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Rapidly Progressive Dementia with Gait Ataxia

Vijay Shanmugam G, Aneesh Basheer, Sudhakar
From the Department of General Medicine, Pondicherry Institute of Medical Sciences, Ganapthychettikulam, Kalapet, Pondicherry, India.

Abstract:

Choroid plexus papillomas are rare neoplasms that arise from choroid plexus epithelium. Most of these lesions appear in children, mainly located in the lateral ventricles. We report a case of a 40 year old male with rapidly progressive dementia and gait ataxia who was found to have choroid plexus papilloma. This condition predominantly affects children and is mostly located in the lateral ventricles. Choroid plexus papilloma accounts for 0.5% of all brain tumors in adulthood with very few case reports in the world which have depicted this unusual presentation.

Key words: Choroid Plexus, Papilloma, Dementia, Gait Ataxia, Brain Neoplasms.

Introduction

Choroid plexus tumors are rare intra-ventricular papillary neoplasms derived from choroid plexus epithelium, which account for only between 0.4-0.6% of all intracranial and 2-3% of pediatric neoplasms [1]. Clinically, choroid plexus tumors tend to cause hydrocephalus and increased intracranial pressure [1]. Choroid plexus papillomas are twice as frequent as carcinomas [2]. It constitutes about 0.5 percent of the adult brain tumors [2].

Case Report

This 40 year old male a known case of psoriasis and type 2 diabetes mellitus, on methotrexate and metformin was brought with complaints of reduced word output and decline in immediate and recent memory with preserved remote memory over past 25 days. He also had swaying to either side while walking. His relatives gave history of reduced speech but no slurring or irrelevant speech. There was no history of fever or seizures or bowel and bladder involvement. Patient had no substance abuse but had a road traffic accident 6 months prior to the current admission. Neuroimaging done then was normal.

On examination, he was conscious, disoriented to time and place, and had reduced spontaneous speech. His Glasgow coma scale was 15/15 (E4M6V5). His attention was reduced and his mini-mental status examination revealed MMSE score of 23. His cranial nerve examination was normal. There was marked hypertonia with grade 4/5 power of upper limbs, and normal tone and power in lower

Corresponding Author: Dr. Vijay Shanmugam G
Email: gvijais85@gmail.com
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limbs. He had an exaggerated left biceps and supinator reflexes with left sided Babinski positive. Cerebellar signs were present in both upper and lower limbs with gait ataxia.

His blood panel revealed a normal study with normal electrolyte and liver enzyme levels. His retroviral status was negative. A computed tomography scan of brain revealed large lobulated heterogeneous lesions with dense calcifications and cystic areas in bilateral ventricles extending into the region of posterior third ventricle and pineal gland causing hydrocephalus suggestive of choroid plexus papilloma [Fig.1,2]. The bone window revealed an iso-dense Hounsfield unit of the hyper-dense region in the lesion when compared with the skull bone suggestive of calcification of the lesion [Fig.3]. Neurosurgery consult was taken and was advised surgery. However the patient was not willing for surgery and was referred to a neurorehabilitation centre.

Our case presented with one of the rare forms of neoplasms of the nervous systems. The presentation was deceiving and had various other differential diagnosis including normal pressure hydrocephalous and subdural hematoma and progressive supranuclear palsy. Moreover the clinical presentation in the form of rapidly progressive dementia and ataxia was also fairly uncommon.

Discussion

Neoplasms of the choroid plexus account for 0.4-0.6% of all intracranial tumors, 2-4% of pediatric

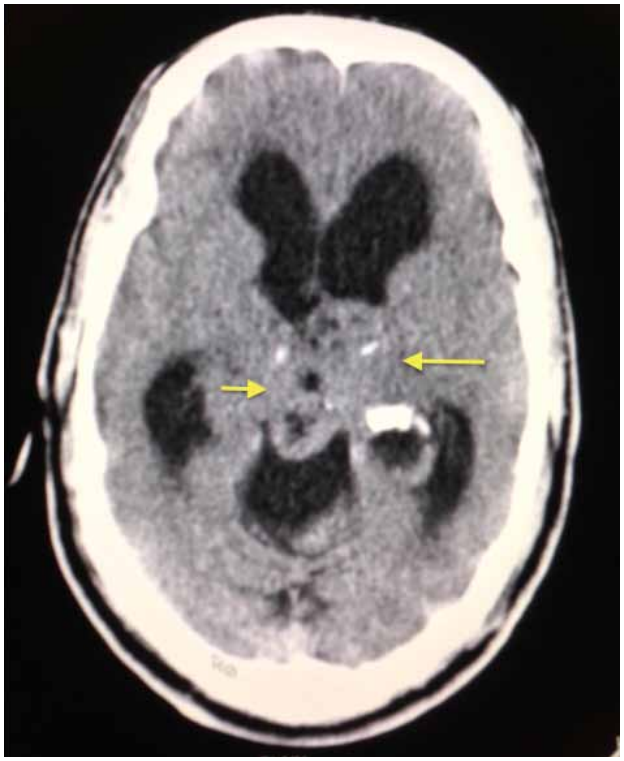

Fig.1: Large lobulated heterogeneous lesions with dense calcifications and cystic areas in bilateral ventricles.

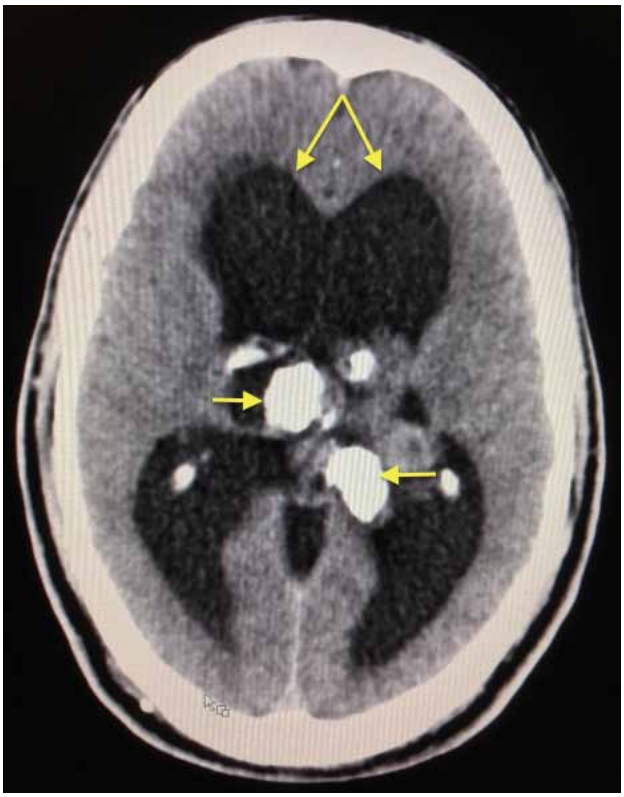

Fig.2: Bilateral calcified mass causing obstruction leading to hydrocephalus.

brain tumors, and 10-20% of brain tumors in children younger than 1 year of age [2,3]. Choroid plexus papilloma usually present with signs of raised intracranial pressure like headache, nausea, vomiting, drowsiness, ocular and gaze palsies, papilledema and blindness [4-6]. In rare circumstances patients present with seizure and psychosis [7-9]. On the contrary our case presented with sub-acute onset and rapidly progressive of dementia and gait ataxia which is an unique and rare presentation. This history, findings and the age of the patient held us in a diagnostic quandary. However the radiologic findings were conclusive and the diagnosis was established.

Tumors that arise in the lateral ventricle are much more common in patients 10 years of age or less, whereas those that arise in the fourth ventricle are fairly evenly distributed among patients 0-50 years of age [3]. In a study conducted by Taconni *et al* the main symptom of choroid plexus papilloma in adults was papilledema, which accounted for 63% and unsteady gait in children, which accounted for 71% [10]. On the contrary a study conducted by Ta Hsieh C *et al*. concluded that choroid plexus papilloma in the lateral ventricle presents with long history of headache, paresis, and cranial nerve palsies, and progressive visual loss is typical in adults [6]. Choroid plexus papillomas in the fourth ventricle or cerebellopontine angle, signs and symptoms of hydrocephalus accompanied by ataxia, lower cranial nerve palsies, and visual disturbances are the most common presentations in all age groups [6,11]. In our case there was neither cranial nerve involvement nor visual disturbances, however there was limited involvement of the pyramidal tract and dementia which is not very frequently accounted before.

The pathophysiology has been attributed to an obstruction to CSF flow by the tumor or the higher production (four to five times) of CSF by the stimulated tumor cells [6,11,12]. It is well documented that

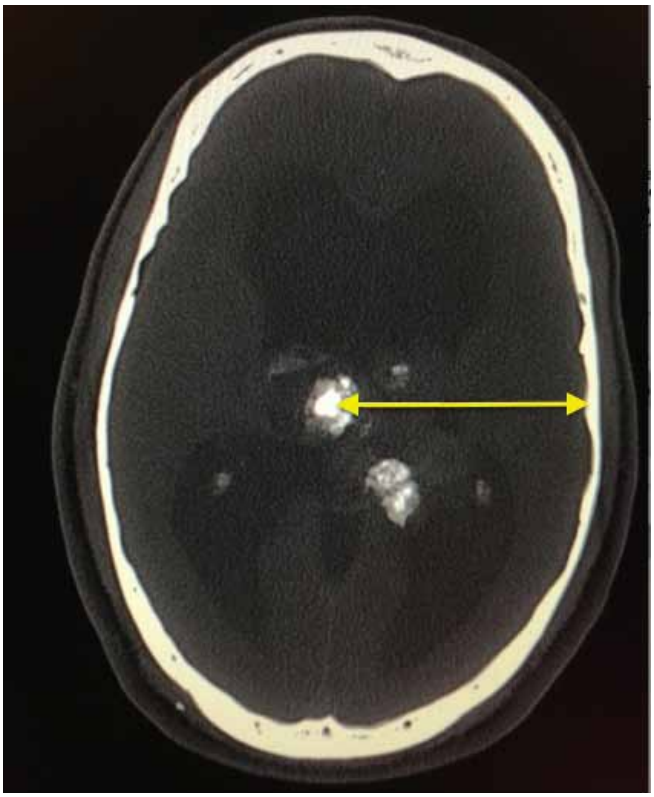

Fig.3: Bone window revealing a lesion of isodense Hounsfield with the skull bone suggestive of calcification.

choroid plexus tumors may produce CSF in amounts far exceeding the average of 450 mL per day that is normally observed [12,13]. In neuroimaging studies, a choroid plexus papilloma usually presents as a well-margined, smooth or lobulated, calcified, iso-dense or hyper-dense mass on pre-contrast computed tomography (CT) and shows dense and usually homogeneous enhancement on post-contrast CT [6,12]. Choroid plexus tumors are soft well-circumscribed cauliflower-like masses with prominent lobulations peripherally. Hemorrhage and cyst formation may be seen. Necrosis and parenchymal invasion are characteristic features for choroid plexus carcinoma [14,15]. Those tumors that have a pedicular attachment may move within the ventricle, giving rise to acute gravity-dependent intermittent ventricular obstruction, and have been associated

with the bobble-head doll syndrome in some cases [15,16].

This case highlights a very rare benign intracranial neoplasm in an adult with a unique presentation of rapidly progressive dementia. This reiterates the role of neuroimaging in cases of cognitive dysfunction even in elderly where it need not always be neurodegenerative disorder.

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