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NODULAR MEDULLOBLASTOMA IN A 23 YEARS OLD MALE PATIENT: A CASE REPORT

Pathology

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ABSTRACT

Nodular medulloblastoma is an embryonal neural tumour arising in the cerebellum and characterized by nodular, reticulin-free zones and intervening densely packed, poorly differentiated cells that produce an intercellular network of reticulin-positive collagen fibres. A 23 yrs. old male patient had a midline cerebellar SOL adjacent to the tentorium. The patient was operated and the lesion was excised. Histopathological examination showed multiple pale areas (nodules) having large cells with euchromatic nucleus in a fibrillary neuropil like background and the nodules were surrounded by densely packed densely packed, undifferentiated, highly proliferative cells with hyperchromatic and moderately pleomorphic nuclei. The nodules in desmoplastic/nodular medulloblastoma show variable expression of neuronal markers, including synaptophysin and NeuN. Desmoplastic/nodular medulloblastoma displays pathological activation of the SHH pathway, which is often caused by mutations in genes encoding members of the pathway, including PTCH1, SMO, and SUFU.

KEYWORDS

INTRODUCTION:

Medulloblastoma is the most common primitive neuroepithelial neoplasm arising in CNS. It is a WHO Grade IV1 tumor. Peak incidence is 3-7yrs1. M/F-1.7:11 It is located mainly in the cerebellum1. On MRI, it is hypointense on T1, iso to hyperintense on T2, also heterogenous due to calcification,necrosis,cyst formation. Nodular medulloblastoma is an embryonal neural tumour arising in the cerebellum and characterized by nodular, reticulin-free zones and intervening densely packed, poorly differentiated cells that produce an intercellular network of reticulin-positive collagen fibres1. In early childhood, it may be associated with Gorlin syndrome1. Nodular medulloblastoma corresponds histologically to WHOgrade IV1. It is also known as desmoplastic medulloblastoma.

CASE REPORT:

A 23 yrs. old male patient presented with complaints of headache, nausea and vomiting, ataxia and blurry vision. On MRI, there was a midline cerebellar SOL adjacent to the tentorium. The patient was operated and the excised tissue was sent for histopathological examination to the Dept. of Pathology, RIMS, Ranchi.

We received soft, friable tissue in multiple bits which were pearly white in colour. The measurement of the bits as a whole was 7cmx4cmx2cm. Histopathological examination showed multiple pale areas (nodules) having large cells with euchromatic nucleus in a fibrillary neuropil like background and the nodules were surrounded by densely packed, undifferentiated, highly proliferative smaller cells with hyperchromatic and moderately pleomorphic nuclei.

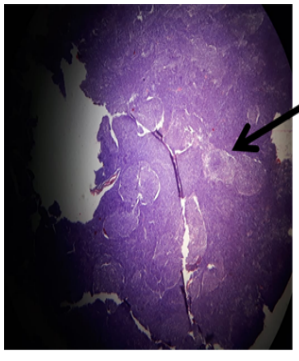

FIGURE 1: H & E scanner view (5x)

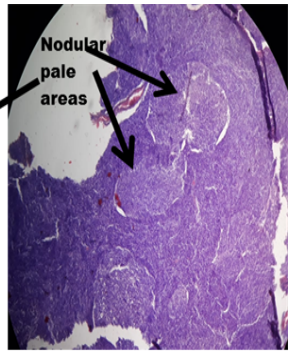

FIGURE 2: H & E low power view (10x)

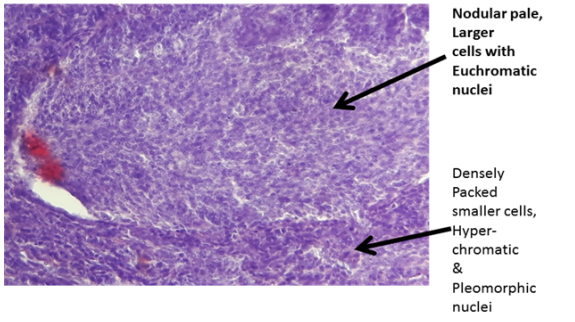

FIGURE 3- H & E high power view (40x)

DISCUSSION:

Desmoplastic/nodular medulloblastomas are estimated to account for 20% of all medulloblastomas[2]. In children aged < 3 years, desmoplastic/nodular medulloblastoma accounts for 47-57% of all cases[3]. Unlike most classic medulloblastomas, which are restricted to the midline, desmoplastic/nodular medulloblastoma may arise in the cerebellar hemispheres and in the vermis. Most medulloblastomas occurring in the cerebellar hemispheres are of the desmoplastic/nodular type[4]. On MRI, desmoplastic/nodular medulloblastomas present as solid, frequently contrast-enhancing masses[1]. Nodular Medulloblastoma can relapse locally, metastasize via cerebrospinal fluid pathways, and in rare cases spread to extra-CNS sites such as the skeletal system. At diagnosis, metastatic disease is found less frequently with desmoplastic/nodular medulloblastomas than with other variants of medulloblastoma[1]. Desmoplastic/nodular medulloblastoma is characterized by nodular, reticulinfree zones (so-called pale islands) surrounded by densely packed, undifferentiated, highly proliferative cells with hyperchromatic and moderately pleomorphic nuclei, which produce a dense intercellular reticulin fibre network[3]. The level of mitotic activity in the nodules is lower than in the internodular areas[1]. Unlike classic medulloblastoma, neuroblastic rosettes are not found in desmoplastic/nodular medulloblastoma[1]. Medulloblastomas that show only an increased amount of reticulin fibres (without a nodular pattern) or that show a focal nodular pattern without desmoplasia are not classified as desmoplastic/nodular medulloblastoma the two characteristic features must occur together for a diagnosis of desmoplastic/nodular medulloblastoma[6]. The nodules in desmoplastic/nodular medulloblastoma show variable expression of neuronal markers, including synaptophysin and NeuN[1]. The Ki-67 proliferation index is much higher in internodular areas than in nodules[1]. Activation of the SHH pathway can be inferred by

immunohistochemistry for specific targets, such as GAB1 and TNFRSF16. These markers are expressed predominantly in internodular areas[7]. Desmoplastic/nodular medulloblastoma displays pathological activation of the SHH pathway, which is often caused by mutations in genes encoding members of the pathway, including PTCH1, SMO, and SUFU[8]. In most cases, desmoplastic/nodular medulloblastoma in early childhood has an excellent outcome with surgery and chemotherapy alone[3].

CONCLUSION:

Nodular medulloblastoma is a histologically defined type of medulloblastoma and its clinical features, histopathological findings and immunohistochemical features must be known to a pathologist.

REFERENCES

1. David N. Louis, Hiroko Ohgaki, Otmar D. Wiestler,Webster K. Cavenee (Eds): WHO Classification of Tumours of the Central Nervous System (Revised 4th edition). IARC; Lyon 2016.
2. Pietsch T, Schmidt R, Remke M, Korshunov A, Hovestadt V, Jones DT, et al. (2014). Prognostic significance of clinical, histopathological, and molecular characteristics of medulloblastomas in the prospective HIT2000 multicenter clinical trial cohort. Acta Neuropathol. 128(1):137-49.PMID:24791927.
3. Rutkowski S, Bode U, Deinlein F, Ottensmeier H, Warmuth-Metz M, Soerensen N, et al. (2005). Treatment of early childhood medulloblastoma by postoperative chemotherapy alone. N Engl J Med. 352(10):978-86. PMID45758008.
4. Buhren J, Christoph AH, Buslei R, Albrecht S, Wiestler OD, Pietsch T (2000). Expression of the neurotrophin receptor p75NTR in medulloblastomas is correlated with distinct histological and clinical features: evidence for a medulloblastoma subtype derived from the external granule cell layer, J NeuropatholExp Neurol. 59(3):229-40.PMID:10744061
5. Giangaspero F, Chieco P, Ceccarelli C, Lisignoli G, Pozzuoli R, Gambacorta M, et al. (1991). "Desmoplastic" versus "classic" medulloblastoma: comparison of DNA content, histopathology and differentiation. Virchows Arch A Pathol Anat Histopathol. 418(3)207-14. PMID:1900966
6. McManamy CS, Pears J, Weston CL, Hanzely Z, Ironside JW, Taylor RE, et al.; Clinical Brain Tumour Group (2007). Nodule formation and desmoplasia in medulloblastomas-defining the nodular/desmoplastic variant and its biological behavior. Brain Pathol. 17(2):151-64. PMID:17388946.
7. Ellison DW, Dalton J, Kocak M, Nicholson SL, Fraga C, Neale G, et al. (2011). Medulloblastoma: clinicopathological correlates of SHH, WNT, and non-SHH/WNT molecular subgroups. Acta Neuropathol. 121(3):381-96. PMID:21267586.
8. Lam CW, Xie J, To KF, Ng HK, Lee KC, Yuen NW, et al. (1999). A frequent activated smoothened mutation in sporadic basal cell carcinomas. Oncogene. 18(3):833-6. PMID:9989836.
9. Giangaspero F, Perilongo G, Fondelli MP, Brisigotti M, Carollo C, Burnelli R, et al. (1999). Medulloblastoma with extensive nodularity: a variant with favorable prognosis. J Neurosurg. 91(6)971-7.PMID:10584843.
10. Helton KJ, Fouladi M, Boop FA, Perry A, Dalton J, Kun L, Fuller C. Medullomyoblastoma: a radiographic and clinicopathologic analysis of six cases and review of the literature. Cancer 2004, 101:1445–1454.