CASE REPORT

Confounding features of ectopic craniopharyngioma: A differential for tumors showing diffusion restriction

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ABSTRACT

Craniopharyngiomas comprise approximately 1-3% of all intracranial tumors and arise from squamous epithelial rests along remnants of Rathke's cleft. They have bimodal age distribution. The ectopic sites reported are the nasopharynx, sphenoid bone, pineal region, cerebellopontine angle and third ventricle. We report a case of ectopic craniopharyngioma in the Foramen of Monro with restricted diffusion, which have is an unusual feature, not commonly reported and is a potential pitfall.

Key words: Ectopic craniopharyngioma, restricted diffusion, T1 hyperintensity

Introduction

Craniopharyngiomas are neoplasms which arise from the craniopharyngeal duct usually located in suprasellar and sellar regions. Ectopic locations include the third ventricle, nasopharynx, sphenoid bone, cerebellopontine angle and the pineal region.[1] We report a case of a squamous-papillary variant of craniopharyngioma essentially in the foramen of Monro with a small stalk in the suprasellar cistern suggesting its true site of origin and its unusual pattern of diffusion restriction.

To the best of our knowledge, craniopharyngioma does not show diffusion restriction and hence, the aim of presenting this case is that craniopharyngioma to be included in suprasellar lesions showing diffusion restriction.

Case Report

A 55-year-old female referred to our hospital with a 10 month history of repeated episodes of headache associated with vomiting that had worsened over the past 1 week, followed by one episode of generalized tonic clonic convulsions. There was fever on and off and there was no visual complaints or limb weakness. On examination, she was conscious and cooperative. Cranial nerves examination revealed left eye nystagmus, normal visual field and acuity. There was no sensory or motor deficit. Fundoscopy revealed bilateral papilloedema. Magnetic resonance imaging (MRI) carried out elsewhere reported as partly thrombosed giant right internal carotid artery aneurysm. She was referred for computed tomography angiogram of the cerebral vessels.

Plain CT brain revealed a midline, well defined round and heterogeneous mass in the region of foramen of Monro showing patchy areas of peripheral hyperdensity with the mean Hounsfield Unit 70 [Figure 1]. There was obstructive hydrocephalus due to pressure effect on foramen of Monro, accompanied by dilatation of the bitemporal horns of lateral ventricles. Posterior fossa and fourth ventricle were unremarkable. The lesion showed minimal enhancement and anterior displacement of anterior cerebral artery (ACA) and its callososeptal branches without encasement [Figure 2].

Review of the MRI Brain revealed a large well-defined lobulated cystic mass lesion in midline in the region of foramen of Monro with extension in to the frontal lobe. The mass was predominantly hyperintense on T1 [Figure 3], heterointense on T2 weighted images and was not suppressed in Fluid attenuated inversion recovery (FLAIR) [Figure 4]. The lesion revealed nodular rim

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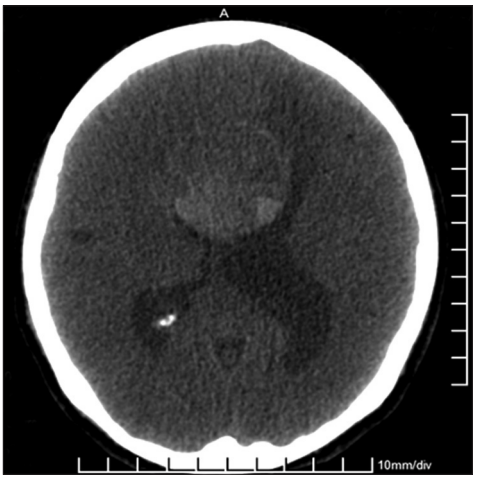

Figure 1: NECT brain revealed a midline, well defined round and heterogeneous mass in the region of foramen of Monro showing patchy areas of peripheral hyperdensity with HU 70

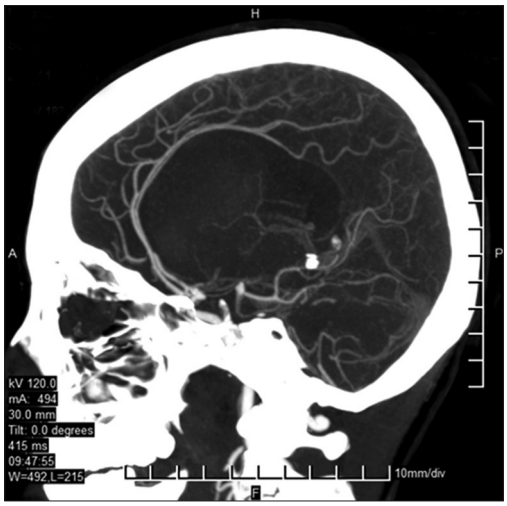

Figure 2: CT angio MIP imageshows anterior displacement of bilateral anterior cerebral artery and callososeptal branches without encasement

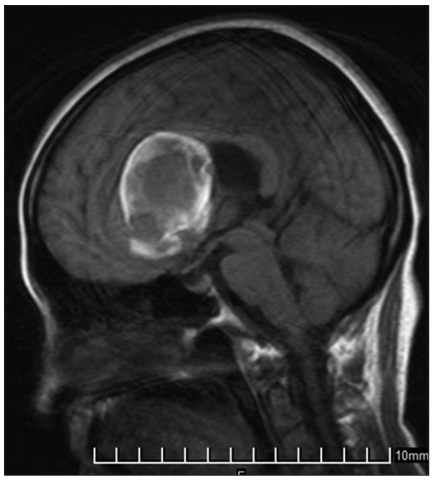

Figure 3: T1WI shows hyperintense mass in the region of foramen of Monro extending superiorly into the frontal lobe

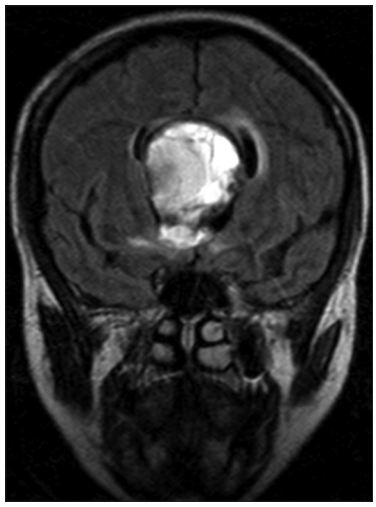

Figure 4: Coronal FLAIR the lesion remains hyperintense

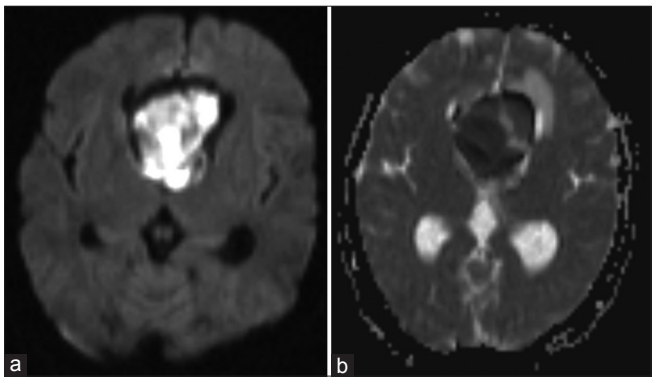

Figure 5: The lesion showed diffusion restriction

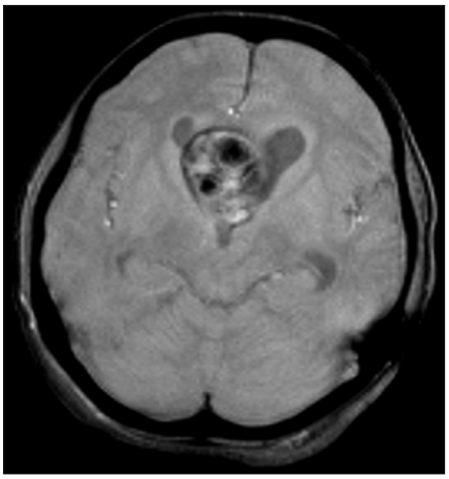

Figure 6: The mass shows blooming in gradient sequence

enhancement on administration of gadolinium. The third ventricle appears to be displaced inferiorly and fourth ventricle appears normal. The major part of the lesion showed restricted diffusion with low-apparent diffusion co-efficient (ADC) values [Figure 5], and peripheral blooming in gradient [Figure 6]. The flow voids of bilateral internal carotid arteries were normal. Magnetic

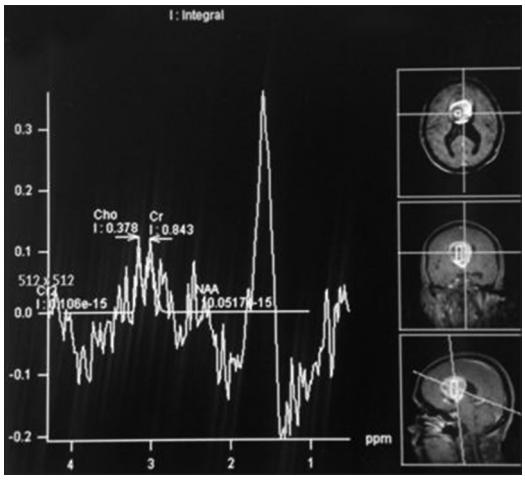

Figure 7: MR spectroscopy reveals prominent peaks centered at 1-1.5 ppm, which probably corresponded to lipid/cholesterol peaks

resonance (MR) angiogram was normal and hence the possibility of thrombosed aneurysm was ruled out. MR spectroscopy revealed prominent peaks centered at 1-1.5 ppm, which probably corresponded to lipid/cholesterol peaks [Figure 7].

Discussion

Craniopharyngiomas comprise approximately 1-3% of all intracranial tumors and arise from squamous epithelial rests along remnants of Rathke's cleft.[2] They have bimodal age distribution with common occurrence in children (peak incidence 5-10 years).[3] There is a smaller second peak in middle aged adults. Most of the craniopharyngiomas arise in suprasellar and sellar regions. The ectopic sites reported are the nasopharynx, sphenoid bone, pineal region, cerebellopontine angle, and third ventricle.[1,4,5]

Craniopharyngiomas are of two histological subtypes; the adamantinomatous type and the papillary type.[3] Adamantinomatous tumors look grossly cystic but may have both solid and cystic components. The cyst content is rich in cholesterol, which is typically described as machine-oil like. Calcification, fibrosis and inflammation are also common features. They are more adherent to surrounding structures and invasive. Recurrence is more common with adamantinous subtype. The papillary type is usually solid, spherical and shows no calcification. They are commonly situated within the third ventricle and usually affect adult age group as in our case.

The presence of calcification certainly has diagnostic value for craniopharyngiomas, and CT is far superior to MRI in the detection of calcification.[6,7] Calcification is more frequent in childhood and adolescence than in adulthood as in our case. However, particularly in adults, a substantial number of craniopharyngiomas do not have calcification and in such cases MRI, including non-contrast T1-weighted images, should play an important role in pre-operative diagnosis.[7] The lesion showed predominant peripheral T1 hyperintensity and T2 hypointensity and blooming in gradient echo sequence which could be due to the intralesional hemorrhage. Hyperintensity in T1 weighted sequences is due to protein, hemorrhage, cholesterol, and other fatty components.[8] With administration of gadolinium the wall and solid portions of the tumor show enhancement.

The major part of the lesion showed diffusion restriction which is a very unusual pattern of presentation of craniopharyngioma. Tumors with high cellularity show diffusion restriction. Diffusion weighted images, ADC maps are useful for determination of consistency of intra-axial brain tumor. Operating time is influenced by tumor consistency, where soft-tumors can easily be resected with gentle manipulation and suction, careful dissection is required for harder tumors. Thus information regarding the consistency of brain tumors certainly aids the surgeon in designing the resection strategy.

In our case, with the imaging characteristics we were able to conclude this intraventricular highly cellular lesion with necrotic and hemorrhagic components showing diffusion restriction along with lipid peak to be either ectopic craniopharyngioma or intraventricular abcess preoperatively. The histopathology confirmed the lesion to be a mixed cystic and solid papillary type craniopharyngioma. Our patient suffered no post-operative endocrine dysfunction as only a partial excision was performed. However, this increases the chances of recurrence.

The differential diagnosis for lesions in anterior third ventricle and foramen of Monro includes, pituitary macroadenoma, meningioma, optic or hypothalamic glioma, suprasellar germinoma, epidermoid, ependymoma, subependymoma, inflammatory masses, and metastasis. Pituitary adenoma often is greater in bulk within the sella and has a tendency to expand the sella. The pituitary gland is separately seen. Meningioma usually shows homogeneous intensity and contrast enhancement without cysts, and has broad-based attachment frequently associated with Dural tail sign. Both optic/hypothalamic glioma and suprasellar germinoma are more common in children, rarely have a sellar component and are rarely calcified. The tumors are usually isointense on T1 and usually lack a cystic component and germinoma show intense contrast enhancement. Both epidermoid and subependymoma do not usually show any contrast enhancement. Inflammations such as sarcoid or histiocytosis may manifest as suprasellar disease. Typically, these lesions are infiltrative, layering along the pituitary stalk and the adjacent undersurface of the brain, but occasionally, sometimes frank granulomas may occur in the suprasellar region giving rise to diagnostic difficulty.

Conclusion

The characteristic morphology and imaging features of the tumor and the extension in to the suprasellar cistern allowed us to make the diagnosis of craniopharyngioma pre-operatively

despite the unusual nature of restricted diffusion, which this tumor usually does not show.

The reason for the restricted diffusion could be due the high cellularity in this tumor. Thus, information regarding the consistency of brain tumors with the help of diffusion weighted imaging certainly aids the surgeon in designing the resection strategy.

Hence, the possibility of ectopic craniopharyngioma should also be considered as a differential for suprasellar tumors showing restricted diffusion in the proper clinical and imaging context.

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Conflicts of interest

There are no conflicts of interest.

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