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Asymptomatic Medulloblastoma Presented as Spinal Cord Lesion - An Unusual Presentation

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Abstracts: ***Medulloblastoma the most common primary brain tumor in children presents clinically with signs and symptoms related to increased intracranial pressure due to blockage of the fourth ventricle. We are presenting this case due to its unusual presentation in which a four and half years old male child presented with multiple swellings in the spinal region without any classical signs and symptoms of medulloblastoma and diagnosed as medulloblastoma histologically after surgery.***

Keywords: Medulloblastoma, CNS tumor, Brain tumor, Tumor in children, Drop metastasis

1. Introduction

Medulloblastoma is the most common type of primary brain cancer in children [1]. Medulloblastoma constitutes 20% of theMalignant brain tumors which are the leading cause of cancer death among pediatric patients [2].Medulloblastoma, a small blue-cell malignant tumor which arises in the posterior cranial fossa and in the cerebellum, mainly in the hemispheres and the vermis.

Signs and symptoms are mainly due to secondary increased intracranial pressure due to blockage of the fourth ventricle and tumors are usually present for one to five months before the diagnosis is made. The child typically becomes listless, with repeated episodes of vomiting, and a morning headache, which may lead to a misdiagnosis of gastrointestinal disease or migraine. [3]

We are presenting the case due to its unusual presentation as multiple swelling in the spinal region without any classical signs and symptoms of medulloblastoma.

2. Case Report

A four and half years old male child presented with multiple swelling in spinal regions in neurology OPD. On CT Scan of the spine, multiple intradural and extramedullary tumors were seen at the Lumbar and Thoracic region of the spinal cord. It was clinically diagnosed as Schwannoma. After surgery sample sent for histopathology study. Gross was greyish-white friable tissue measuring 2x2x1 cm. Microscopy findings-On [H&E X100] Small undifferentiated cells arranged in swirling and fascicular architecture. Nuclei are densely hyperchromatic, angulated with scanty cytoplasm. On [H&E X400] pale islands of cells with nuclear pleomorphism are seen. Microscopic Features were consistent with Medulloblastoma, so after one month when CT Scan of Brain was done, a posterior fossa tumor was diagnosed with drop metastasis to the Spinal cord.

3. Discussion

Medulloblastoma is the most common type of primary brain cancer in children. It originates in the part of the brain that is towards the back and the bottom, on the floor of the skull, in the cerebellum, or posterior fossa. [4] Historically medulloblastomas have been classified as a primitive neuroectodermal tumor (PNET), but it is now known that medulloblastoma is distinct from supratentorial PNETs and they are no longer considered similar entities. [5] **Medulloblastomas are invasive, rapidly growing tumors that, unlike most brain tumors, spread through the cerebrospinal fluid and frequently metastasize to different locations along the surface of the brain and spinal cord. Metastasis down to the cauda equina at the base of the spinal cord is termed "drop metastasis".**

Medulloblastomas usually found in the vicinity of the fourth ventricle, between the brainstem and the cerebellum. Tumors with similar appearance and characteristics originate in other parts of the brain, but they are not identical to medulloblastoma. [6] Medulloblastomas are thought to originate from immature or embryonal cells at their earliest stage of development, the cell of origin depends on the subgroup of medulloblastoma. WNT tumors originate from the lower rhombic lip of the brainstem, while SHH tumors originate from the external granular layer. Medulloblastoma is composed of four distinct molecular and clinical variants termed WNT/β-catenin, Sonic Hedgehog, Group 3, and Group 4. [7]Of these subgroups, WNT patients have an excellent prognosis, and group 3 patients have a dismal prognosis. Medulloblastomas are also seen in Gorlin syndrome as well as Turcot syndrome. Recurrent mutations in the genes *CTNNB1*, *PTCH1*, *MLL2*, *SMARCA4*, *DDX3X*, *CTDNEP1*, *KDM6A*, and *TBR1* were identified in individuals with medulloblastoma. [8] Additional pathways disrupted in some medulloblastomas include MYC, Notch, BMP, and TGF-β signalling pathways.

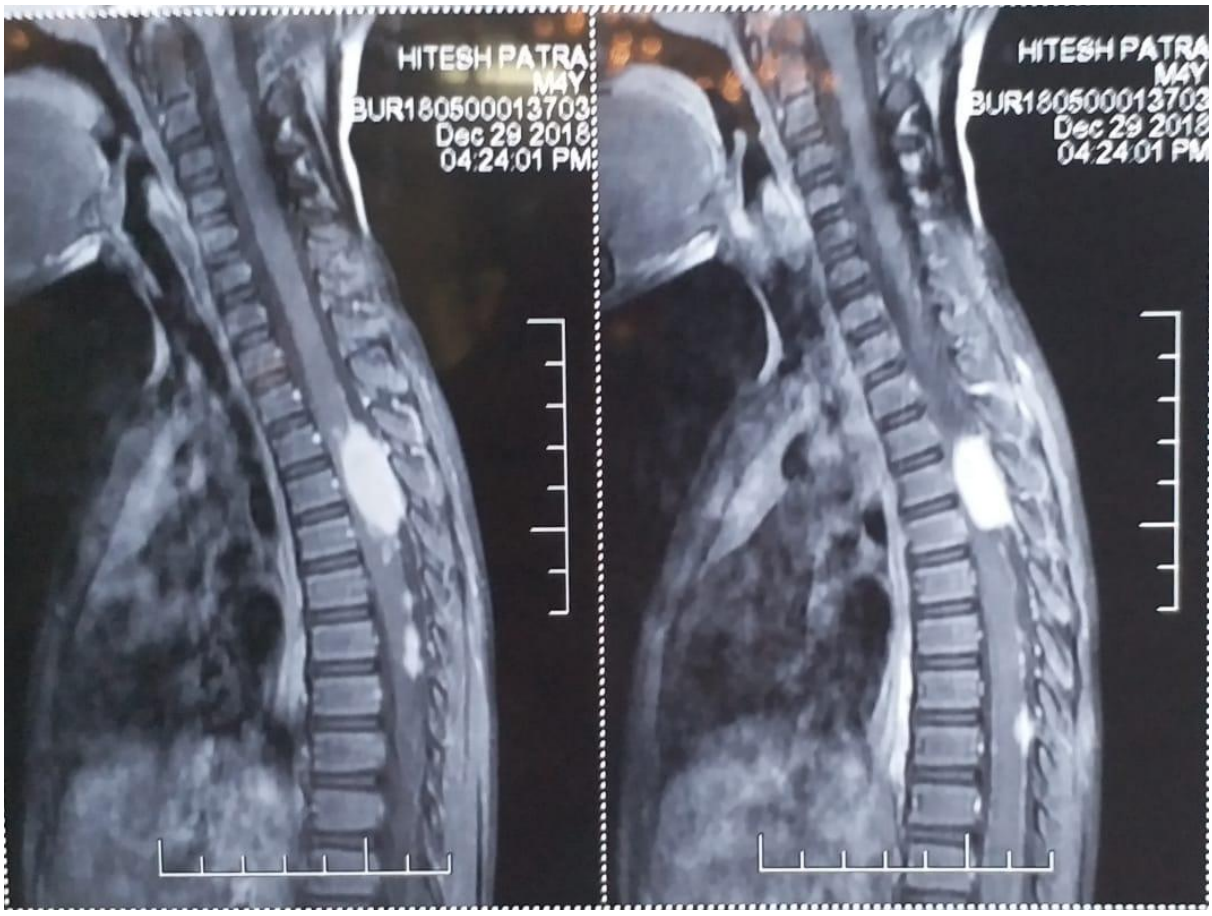
Risk stratification is based on the patient's age, metastasis, the extent of resected tissue and anaplasia in histology [9]. Patients over 3 years with localized lesions, no anaplasia, no metastases, and residual tumor tissue below

1.5 cm have a good predicted outcome and are classified as the standard-risk group. The high-risk group with poor outcome prognosis consists of very young children with dispersed metastases and unresectable tumor or residual tumor after surgery over 1.5 cm [9]. Currently, the treatment is risk-adapted. Maximum surgical resection is recommended, always followed by chemotherapy and neuroaxis radiotherapy [9]. The treatment of MB causes serious consequences for the developing CNS. Thus, patient stratification should include progression risk and aggressiveness of the disease and, consequently, the type and intensity of therapy. This underlines the importance of molecular research which leads to MB subtype identification

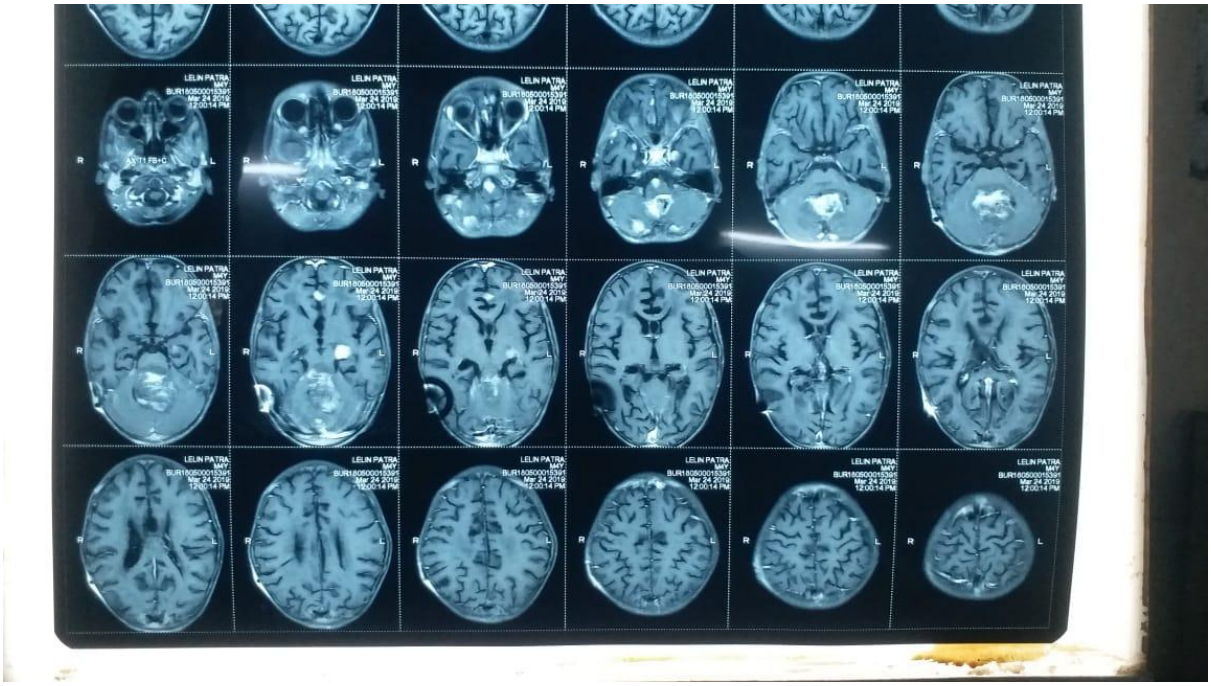
4. Conclusion

Brain tumors, after leukemia, are the major cause of death in pediatric oncology. The most common tumor, constituting 20% of all pediatric brain tumors, is MB, a small blue-cell malignant tumor that arises in the posterior cranial fossa and the cerebellum, mainly in the hemispheres and the vermis [10, 11, 12]. All variants of MB arise from primitive neuroectodermal cells. Patient symptoms result from alterations in the cerebellum (e.g. ataxia, hypotonia) and increased intracranial pressure (e.g. headache, vomiting withoutnausea, ocular palsies). Metastases spread via cerebrospinal fluid, rarely localizing outside the central nervous system. According to the new WHO classification (2007), medulloblastoma is divided into five groups: classic type (CMB), desmoplastic/nodular type (DN), medulloblastoma with extensive nodularity (MBEN), anaplastic type and large cell medulloblastoma (LC) [13]. As anaplastic medulloblastoma and large cell medulloblastoma often appear together, sometimes they are named large cell/anaplastic medulloblastoma (LC/A) [13]. Because of the similar histological pattern, DN and MBEN are classified as desmoplastic medulloblastoma (DMB)

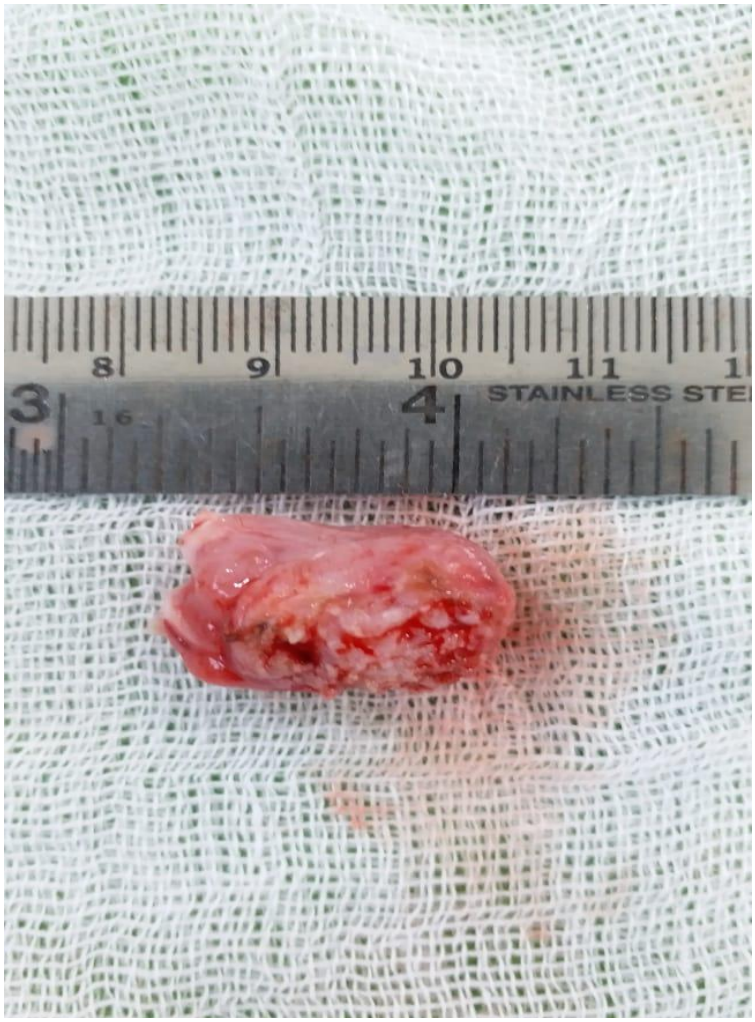
Photos



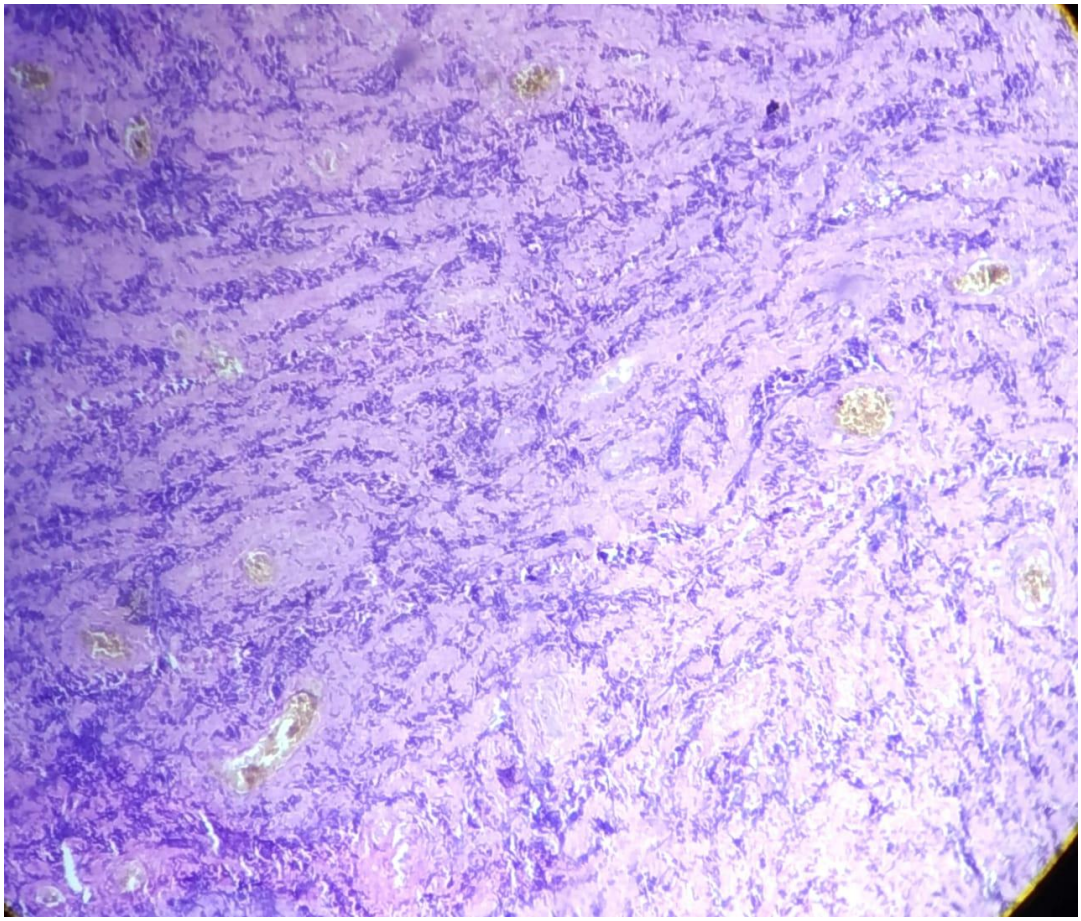
CT Scan Image of Thoracic and Lumbar Spine Area



Posterior fossa tumor in CT SCAN

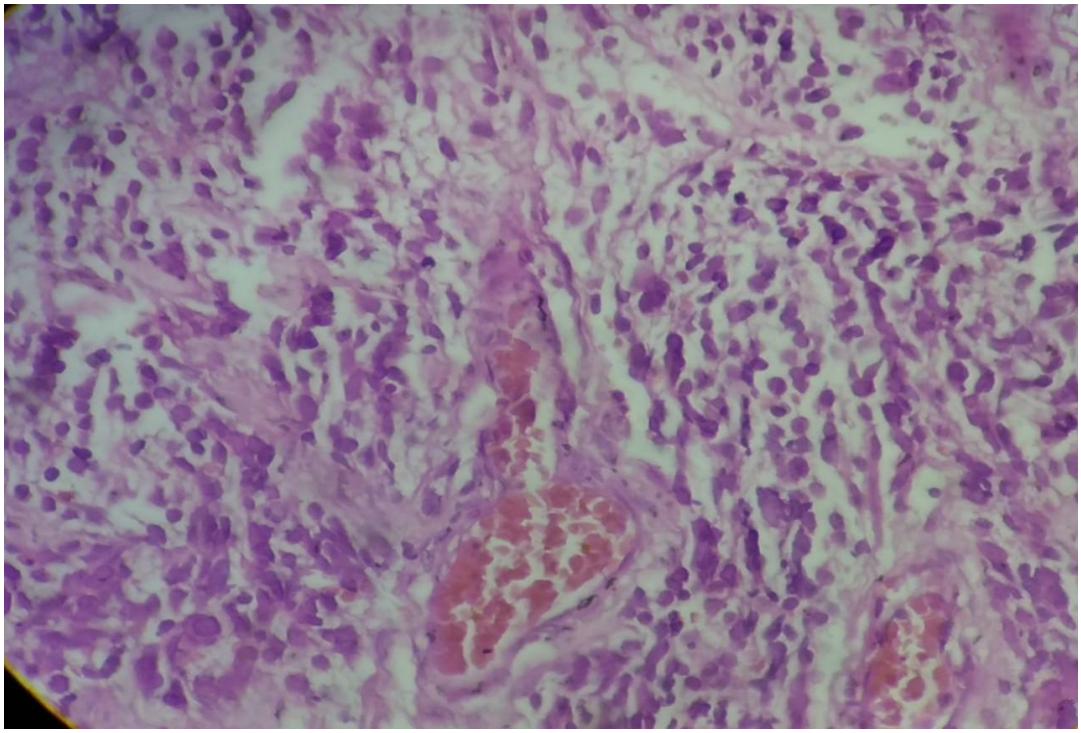


Gross -Red -White friable tissue mass measuring 2x1x1 cm



H&E X100

Small undifferentiated cells arranged in swirling and fascicular architecture. Nuclei are densely hyperchromatic, angulated with scanty cytoplasm.



H&E X400

Pale islands of cells with nuclear pleomorphism.

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