Medulloblastoma, WNT-activated

Typical in older childhood (hardly ever in infancy). Account for ≈ 10% of all MDBs and ≈ 15% of adult MDBs.

Male: female = 1:2. Predominant histologic variant: classic (almost all). May favor the midline of the cerebellum. The putative cell of origin: lower rhombic lip progenitor cells. Frequent genetic mutations: CTNNB1, DDX3X, TP53.

▶ Classic morphology. Prognosis: low-risk tumor. Almost all WNT-activated MDBs have classic morphology.

▶ Large cell/anaplastic morphology. Very rare. Uncertain clinical significance.

Epidemiology

Medulloblastoma - wingless (WNT) subgroup tumors are the least common medulloblastoma subgroup

They are usually seen in children and, to a lesser degree, adults. WNT tumors account for only 11% of all medulloblastomas, and have, unlike group 3 and 4 tumors, no particular predilection for males, with ~1:1 male to female ratio 1,2.

They are most frequently encountered in children (peak incidence 10-12 years of age), and are also encountered in adults, but almost never in infants 1.

Pathology

WNT tumors arise from progenitor cells of the lower rhombic lip that migrate laterally along the anterior extramural migratory streams (AES) 15. This accounts for the location of these tumors 15:

foramen of Luschka: 75% fourth ventricle: 70% cisterna magna: 30% cerebellopontine angle cistern: 20% WNT tumors are almost invariably (97%) of classic histology, with the remainder being of large cell/anaplastic morphology 1,2. Interestingly even those with large cell/anaplastic histology have a good prognosis, which is not the case with other subgroups 2.

Most WNT medulloblastomas are sporadic, most frequently the result of mutations of CTNNB1 which encodes for beta-catenin, although numerous other mutations have been described 2. In patients with Turcot syndrome the mutation is of the APC gene 2.

Radiographic features

They typically arise from the region surrounding the foramen of Luschka and middle cerebellar peduncle.

Interestingly, and helpful in preoperative assessment, the WNT subgroup has a distinct predilection

for the cerebellar peduncle, bulging into the cerebellopontine angle, which is not usually encountered in other molecular subgroups. They tend to have prominent contrast enhancement 3.

For more details on radiographic features please refer to the general article on medulloblastomas.

Treatment and prognosis Surgery is the first line of therapy (as is the case in all subgroups) with the aim being histological proof, molecular subtyping and maximal tumor resection, with adjuvant therapy depending on an overall risk profile (see general article on medulloblastoma) 2.

The incidence of CNS metastatic disease in the WNT subgroup at diagnosis is uncommon, found in only 10% of children, and not seen in adults 1.

WNT tumors have a much better prognosis than all other subtypes of medulloblastoma, in all age groups 1:

95% 10-year overall survival of in children 100% 5-year overall survival of in adults

Outcome

They have by far the best prognosis.

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