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Case Report

An unusual case of stroke in a young Indian male, presenting with recurring hemiparesis-diagnosis aided by the use of susceptibility weighted magnetic resonance imaging and computed tomography

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ABSTRACT

Germinoma is the most common and least malignant intracranial germ cell tumour, usually found in the midline. Basal ganglia and thalamic germinoma, called ectopic germinoma, is a rare and well documented entity representing 5% to 10% of all intracranial germinomas and has been reported extensively in young Asian males. The association of cerebral and/or brain stem atrophy with basal ganglia- thalamic germinoma on computed tomography (CT) and magnetic resonance imaging **(**MRI) is found in 33% of the cases. Although early treatment with chemo radiotherapy can be curative and significantly improve the quality of life, medical attention is often delayed due to the unusual imaging morphology of these lesions. We describe a case of a young 19-year-old Indian male who presented initially with recurring monoparesis after an episode of trauma, which then progressed slowly to hemiparesis and unusual features over a period of 2 years. The initial and subsequent imaging findings are described in this article and the value of susceptibility weighted MRI and CT imaging in arriving at the diagnosis is highlighted. The diagnosis was proven by positron emission tomography/CT (PET-CT) and biopsy, following which the patient was started on chemo-radiotherapy and is under follow up.

Keywords: Basal ganglia-thalamic, Germinoma, Hemi atrophy, Susceptibility weighted imaging

INTRODUCTION

Germinomas, which are believed to arise from the primordial germ cells, are the least malignant and most common of all germ cell tumors, and most of these tumors are found at the midline in the pineal or suprasellar regions.[1]

Germinoma arising in the basal ganglia-thalamus is a rare, well documented entity representing 5% to 10% of all intracranial germinomas.[2,3] There is a higher incidence in Asian populations, with an overall male predominance, and it usually occurs in the second decade of life.[4] There have been limited case reports in Indian males, with one of the cases being a bilateral lesion.[5-7] A clinical history of slowly progressive hemiparesis and associated neuropsychiatric symptoms such as dementia, psychosis and cognitive decline are commonly observed. However, presentation as recurring neurological illness mimicking stroke in young has been rarely described. The association of cerebral and/or brain stem hemi atrophy with basal ganglia germinoma (BGT) on CT and MRI is found in up to 33% of the cases and correlates with clinical features, although its aetiology is unclear.[1,3] Typical radiological features of advanced disease are well described and easily diagnosed due to formation of a large mass, but MRI revealing the presence of hemi cerebral atrophy, a reliable early sign, and use of susceptibility weighted MRI to aid in early diagnosis are not commonly described.[8]

Only 3 cases in Indian males have been reported previously and in this report of an Indian adolescent, we

would like to highlight the use of susceptibility weighted imaging and CT imaging which aid in the early diagnosis of difficult cases.

CASE REPORT

A 19-year-old Indian male sustained trauma with head injury, left tibial fracture in a minor road traffic accident in August 2017 at 17 years of age. He was put in a cast and on removal after 4 weeks, he was noticed to have left dragged gait. After a month he developed headache with persistent monoparesis, for which the initial MR imaging was performed elsewhere (Figure 1 A). It was reported as a right gangliocapsular subacute infarct, and the patient was investigated with a CT angiogram of the head and neck vessels and cardiac 2D echocardiogram to assess for the cause of stroke, both of which were normal. The patient was started on antiplatelets and was on follow up and physiotherapy. Later on, in the month of November 2017 he developed left sided focal motor seizures with residual hemiparesis. A diagnostic cerebral digital subtraction angiography (DSA) revealed no abnormalities. Over the next 2 years, the left hemiparesis had progressed and he presented to us with diplopia, ptosis, and focal seizures with secondary generalization. He had a repeat MRI done elsewhere (Figure 1 B) just before presentation, which was interpreted as gliotic and old haemorrhagic changes in the right gangliocapsular and thalamic region with right sided brainstem Wallerian degeneration. We repeated the MRI due to unusual features, along with susceptibility weighted sequences (Figure 1 C), which we interpreted as a diffuse ill-defined lesion in the right basal ganglia and thalamus with "blooming" on SWI, associated with left hemi cerebral and left brainstem atrophy. CT imaging (Figure 1 D) further aided in the diagnosis since, the lesion appeared as an ill-defined hyper dense mass in the basal ganglia and thalamus and did not show expected features of gliosis. For confirmation, patient underwent 18F-DOPA PET CT (Figure 1 E), which showed increased uptake by lesion in right basal ganglia and thalamus. This was followed by stereotactic assisted biopsy, with confirmation of diagnosis on histopathology, which showed tumour cells with large, round nuclei with prominent nucleoli, frequent mitoses and varied cytoplasm which was predominantly eosinophilic (Figure 1 F).

As regards treatment, he underwent chemotherapy with 5 cycles of cisplatin, etoposide, followed by radiotherapy. Following, chemo radiation, the patient was in remission with regular follow up and rehabilitation and is doing well on 5 year follow up clinically.

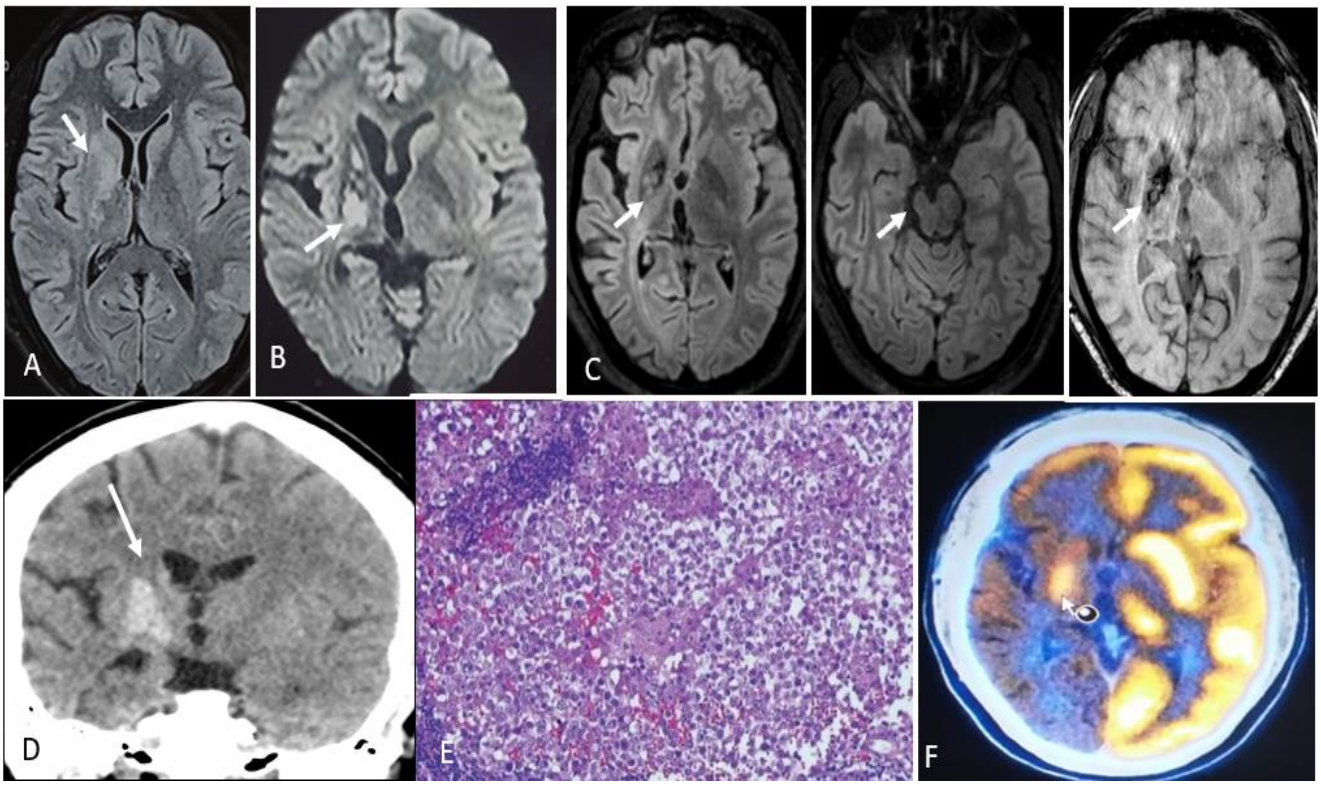

Figure 1 (A-F): A-Axial T2FLAIR, 2017-(white arrow) right basal ganglia signal change labelled as infarct. B-Axial T2 FLAIR, 2018-(white arrow), gliotic changes with haemorrhage in right basal ganglia. C-Axial T2FLAIR (2 sections), and Axial SWI done at our institute (white arrows) right basal ganglia lesion with right cerebral peduncle atrophy. Lesion shows "blooming" on SWI (white arrow). D-Coronal CT scan shows hyper dense lesion right basal ganglia (white arrow). E-H and E stained HPE specimen of the lesion showed tumour cells with large round nuclei and nucleoli with frequent mitosis. F-Axial DOPA PETCT (arrow) hypermetabolic lesion right basal ganglia, with right cerebral diffuse hypo metabolism.

DISCUSSION

Germinomas, although common, are the least malignant intracranial germ cell tumour, usually found in the pineal and suprasellar region and are thought to arise from a midline stream of Toti potential cells very early in the period of rostral neural tube development.[9] Primary thalamic and BGT, also called ectopic germinoma, have been described in the literature, occurring in 5% to 10% of all intracranial germinomas, and they are more common in Asian populations, with only 3 case reports in Indian males.[3,5-7] They tend to have a protracted clinical presentation, dominated by progressive hemiparesis, cognitive decline and psychosis, resulting in delayed diagnosis. On the contrary, our index case, had recurrent episodes of neurological dysfunction mimicking "stroke in young" with preserved cognition. This had led to probable delay in diagnosis and on presentation to us, the interesting clinical features and hemi atrophy of brain stem led us to clinch the diagnosis. All the cases described in literature also had hemispheric and brainstem atrophy and it has been proposed, that atrophy occurs as a result of tumour involvement of the internal capsule fibres or thalamic ganglion cells with subsequent interruption of thalamo-cortical connections.[1,10]

Early recognition of BGT germinoma is important because it is highly radiosensitive and potentially curable by RT alone, if correctly diagnosed at its early stage.[11] BGT germinoma is one of the differential diagnoses when a lesion in the BGT is ill-defined, hyper intense on T2WI, and without mass effect or prominent contrast enhancement in young patients. However, these changes are subtle and inconspicuous on conventional MR imaging, and if germinoma is not suspected initially, then lesions could progress to form a large mass.[3,8]

SWI exploits the magnetic-susceptibility effects generated by local inhomogeneity of the field. It can be used to detect blood products and biologic metal accumulation and provides additional information in brain tumour evaluation, which is valuable in BGT germinoma due to its characteristic intra tumoural haemorrhage.[12] Although early BGT germinomas have some distinct characteristics, (i.e. hyper intense T2 signal intensity and no enhancement), they are more strikingly visualized and appear larger on SWI sequences than on conventional MRI.[11]

In addition to intra-tumoural haemorrhage, signal-intensity change of germinomas on SWI may also be explained by 2 other mechanisms: First, BGT structures, especially the internal capsule and globus pallidus, are rich in biologic metals such as iron. Tumour invasion may lead to disruption in axonal transport of transferrin, resulting in abnormal iron accumulation. Second, it is possible that transferrin secreted by the tumour cells binds to transferrin receptors on neurons and glial cells, which are then transported into the globus pallidus.[12]

CONCLUSION

Primary thalamic and BGTs have an unusual clinical presentation of progressive hemiparesis. Although this entity has been extensively reported in Asian males, only 3 reported cases in Indian males exist upon review of literature. These lesions show a unique constellation of imaging findings of progressive cerebral and brainstem hemi atrophy associated with an ill-defined non-mass-like lesion in the thalamic and basal ganglia region which shows T2 shortening on susceptibility weighted imaging. In our case, the patient had undergone two prior MRI studies which did not include SWI sequences and hence the T2 shortening/ "blooming" property of the lesion was not assessed adequately. On the MRI done at our institute, the SW images showed pronounced T2 shortening within the lesion, which prompted us to also perform a CT which showed an ill-defined hyper dense mass in the right BGT region. This led to a relatively early diagnosis since the primary lesion was still small in size (approximately 12-15 mm) which was further confirmed on PET-CT and on biopsy, and the patient received adequate treatment.

The knowledge of these key points aids in the early diagnosis of these lesions which are known to be highly radiosensitive, thereby leading to a favourable outcome.

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