

Choroid Plexus Papilloma in Adult as a Cause of Sudden Death-A Rare Case Report and Review of Literature

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Abstract: Choroid plexus tumors are rare intracranial tumors which account for only 0.4-0.6 % of all brain tumors. These are intraventricular papillary neoplasm derived from choroid plexus epithelium and range from choroid plexus papilloma [World Health Organisation (WHO) grade I] to choroid plexus carcinoma (WHO grade III). Choroid plexus papilloma (CPP) is a rare, benign neoplasm, relatively more common in childhood. Sudden deaths have been reported, but are very unusual. Here we present a case of 21 year old male, brought for medico-legal autopsy examination on grounds of sudden death. He was reported to have headaches over a long period of time. On autopsy examination, diffuse subarachnoid haemorrhage was seen over bilateral cerebral hemispheres and cerebellum extending up to base of brain. On gross examination the right lateral ventricular cavity showed a friable grey brown lesion, revealing grey brown cut surface. On histopathological examination, it was found to be a choroid plexus papilloma grade I. Both cerebral hemisphere revealed marked congestion and sub-arachnoid haemorrhage. The present case reveals an unusual cause for sudden death in an adult male. The pathology is rare and a suspicion for this pathology in the adult male was not expected. So here we present a rare case with unusual presentation/location in an adult with review of literature.

Key Words: Choroid Plexus Papilloma, Sub-Arachnoid Haemorrhage, Intracranial Tumours,

Date of Submission: 27-07-2018

Date Of Acceptance: 09-08-2018

I. Introduction

Choroid plexus tumors are intravascular papillary neoplasms derived from choroid plexus epithelium and account for 0.4-0.6% of all brain tumors.[1]

They are relatively more common in childhood and constitute 1.5 to 4% of intracranial tumors. Choroid plexus papillomas are rare, accounting for less than 1% of all intracranial tumors in adults. [2]

Most of these lesions appear in children, mainly located in the lateral ventricles. Those from the fourth ventricle are more frequent in adulthood.[3] They are neuroectodermal in origin and similar in structure to a normal choroid plexus in the form of multiple papillary fronds mounted on a well vascularised connective tissue stroma. CPPs are often associated with a vascular stalk connected to the choroid plexus, allowing mobility within the ventricular system. Grossly, they may appear as reddish cauliflower like growths, which often become heavily calcified. [4]

The tumor's presence is often heralded by non-specific signs and symptoms of increased intracranial pressure. In adults, headache is the most common presenting symptom, which may be related to an alteration in head position. The tumor is liable to spontaneous hemorrhage, resulting in blood-stained or xanthochromic CSF. Hydrocephalus is a common complication which may result from direct tumor obstruction of the outlet of CSF or due to excessive production of CSF. [4] CT and MRI are the investigative procedures of choice in the evaluation of CPPs. Surgery with complete resection can be curative in papilloma with 5-year survival rates close to 100% and occasional recurrences.[3]

Case report:

A 21 years old male was brought for medico-legal autopsy examination on grounds of sudden death. He was reported to have found unconscious in washroom of hostel and was declared dead after unsuccessful resuscitation in casualty department. Nothing suspicious was found around him. He was known to complain of headache quite often. On autopsy examination, no injuries were seen on the body. On opening the cranial cavity,

duramater was markedly tense and congested. Massive subarachnoid hemorrhage was present all over on both cerebral hemispheres and cerebellum. Other abdominal and thoracic organs were normal and healthy. Different viscera were sent for chemical analysis and histopathological examination. Pathological findings: Grossly, the specimen of brain including right and left cerebral hemisphere and cerebellum, measured 17.0x15.0x8.0cms and weighed 1519gms. The external surface revealed diffuse areas of subarachnoid haemorrhage, more pronounced on fronto-basal and left temporo-parietal region and congested blood vessels[fig-1]. A friable grey brown lesion measuring 2.5x1.5x0.5cms was identified in right lateral ventricular cavity attached to tela choroidea. Cut surface was grey brown. Histologically, the lesion simulated the normal architecture of the choroids plexus. The papillary fronds showed fibrovascular cores. The epithelial cells lining the fronds were crowded and mildly pleomorphic. Numerous calcified psammoma bodies were also seen[Fig-2]. The histological diagnosis was choroid plexus papilloma, grade I. Sections from left lateral ventricular cavity were unremarkable. Microsections examined from representative portions of both cerebral hemisphere including fronto-basal, frontal, temporo-parietal and occipital region revealed marked congestion and sub-arachnoid hemorrhage. Microsections examined from right and left vertebral arteries, from the base, were unremarkable. Gross examination as well as microscopic findings from other organs did not reveal any significant pathological findings. Chemical analysis report of viscera was also nil.

II. Discussion

Choroid plexus tumors are rare intracranial tumors which account for only 0.4-0.6% of all brain tumors. These are intraventricular papillary neoplasms derived from choroid plexus epithelium and range from choroid plexus papilloma (WHO grade I) to choroid plexus carcinoma (WHO grade III).[5]

There is a regional difference between adult and childhood tumors with most choroid plexus tumors in children arising in the lateral ventricles and those in the adult more common in the fourth ventricle with a tendency to grow through the foramen of Luschka into the cerebello-pontine angle. [6] In the present case, CPP was seen in an adult male in the lateral ventricular cavity of brain.

The median duration of symptoms is reported to be about one month with approximately one-third of patients presenting within two weeks. [7] In the present case, as retrieved from friends and neighbours, the symptoms extended over a period of months.

The clinical progression is usually one of gradual deterioration. Complications that can occur with CPP include spontaneous hemorrhage from the tumor, dissemination of tumor fragments and hydrocephalus.[5] In adults, headache is the most common presenting symptom. In the present case also, off and on headache was the only symptom reported for months. The primarily intra-ventricular growth is responsible for the paucity of symptoms in the early stages of the disease. [8]

Choroid plexus tumor can also manifest, manifesting as anorexia nervosa in adult, may be a victim of diagnostic error.[9]

Grossly, choroid plexus tumors are generally described as a well-circumscribed, brownish-red, cauliflower-like mass, the carcinoma being invasive, appearing hemorrhagic or necrotic. Histologically, there is delicate connective tissue fronds covered by a single layer of cuboidal epithelium. The papilloma resembles normal choroid plexus but the cells are more crowded and elongated. The carcinoma is far more cellular and displays signs of anaplasia. Rarely, these tumors can exhibit mucinous degeneration, melanization, tubular glandular architecture or osseous and cartilage metaplasia. [8]

In adults, most CPP are heterogeneous secondary to cystic and /or calcific degenerations. The present case also showed calcification on microscopic examination, cystic change was not evident. There has been a case report of sudden death in which the tumor involved the third ventricle and caused acute ventricular obstruction. [10]

There is also a case report of sudden death in a adult with massive sub arachnoid haemorrhagediagnosed as cystic choroid plexus papilloma grade I of right lateral ventricle.[8]

In the present case, there was massive sub-arachnoid hemorrhage leading to sudden death. The pathology could have been diagnosed easily by CT scan or MRI. When diagnosed, it has good survival rate, the morbidity depends on the extent of pathological effects. In the present case, the tumor was designated as choroid plexus papilloma, Grade I (WHO). The case is being presented for its rarity and some features being distinct from expected findings in such a case.

III. Conclusion

The present case was adult, with the lesion in lateral ventricular cavity and occasional sign and symptoms of headache only. The pathology is rare, and a CT scan can diagnose the case and could be treated. Death could be preventable by surgical treatment. So the case is being presented for its rarity and some features being distinct from expected findings in such a case.

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- [11]. Figure No. 1 a, band c : External and cut surfaces of brain showing markedly tense and congested duramater. Massive subarachnoid haemorrhage present all over on cerebral hemispheres.
- [12]. Figure No. 2 a, b, c and d: Microscopic picture of CPP showing severe subarachnoid haemorrhage, complex branching papillary fronds with central fibro-vascular core (H & E low power view) and concentrically laminated psammoma bodies, choroid plexus papilloma in high power showing epithelial cells with mild nuclear atypia lining the papillary fronds (H & E low and high power view)

Pawan Singh "Choroid Plexus Papilloma in Adult as a Cause of Sudden Death-A Rare Case Report and Review of Literature." IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 17, no. 8, 2018, pp 65-67.