Annals of Short Reports and Clinical Images

Clinical Image

Group 4 Medulloblastoma

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Clinical Image

A 7 years old girl presented with occipital headache, vomiting and gait unsteadiness of 1 month duration. On examination she had truncal ataxia, cerebellar signs and papilledema. On Magnetic Resonance (MR) imaging, tumor was seen arising from the vermis extending into the fourth ventricle. It was hypointense to isointense on T1WI (Figure 1A), hyperintense on T2WI (Figure 1B) with mild homogenous enhancement on contrast (Figure 1C and D). The superior recess of the fourth ventricle was dilated (Figure 1C). The lateral ventricle and third ventricle were dilated with periventricular ooze. There was no evidence of leptomeningeal dissemination. The tumor was excised totally. Histopathology was reported as classical medulloblastoma. Molecular analysis of the tumor tissue was consistent with Group 4 Medulloblastoma. Postoperative she received craniospinal radiation - 36 Gy in 20 fractions @ 1.8Gy/# to the craniospinal axis followed by 18 Gy boost to posterior fossa. 1 year later she presented with recurrent headache and gait unsteadiness. MR imaging of the brain showed tumor recurrence at the vermis (Figure 2A) with concurrent nodular metastasis at suprasellar, pineal region and ependymal lining of lateral ventricles (Figure 2B-D). Mismatching pattern of enhancement was seen between the recurrent tumor and nodular tumor deposits. The metastatic tumor deposits demonstrated intense enhancement on contrast. However the recurrent vermian tumor demonstrated mild enhancement on contrast similar to the primary tumor (Figure 2B-D). Above imaging features are classical of Group 4 medulloblastoma with metastatic deposits. Medulloblastoma is the most common primary malignant tumor in children accounting for 20%-25% of all childhood brain tumors [1]. Medulloblastoma is now no longer considered a single entity. It represents a heterogeneous group of diseases comprising four different molecular subgroups as per 2016 WHO classification. Each subgroup has distinct developmental origins, unique transcriptional profiles, diverse phenotypes and varying clinical outcomes [2]. Preoperative MR imaging can predict the molecular subgroup of medulloblastoma accurately based on specific imaging features [3]. Predicting the molecular subgroup of medulloblastoma on imaging can serve as independent predictive and prognostic biomarker [3]. Group 4 medulloblastomas have very specific imaging features. These tumors arise from the inferior medullary velum with resultant dilation of the superior recess of the fourth ventricle [1-3]. They demonstrate very minimal or no enhancement [1,2,4,5]. Medulloblastomas are known to have leptomeningeal metastasis through CSF spread. Lumbosacral drop metastasis in the spinal subarachnoid space is commonly reported in Group 3 medulloblastoma [1,5]. However Group 4 medulloblastomas often have supratentorial metastasis involving suprasellar and/or infundibular recess of the third ventricle, pineal recess and ependymal lining of the ventricles [1]. Metastasis in these recess is considered to be specific marker of group 4 medulloblastoma [5,6]. Metastatic deposits from group 4 tumors are more nodular and associated with a "mismatching pattern" i.e. lack of or minimal contrast enhancement but restricted diffusion [5]. In the present case all the MR imaging features of the primary, recurrent tumor with metastatic deposits were classical of Group 4 medulloblastoma. Predicting the molecular subgroup of medulloblastoma based on preoperative MR imaging enables prognostication of the tumor biology and behavior.

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Citation: Sachin Baldawa. Group 4 Medulloblastoma. Ann Short Rep Clin Image. 2021; 3(1): 1022.

Publisher Name: Medtext Publications LLC

Manuscript compiled: Oct 07th, 2021

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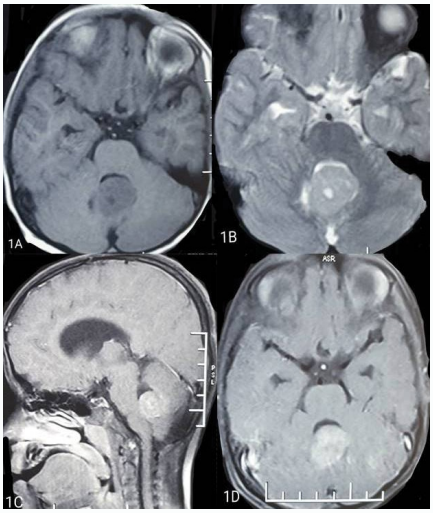

Figure 1: MR imaging of the brain demonstrated the tumor arising from the vermis inferiorly extending into the fourth ventricle. It was hypointense to isointense on T1W1 (1A), hyperintense on T2WI with mild homogenous enhancement on contrast (1C, 1D). The superior recess of fourth ventricle was dialated (1C).

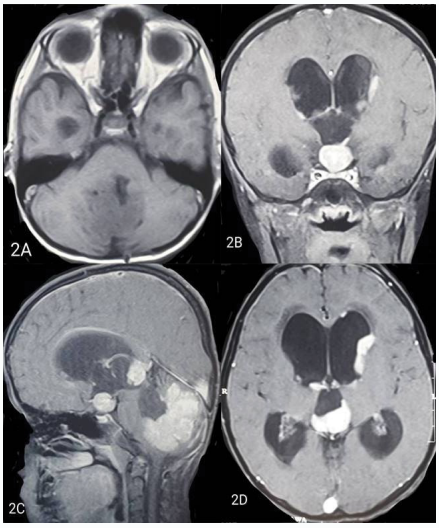

Figure 2: MR imaging of the brain showed tumor recurrence at the vermis (2A) with nodular metastasis at suprasellar, pineal region and ependymal lining of lateral ventricles (2b, 2C, 2D). Mismatching pattern of enhancement was seen between the recurrent tumor and nodular tumor deposits. The metastatic tumor deposits demonstrated intense enhancement on contrast. However the recurrent vermian tumor demonstrated mild enhancement on contrast similar to the primary tumor (2B, 2C, 2D).