic chiasm [4,27–31]. Other alternatives include radiosurgery, intra-axial radiation, bleomycin administration, and salvage surgery for solid lesions that pose a serious threat to life [32,33]. In any event, each patient should receive individualized care while choosing a course of treatment for recurrent illness.

Conclusions

This report presents a rare recurrence of craniopharyngioma with a 50-year interval and highlights the challenges in diagnosis and the multidisciplinary approach required for patient diagnosis and management.

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