Pediatric Pathology & Laboratory Medicine

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Neuroblastoma is o en used as an omnibus term for all types of peripheral neuroblastic tumors including neuroblastoma, ganglioneuroblastoma, and ganglioneuroma. Tumors in this group are biologically diverse: Molecular/genomic properties of individual cases are closely related to their unique clinical behaviors. Biologically favorable tumors have a potential of spontaneous regression or tumor maturation and are o en associated with a hyperdiploid pattern (whole chromosomal gains without structural abnormalities). Biologically favorable tumors have a potential of spontaneous regression or tumor maturation and are o en associated with a hyperdiploid pattern (whole chromosomal gains without structural abnormalities). For neuroblastoma clinical trials, the children's oncology group utilizes their risk-grouping system for patient strati cation and protocol assignment based on the combination of clinical stage, age at diagnosis, International Neuroblastoma Patholog Classi cation,MYCN status, DNA index, and segmental chromosomal aberrations. Estimated survival rate for the non-high-risk patients is ~90% with surgery alone (low risk) or with biopsy/surgery and moderate chemotherapy (intermediate risk). In contrast, estimated survival rate for the high-risk patients remains as low as 45~50% even a er intensive treatment followed by stem-cell transplantation. Continuous e orts are being made for discovery of actionable/druggable targets in high-risk neuroblastomas. ose potential targets includeALK activating mutation/ampli cation (dysregulating cell signaling and leading to uncontrolled proliferation of neuroblasts),TERTrearrangement andATRX/DAXXmutation (preventing neuroblasts from telomere-mediated senescence); and MYC family protein overexpression- a new concept of highly aggressive "MYC family-driven neuroblastomas" with augmented expression of MYCN or MYC protein, also morphologically characterized by nucleolar hypertrophy (promoting MYC/MAX heterodimer formation for activating down-stream gene targets).

Hiroyuki Shimada has completed his MD and PhD from the Yokohama City University, School of Medicine and Ohio State University College of Medicine, respectively. He LV D 3URIHVVRU RI 3DWKRORJ\ DW WKH 8QLYHUVLW\ RI 6RXWKHUQ &DOLIRUQLD .HFN 6FKRRO RI 0HGLFLQH) of COG Neuroblastoma Pathology Reference Laboratory. He has been reviewing ~700 neuroblastoma cases per year from US, Canada, Australia and New Zealand and SDUWLFLSDWLQJ LQ YDULRXV FOLQLFDO DQG WUDQVODWLRQDO UHVHDUFK DFWLYLWLHV LQ WKH ¿HOG RI 3H

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