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Case Report

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Fourth Ventricle Papilloma and Cranio-Cervical Junction Meningioma: Coincidental Tumors: a Case Report and Review of the Literature

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Abstract

The co-existence presence of meningioma and choroid plexus papilloma is very rare. Few cases of simultaneous tumors at the 4th ventricle described in the literature but there is no a single craniocervical, 4th ventricle combined tumour case reported in the recent neurosurgery literature. The condition is difficult to be diagnosed clinically as there are discrepences between clinical and radiological findings, the best outcome was reached by the simultaneous removal of both tumors. Meningiomas with concurrent papillomas are thought to be different primary brain tumours arising in the same individual at random.

Keywords: *Concurrent Intracranial Tumours; Meningioma; Choroid Plexus Papilloma; Ventricular Tumours*

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Introduction

Choroid plexus papillomas (CPPs) are rare tumors originating from the neuroectoderm, they comprise less than [1]. It has been found that children are the most heavily affected portion of the population (70% of C.C.Ps) with 50% presenting before the second year of life [2]. A very few reports of CPP in adult are present in the literature [2–5].

Skull basal meningioma's uncommonly affect the foramen magnum. Despite the accelerating advances in imaging modalities, the tumor location and rate of recurrence along with rate of vertebral artery (V.A) encasement and surgical approaches are still open to debate. We report an unusual case of an intraventricular choroid plexus papilloma (CPP) occupying the fourth ventricle occurring together with Cranio-cervical junction meningioma. The clinical presentation, preoperative imaging, surgical treatment and histologic features of the two tumors are discussed.

Case Report

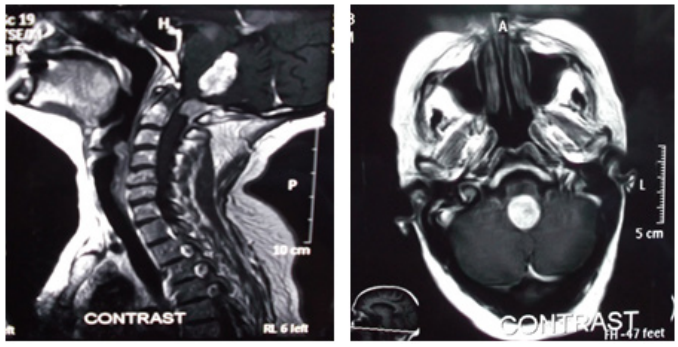
Clinical presentation

This study presents a woman aged 45-year-old complaining of progressive confusion, altered facial sensation, vomiting for 2 months, gait unsteadiness and both L.L weakness. The medical history is insignificant, Neurofibromatosis was excluded. On examination, she had an unsteady, wide.

Radiological findings

Brain and craniocervical MRI reveals a well-defined elongated mass measures 44*21 mm with homogenous contrast enhancement which is filling the 4th ventricle, anteriorly it is seen markedly indenting the posterior aspect of the brain stem.

It shows also LT SIDES Dural based homogenous contrast enhanced S.O.L at craniocervical junction with homogenous contrast enhancement measures 23*21 mm. Figure (1a, 1b).

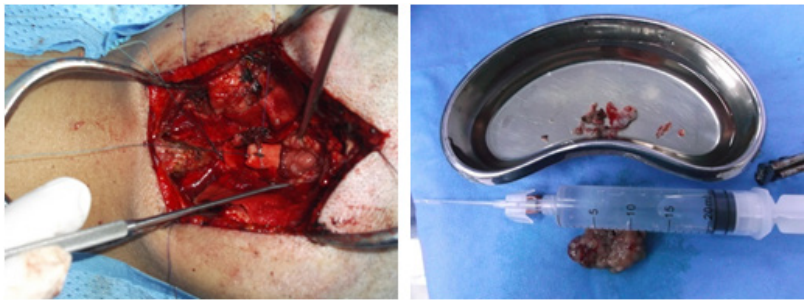


(A) (B)

Figure 1: *Brain and craniocervical junction preoperative MRI images with contrast sagittal (a) and axial (b) views, craniocervical junction shows Fourth ventricle papilloma (red arrow) and Cranio-Cervical junction meningioma(blue arrow).*

Surgical intervention

The patient was underwent midline sub-occipital craniotomy. Opening the cistern magna and separating the tonsils to reach the tumours. It was shrunken by coagulation and irrigation and then removed. C1 laminectomy and gross total excision of the meningioma was established. Figure 2



(A) (B)

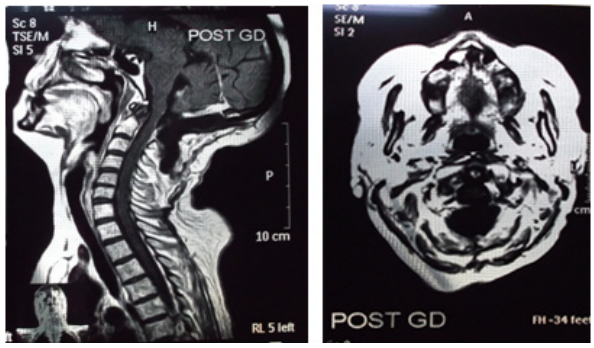
Figure 2: *Intraoperative image shows exposed tumor filling the 4th ventricle (a), (green arrow), and both tumors samples (b), (black arrows).*

Pathology

Histopathology revealed a CCJ meningioma's well as a choroid plexus papilloma.

Postoperatively

The patient evolution was satisfactory, she was admitted in INTENSIVE CARE UNIT for two days for observation, she was underwent motor rehabilitation with good clinical evolution. It is debatable whether or not to treat C.C.P with post-operative chemotherapy or radiation, therefore we didn't recommend her to do. Figure 3



(A) (B)

Figure 3: *Brain and craniocervical junction postoperative MRI images with contrast sagittal (a) and axial (b) views at level of craniocervical junction shows total resection of both tumors. (white arrows).*

Discussion

Histopathologically, CCPs are rare benign neoplasma, the origin of which is the choroid plexus epithelium of the 4th ventricle in adults and the lateral ventricles in children are the most common sites. CCP are usually treated by complete microsurgical resection, a surgery which is argued to be curative, however the role of chemotherapy and radiotherapy is open to be dispute.

As to meningeal tumors The second most frequent location for meningioma's of posterior fossa is the Craniovertebral junction meningiomas which is either situated in the ventral or dorsal aspect of the foramen magnum, ventral foramen magnum lesions are obviously more difficult to approach surgically, as the brain stem, lower cranial nerves vertebral artery and the occipital condyles, all hinder proper exposure. Multiple intracranial tumors represent 4 to 8% of all brain neoplasm.

However it is well recognized that the association of CPP and meningioma a rare event. Regarding pathogenesis, there are several hypothesis.one of which was the COHENHELM THEORY suggesting a multipotential oncogenic role of embryonal rests 6, 7. There was also a hypothesis proposing a genetic link between the two tumors 11, although, on a histopathological level, they seem to be different. However, under close scrutiny both of these hypothesis were refuted 2, 12.

Although, most authors consider the concurrence of meningiomas and CCP to be a more coincidence, the fact that a third of reported cases of the two tumors stated that they were in adjacent locations demands a better explanation. It's possible that the tumor occurring first could induce the formation of the other by acting as a local irritant 9, 10 having both tumors in close proximity to each other facilitates removal of both in one session.

Conclusions

This is a rare occurrence of concomitant meningioma and CPP. It has been speculated that the association of multiple different intracranial tumours might represent a "forme fruste" of central neurofibromatosis. But no proof of this hypothesis has been given so far. It also has been suggested that the meningioma itself might have acted as oncogenic stimulus to the development of the CCP tumor or "vice versa" in those cases, in which the two tumours are adjacently located. But the fact, that adjacent location occurs only in about one third of the reported cases, speaks against this assumption. It also cannot be supported by any other findings.

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