# Fourth Ventricular Medulloblastoma – A Foremost case in ISTH Irrua

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## ABSTRACT

**Introduction**: Medulloblastoma (MB) though most common malignant brain tumor affecting children and are usually high grade tumours, are only metastatic within the central nervous system and rarely, beyond the neuraxis. This case is presented as the first ever seen in our department despite been in operation in about two decades. The traditional therapeutic mainstay for medulloblastoma includes a multimodal approach which include surgery, radiation, and multiagent chemotherapy. As we discover more about the molecular basis of medulloblastoma, efforts to adjust treatment approaches based on molecular risk stratification are under active investigation. Certainly, the known neurological, developmental, endocrine, and psychosocial injury related to medulloblastoma and its associated therapies motivate ongoing research towards improving treatment for this life-threatening tumor while at the same time minimizing long-term side effects.

**Case Presentation**: We present the case of an 11year old boy who presented at the neurosurgical unit of our hospital with cerebellar physical symptoms of gait problems without IQ sequelae and had CT scan and MRI radio-diagnosis of a 4$^{th}$ Ventricular tumour probably a Medulloblastoma. He subsequently had surgery and histologically, a diagnosis of Medulloblastoma, desmoplastic variant, W.H.O Grade 4 was made.

**Conclusion**: Medulloblastoma according to the 2016 WHO Classification of CNS tumour as a W.H.O Grade 4 tumour with malignant features and tendency. Therefore, prompt and early diagnosis and proper management including financial and/or health insurance is essential to achieving a good outcome.

**Keywords**: Medulloblastoma, WHO, Laminectomy, CNS, weighted images

**Medulloblastoma – A case report of an 11year old boy**

## INTRODUCTION

Medulloblastoma is said to be the most common paediatric malignant tumour of the central nervous system (CNS) accounting for about 20% and 40% of tumours in the posterior fossa.[1] It occurs most commonly in the fourth ventricle and cerebellar parenchyma but sources have reported rare sites such as the meninges of the brain.[2,3] The peak incidence is between five and nine years.[4] Overall ratio tend to be 1.5:1 for males and they also tend to have poorer prognosis.[1] In terms of morbidity, hydrocephalus seems to be the most common complication. Others are cerebellar dysfunction and leptomeningeal dissemination. Cognitive and IQ symptoms and sequelae could follow the presenting complains and improve following treatment respectively.

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Research suggests that attention disorders, working memory disorders and lower general IQ might occur as the most crucial long-term cognitive symptoms.[1,12]

We report a case of 4th ventricular medulloblastoma in an 11-year-old male. The clinico-radiologic diagnosis was that of a 4th ventricular tumour (? medulloblastoma). The diagnosis of medulloblastoma was confirmed on histology following complete excisional biopsy though patient succumbed to the stress of surgery and other negating parameters as regards CNS tumours generally. This is the first of such case seen in our practice and our hospital hence the interest to report it.

**Case Presentation**

Master E.E was an 11-year-old school boy who presented to the neurosurgical clinic of the Irrua Specialist Teaching Hospital with complaints of a two-week history of progressively worsening unsteadiness of gait, not favoring any particular side, and a ten-day history of slurred speech. He reported no headache, seizures, nausea or vomiting, focal body weakness or visual difficulties. Academically, he had been performing well above average.

On examination, he was fully conscious with stable vital signs. Pupillary size and reactivity to light were normal, as were his visual acuity and fundoscopy findings, with no evidence of nystagmus. His speech was slurred, and he exhibited cerebellar signs, including ataxic gait, dysmetria, and bilateral dysdiadochokinesia.

An initial cranial CT scan available at the time of presentation showed a large enhancing hyperdense mass in the midline in the fourth ventricle with mass effect on the brainstem and the cerebellum. There was mild hydrocephalus

Follow-up brain MRI **(Fig.1)** identified a T1 hypointense, T2 heterointense lesion in the fourth ventricle with minimal contrast enhancement; and with mass effect on the brainstem and in the cerebellum. It showed restricted diffusion seen as high DWI signal and correspondingly low ADC values. Hydrocephalus was absent.

The clinico-radiological diagnosis was that of a fourth-ventricular tumour, likely medulloblastoma.

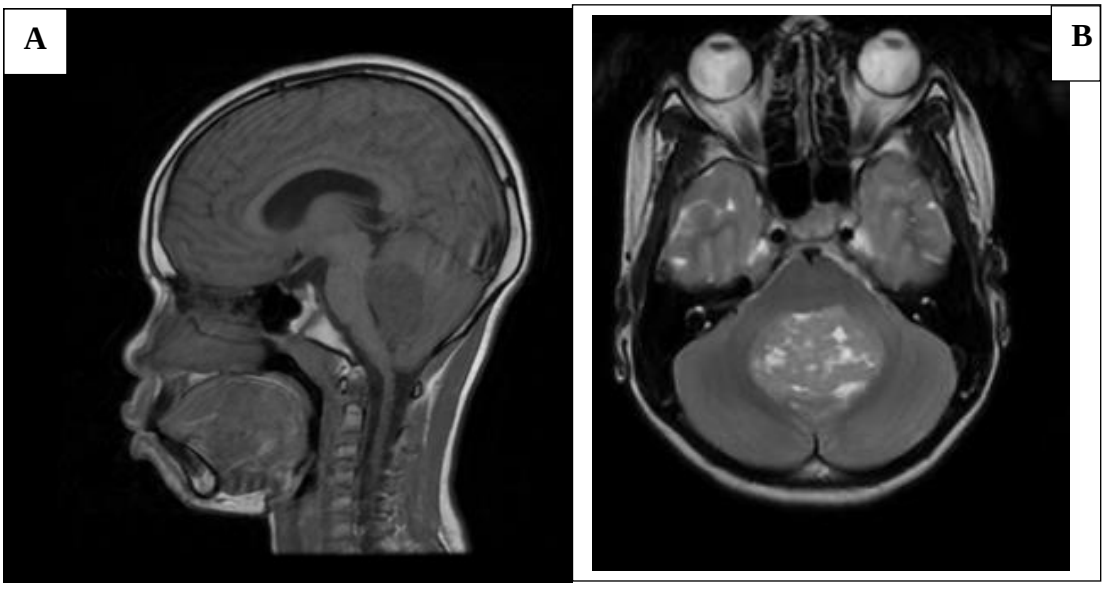


Fig. 1: MRI images showing T1 hypointense (A); T2 heterointense (B) lesion in 4th ventricle.

The patient underwent a suboccipital craniotomy with C1 laminectomy and gross total excision of the fourth ventricular tumour through a telovelar approach. During surgery, the tumour, which completely filled the fourth ventricle, was observed to be pinkish, soft and suckable with areas of haemorrhage. A sample was sent for histopathological analysis.

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Histology report **(Fig.2)** described a small round cell lesion composed of densely packed undifferentiated cells disposed within scanty hyalinized fibrocollagenous connective tissue stroma. The tumour cells have mildly pleomorphic hyperchromatic nuclei with scanty eosinophilic cytoplasm. There are few atypical mitotic activities foci of vague Homer-Wright rosettes and apoptotic bodies seen **(Fig. 2).** A diagnosis of medulloblastoma, desmoplastic / nodular variant, W.H.O Grade 4 was made.

Postoperatively, the patient was transferred to the ICU for elective ventilation with paralysis and sedation. Although his immediate postoperative condition was stable, he unfortunately deteriorated unexpectedly and passed four days following surgery.

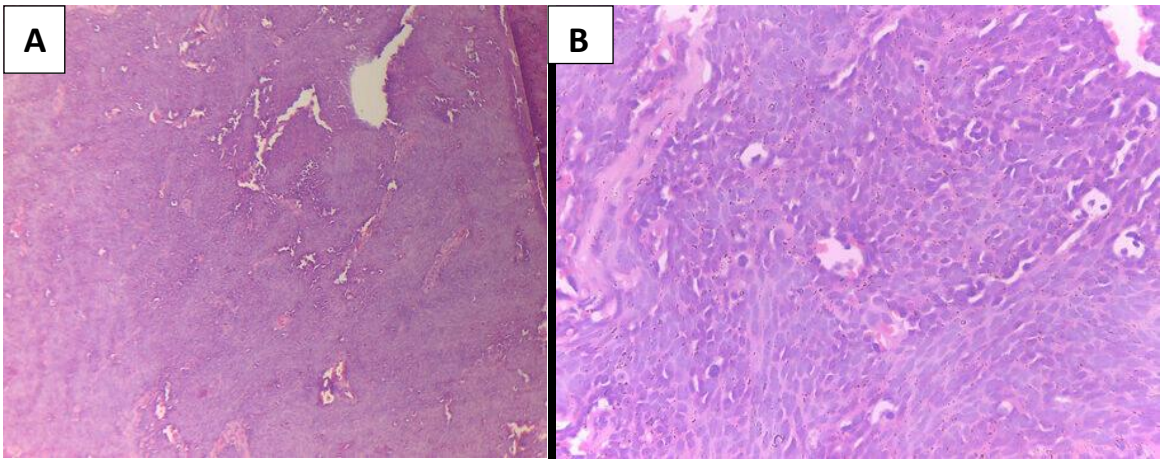


Fig. 2: H & E stained Photomicrograph showing small round blue cells arranged in syncytium with vague Homer-Wright Rosettes (seen better in B)

Table 1: Comparing and contrasting some clinical features of Medulloblastoma as seen in children versus index case.

| S/N | **Clinical Feature** | As seen in children | As seen in index case |
|---|---|---|---|
| **1.** | **Age at Presentation** | 5 – 9 years | 11 years |
| **2.** | **Gender preponderance** | Commoner in males | Patient is male |
| **3.** | **Symptoms** | Commonest being headaches then gait abnormalities | Presented with only gait abnormality |
| **4.** | **Slurred speech** | Not commonly present | present |
| **5.** | **IQ sequelae** | Commonly present | Absent |

## DISCUSSION

Medulloblastoma (MB) is a primitive neuroectodermal tumour accounting for approximately 30% of paediatric tumours and 7-8% of intracranial tumours including those in the posterior fossa of the brain.[1] The mean age of affectation is around 9 years with peak incidence of 3 – 7 years. However, a second peak occurs in adulthood accounting for about 25% of all cases.[5,6]

MB may be seen as a component of nevoid basal cell carcinoma syndrome i.e Gorlin syndrome, Li-Fraumeni syndrome, Turcot syndrome, Rubinstain-Taybi syndrome and Nijmegen breakage syndrome.

MB was thought to have originated from “medulloblast”, an undifferentiated cell within the ependymal lining of the ventricular system.[4] Some sources have claimed that it was first described in 1925 but added in to the

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histogenetic classification scheme by Baily and Cushing later on based on the morphologies of the cells of the developing brain.[4]

It is an embryonal neuroepithelial tumour arising most likely in the cerebellum (roof of fourth ventricle, midline of the cerebellum, the vermis, and cerebellar hemispheres) as well as the dorsal brain stem consisting of densely packed small round undifferentiated cells with mild to moderate nuclear pleomorphism and a high mitotic count.[1,2-4] Our patient had a tumour completely filling up the 4th ventricle however other symptoms of gait disturbances may have alluded to the fact that there may be cerebellar involvement especially the brain stem but visualized as a mild mass effect which was not sufficient enough to cause other clinical effects **(Table 1).**

These tumours can grow rapidly leading to obstruction of the flow of CSF and ultimately hydrocephalus[2,5] Our patient had no feature to suggest hydrocephalus despite involvement of the pathway of the flow of CSF. This may be due to the fact that the lateral ventricles not involved with the tumour may have compensatory outflow. It can also spread locally through the CSF to affect the meninges, ventricles and subarachnoid space including distant involvement of extraneural sites such as the lungs, liver, vertebrae and pelvis.[4]

Medulloblastoma arises from remnants of the primitive neuroectoderm in the roof of the fourth ventricle. It grows in cerebellar vermis and fills the ventricle, often invading through the ependyma in the floor of the ventricle to enter the brainstem. Less commonly, the tumor arises in the cerebellar hemisphere.[1,2,9] It's pathogenesis have been linked to deletions of 17p and isochromosome 17q. Genomic wide studies have revealed alterations in signaling pathways involved in normal cerebellar development, such as the sonic hedgehog-patched (SHH) and Wnt/ß-catenin pathways. Fourth ventricular as well as middle cerebellar peduncle tumours are often associated with Wnt pathway alteration while tumours in the vermis or lateral cerebellum are usually associated with SHH pathway.

According to the W.H.O Classification of CNS Tumours 2016, MB have been identified which have been regarded as a Grade IV (malignant) tumour and this has prognostic, therapeutic and clinical significance. [2,3,7] MB can be histologically classified as classic MB, desmoplastic/nodular *(DN)* MB , MB with extensive nodularity *(MBEN)* and anaplastic/large cell *(LCA)* MB.[5,7] The histopathological classification remains relevant due to its clinical utility especially where genetic analysis is limited or not feasible. These have been incorporated into the molecular classification according to the W.H.O as WNT activated MB (classic & large cell – rarely), SHH-activated - TP53 mutant MB (large cell & desmoplastic – rarely), SHH-activated - TP53 wild type (classic, desmoplastic, extensive nodularity, MB (large cell & desmoplastic – rarely), non-WNT/non-SHH group 3 (classic and *LCA*) and non-WNT/non-SHH group 4 (classic and *LCA*). Sometimes, smaller biopsies especially from the fourth ventricle may have difficulty in classifying the tumour. These are termed MB Not Otherwise Specified (NOS).[4,5,7]

The 5-year overall survival for MB is approximately 75%. However, long-term therapy-related morbidity remains a significant concern despite advances in the biology and genomics of this tumour earning it as one of the prototypes in clinicopathologic diagnosis and management of this disease.[4,5] The boy succumbed four (4) days following surgery and post mortem examination was denied for cultural reasons.

Diagnosis of MB though largely histological, radio-diagnostic significance in determining type, site and operability of the lesion cannot be overemphasized. The diagnostic efficiency of MRI in solid CNS tumours especially posterior fossa tumours like MB could be as high as 75%.[8] MB on MRI shows restriction in diffusion weighted images.[8] This imaging was corroborated in our patient.

Like most other reported cases, the index case was in the cerebellum – 4th ventricle which is one of the commonest sites for its location. Though, some of them have been reported to have symptoms of raised intracranial pressure, gait and cognitive, features of obstructive CSF drainage leading to hydrocephalus as well as intellectual dysfunction. This one, however, presented with mainly gait problems.[1] This patient however did not have features of hydrocephalus **(Table 1).**

Clinically, the differential diagnoses often considered in such lesion as this include ependymoma, atypical teratoid /rhabdoid tumour, embryonal tumour with multilayered rosettes amongst other rare CNS tumours. In ependymoma which is the closest differential, the tumour is mainly cerebral or in the spinal cord and composed of monomorphic round to oval cells that is sharply circumscribed.[2-5,9-11] Other differentials too share similar features as MB but mainly supratentorial as well as in ages much less than the index case. This is where a good knowledge of the clinical, radiologic as well as histology becomes invaluable in clinching the diagnosis. Once diagnosis is confirmed, definitive treatment is mainly surgical with attendant adjuvant therapy after subsequent MRI image monitoring as well as the improvement in clinical outcomes.[1,9] The current case had a laminectomy and passed four days post operatively after diagnosis was confirmed.

## CONCLUSION

This patient did not have hydrocephalus which is the commonest presentation and also was there any sequelae to his academic performance or his IQ.

We conclude that this case was aptly diagnosed in our facility and confirmed with histology which is the foremost in our more than 25years of existence.

The patient did not have chemotherapy, follow-up MRI, immunohistochemistry and other ancillary investigations like molecular studies due to financial constraints and not on any insurance cover. These may have contributed to the demise of the patient as well as cultural beliefs necessitating the parents to decline autopsy.

We also advocate for social and/or health insurance to cover rural communities such as ours which its coverage to include everyone and not only to government workers.

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