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Case Report

Ectopic clival craniopharyngioma with intratumoral hemorrhage: A case report

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ABSTRACT

Craniopharyngiomas located outside the intra/suprasellar region are rare and the incidence of intratumoral hemorrhage is also rare. A 45-year-old woman with visual field defect underwent subtotal resection of the tumor located in the skull base. CT revealed a well-defined mass with calcifications involving the clivus, and MRI showed anterior displacement of a normal pituitary gland by the mass consisting cystic portions with some fluid-fluid levels and enhancing solid portions. Surgery and histopathological examination revealed an ectopic adamantinomatous craniopharyngioma arising from the clivus with intratumoral hemorrhage. Though this condition is rare, detailed analysis of symptoms in association with imaging findings should alert the physician to the possibility of craniopharyngiomas.

Introduction

Craniopharyngiomas are benign tumors and comprise 1.2%-4.6% of all intracranial tumors, accounting for 0.5-2.5 new cases per million per year. The majority of these tumors have both intra- and supra-sellar components. Primary presentations outside this region are rare [1]. Craniopharyngiomas can be accompanied by cystic changes that rarely represent intratumoral hemorrhage [1], only a few cases of which have been reported [2]. We, herein, report a rare case of primary ectopic clival craniopharyngioma with intratumoral hemorrhage and review relevant published reports.

Case report

A 45-year-old woman was incidentally found to have visual field defect at a previous hospital and was referred to our hospital. She had undergone colectomy for sigmoid colon cancer 6 years earlier. On physical examination, she was noted to

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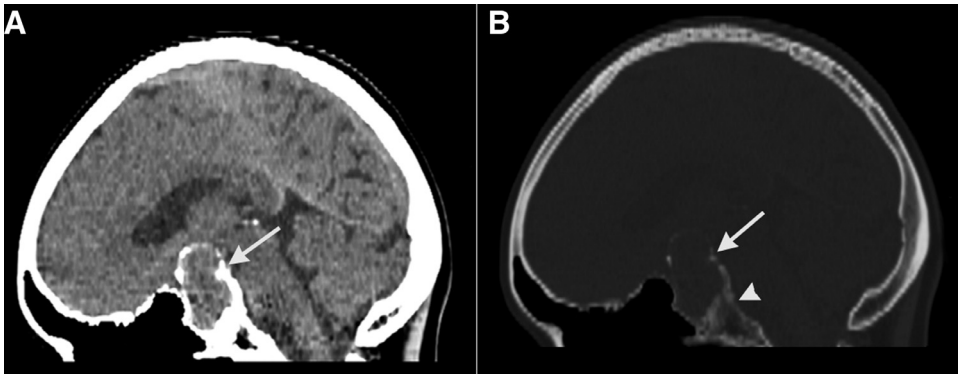

Fig. 1 – Sagittal unenhanced CT images. Brain window image (A), bone window image (B), showing a well-defined mass (arrows) involving the skull base and clivus with peripheral calcifications. Bone windowing (B) shows a bulge in the clivus (arrowhead) adjacent to the mass.

have a right upper visual field defect but was otherwise neurologically intact. Laboratory studies, including pituitary hormones, were normal.

Computed tomography of the brain showed a well-defined mass involving the skull base and clivus with peripheral calcifications (Fig. 1). Computed tomography angiography revealed a mass without significant abnormal vascularity. Magnetic resonance imaging of the brain revealed a well-defined mass ($3.7 \times 2.1 \times 2.4$ cm) involving the clivus, the mass being heterogeneously hyperintense on T1-weighted images and heterogeneously hypointense on T2-weighted images. It consisted of cystic portions with some fluid-fluid levels, and solid portions with contrast enhancement of some of the solid parts after administration of gadolinium. Sagittal images showed anterior displacement of a normal pituitary gland and stalk; thus, the tumor was arising from the dorsum sellae and clivus, not from the intra/suprasellar region (Fig. 2). The radiological differential diagnoses included ectopic pituitary adenoma with hemorrhage, ectopic craniopharyngioma, giant cell tumor/giant cell reparative granuloma with aneurysmal bone cyst (ABC) changes, and fibrous dysplasia with ABC changes. The patient underwent subtotal resection via an endoscope-assisted, endonasal, transsphenoidal approach. The tumor was located in the clivus and clearly separate from the pituitary gland and infundibular stalk. The cystic portion contained yellow fluid and some blood clots. Histopathological examination of the resected specimen revealed typical features of adamantinomatous craniopharyngioma with nodules of wet keratin. Macrophages containing hematoidin and hemosiderin depositions were noted, this being suggestive of intratumoral hemorrhage (Fig. 3).

There were no obvious interval changes during 3.5 years of follow-up.

Discussion

Ectopic craniopharyngiomas can be located in the clivus away from the intra/suprasellar region and pituitary gland. Craniopharyngiomas may arise anywhere along the path of the craniopharyngeal duct, especially in the intra/suprasellar regions. Primary ectopic craniopharyngiomas arise in the absence of any previous surgery whereas secondary ectopic craniopharyngiomas are ectopic recurrences of a previously surgically-managed tumor that arise from tumor cell spillage along the resection corridor or via the cerebrospinal fluid; secondary ectopic craniopharyngiomas being a rare but more common than primary ectopic craniopharyngiomas [3]. There is no consensus as to the mechanism of development of primary ectopic craniopharyngiomas; one possible explanation for the tumor site in the present case is migration of squamous epithelial cell remnants of the obliterated craniopharyngeal canal [4]. The infrasellar region is the most common of the unusual locations of craniopharyngiomas, approximately 50 such cases having been reported. The most common site for infrasellar craniopharyngiomas is the sphenoid sinus, either alone or combined with other sites such as the nasopharynx, sella turcica, suprasellar, ethmoid sinuses, or maxillary sinus [5]. However, it is sometimes difficult to distinguish between primary infrasellar craniopharyngiomas and intra/suprasellar craniopharyngiomas developing caudal to the infrasellar region, especially when the tumor is large. The second most common extrasellar location is the cerebellopontine angle (13 reported cases), followed by the frontotemporal region (4), fourth ventricle (3), pineal region (2), and corpus callosum (1) [6]. Only one patient with an ectopic clival craniopharyngioma completely separate from the sellar region has been reported [4].

Craniopharyngiomas can present with intratumoral hemorrhage. Craniopharyngiomas with intratumoral hemorrhage are rare, only 21 cases (adamantinomatous type, 2; papillary type, 8; and unknown, 11) have been reported. Seventeen of these tumors were located in the intra/suprasellar region; the location was not specified for the other four [2]. No ectopic craniopharyngiomas with intratumoral hemorrhage have been reported. The symptoms are similar to those of pituitary apoplexy. Causes of intratumoral hemorrhage can be classified as spontaneous or secondary. The pathogenesis of spontaneous intratumoral hemorrhage in craniopharyngiomas is unknown but may include degenerative changes in blood vessels and the presence of numerous immature

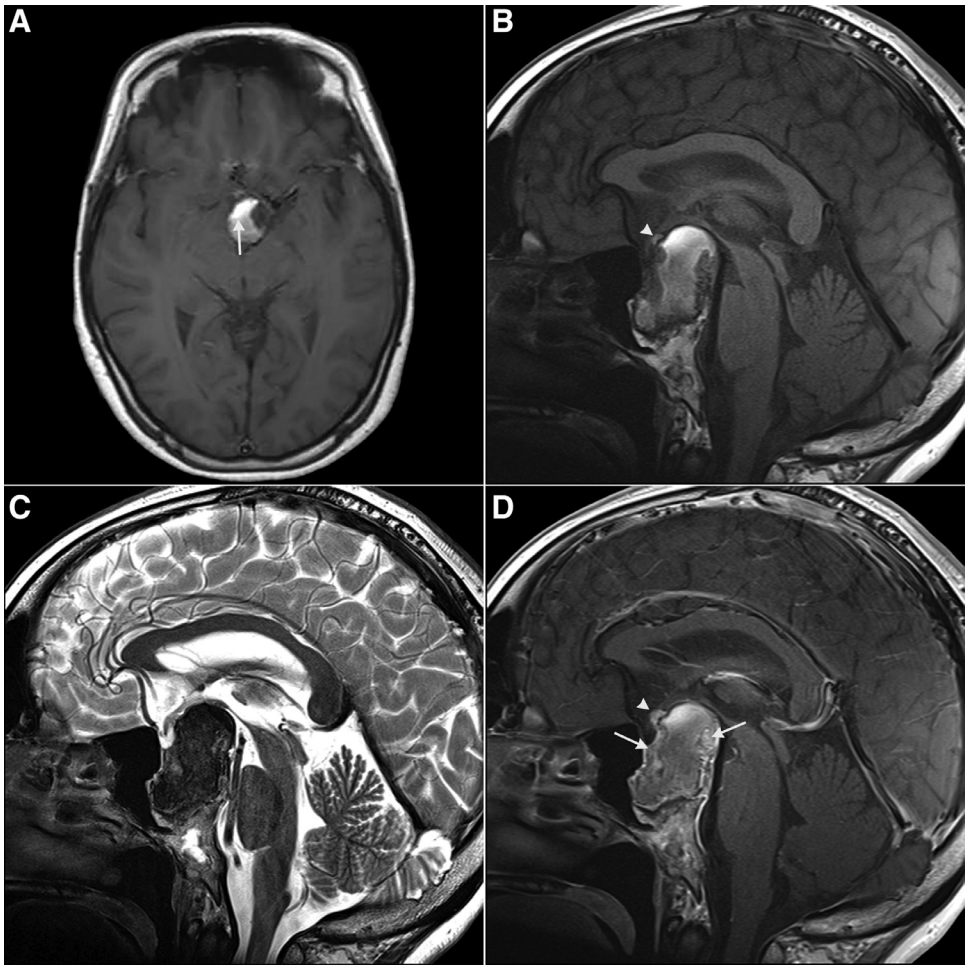

Fig. 2 – Magnetic resonance imaging (MRI) showing a well-defined mass (3.7 x 2.1 x 2.4 cm) involving the clivus, the mass being heterogeneously hyperintense with some fluid-fluid levels (A: arrow) on T1-weighted images (A,B), and heterogeneously hypointense on T2-weighted images (C). The mass consists of cystic and solid portions with contrast enhancement of some of the solid parts (arrows) on T1-weighted images with gadolinium enhancement (D). Sagittal images showing a normal pituitary gland and stalk (B,D: arrowheads) that have been displaced anteriorly by the lesion.

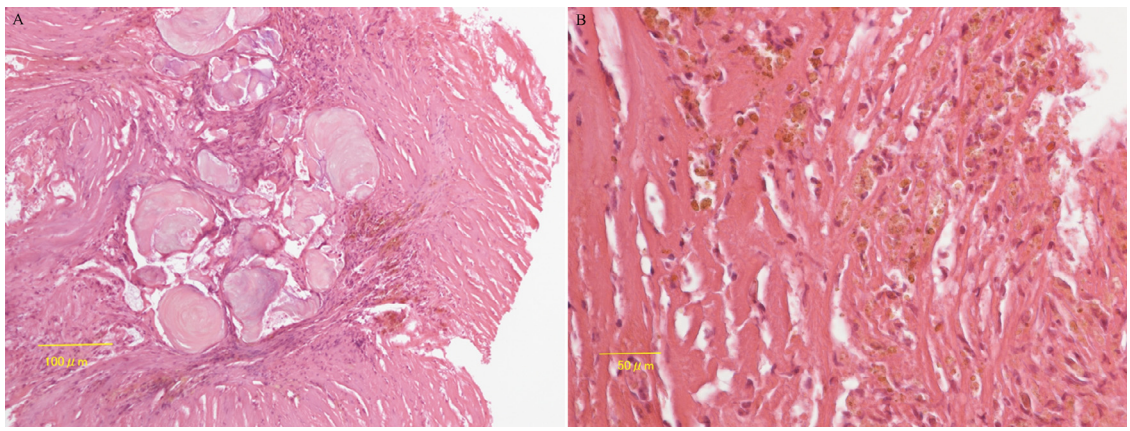

Fig. 3 – Hematoxylin and eosin-stained sections (A: x 200, B: x 400) showing nodules of wet keratin (A) and macrophages containing hematoidin and hemosiderin deposition (B).

blood vessels [2]. Secondary intratumoral hemorrhage has been associated with minor head injury, lumbar puncture, and surgical intervention [7]. Six of the previously reported 21 patients with intratumoral hemorrhage had spontaneous bleeding and the other 15 had secondary or unknown type of bleeding [2]. Asymptomatic intratumoral hemorrhage in craniopharyngiomas may be more common; no published reports have reported possible causes of asymptomatic intratumoral hemorrhage in craniopharyngiomas.

The differential diagnoses of hemorrhagic tumor of the intra/suprasellar region or clivus include hemorrhagic pituitary adenoma, hemorrhagic Rathke cleft cyst, giant cell tumor/giant cell reparative granuloma with ABC changes, and fibrous dysplasia with ABC changes. Hemorrhagic pituitary adenomas are well known; one study reported that the incidences of subclinical pituitary adenoma hemorrhage and pituitary adenoma with clinical apoplexy are 14.3% and 3.4%, respectively [8]. Giant cell tumors, giant cell reparative granulomas, and fibrous dysplasias of the clivus are extremely rare. Only one case of giant cell tumor with ABC changes in the skull base has been reported [9], and there have been no case reports of giant cell tumor, giant cell reparative granuloma, or fibrous dysplasia with ABC changes in the clivus.

Conclusion

Ectopic craniopharyngiomas can be located in the clivus separate from the intra/suprasellar region and pituitary gland, and ectopic craniopharyngiomas can present with intratumoral hemorrhage. Craniopharyngiomas located outside the intra/suprasellar region are rare and the incidence of intratumoral hemorrhage is not clear. However, detailed analysis of symptoms in association with imaging findings should alert the physician to the possibility of a craniopharyngioma, thus enabling formulation of an appropriate treatment plan for this potentially curable tumor.

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