Paraneoplastic neurological syndromes in childhood: a single institution 8-year case series

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Objectives

Paraneoplastic neurological syndromes (PNSs) are remote neurological immune-related effects of tumors. The diagnostic criteria for PNSs were updated in 2021. However, the clinical characteristics of pediatric PNSs remain unclear. We retrospectively examined the clinical characteristics of pediatric cases of PNSs and assessed the performance of the 2021 diagnostic criteria in children.

Methods

Patients who were hospitalized in the Department of Neurology or Medical Oncology at Beijing Children's Hospital between June 2015 and June 2023 and met the following criteria were retrieved from the Medical Records System: (1) age of onset < 18 years; (2) suspected immune-mediated neurological syndrome; (3) positive PNSs antibodies or a clear tumor diagnosis during hospitalization or follow-up. Patients who met the above criteria were evaluated using the 2004 diagnostic criteria for PNSs, and those who fulfilled the description of "definite" were finally included in the study. The clinical characteristics of included patients were retrospectively analyzed and the 2021 diagnostic criteria were applied to re-diagnostic stratification of these patients.

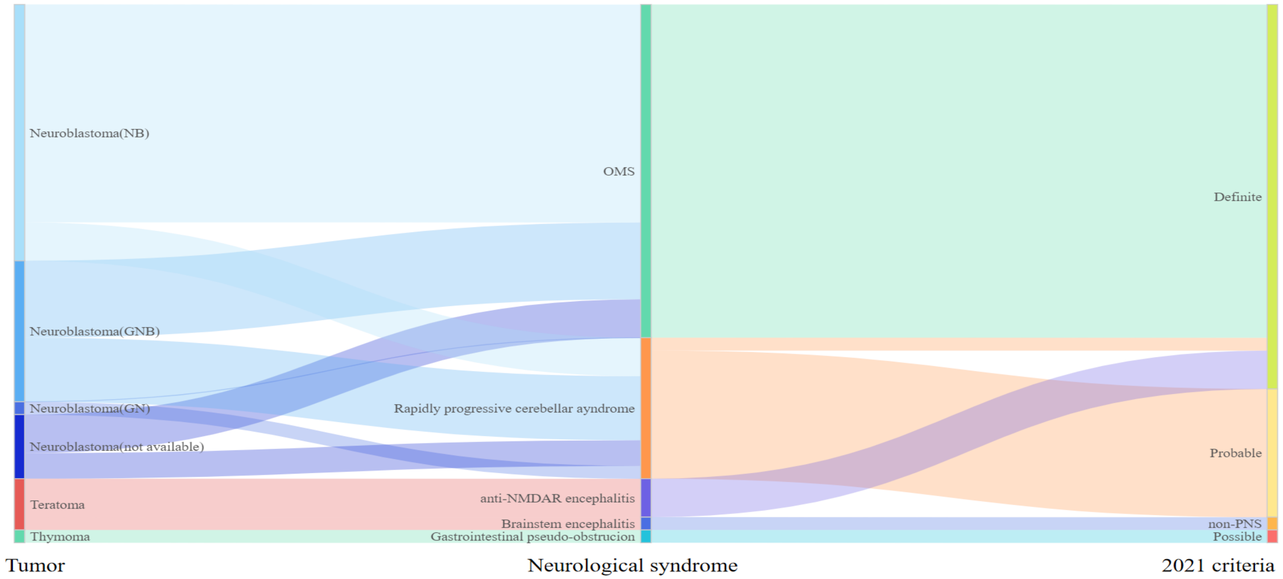

Figure 1: Diagnostic performance of 2021 criteria for paraneoplastic neurological syndromes.

Results

(1) General information

42 patients (20 male) with a median onset age of 1.75 (1.00–15.75) years were included. The most common neurological syndrome was opsoclonus-myoclonus syndrome (OMS) (62%), followed by rapidly progressive cerebellar syndrome (26%). A majority of tumors were neuroblastomas (88%), with few being ovarian teratomas (10%). 71% (30/42) of patients were classified as "definite" and 24% (10/42) as "probable" according to the 2021 criteria.

(2) Performance of the 2021 diagnostic criteria

71% (30/42) were classified as "definite," 24% (10/42) as "probable," 2% (1/42) as "possible" and 2% (1/42) as "non-PNS" using the 2021 criteria. All cases judged as "probable" were rapidly progressive cerebellar ataxia with neuroblastoma.

(3) Treatment of neuroblastomas associatied PNSs

For OMS, chemotherapy according to the risk stage of the tumor and regular infusion of IVIG and oral steroids were administered after tumor diagnosis. Twenty-one patients were follow-up for ≥6 months, with a median follow-up of 4.92 (0.58–7.58) years. The median score of 12 (7–14) on the Mitchell and Pike OMS rating scale at first hospitalization and 0 (0–5) at the final follow-up. For rapidly progressive cerebellar syndrome, therapy regimen was similar. Nine patients were regularly followed-up with 4.42 (1.17–7.50) years. The mean mRS score at first hospitalization was 4(3-4) and 1 (0–4) at the last follow-up. Poor response to this regimen were observed in only 17% (5/30) of the patients from the two groups. Of the five patients, four were in the low-risk group (without chemotherapy), of whom, three were subsequently treated with RTX with significant improvement of symptoms.

CONCLUSIONS

- ✓ OMS followed by rapidly progressive cerebellar ataxia are the most common form of PNSs in children and are associated with neuroblastoma.
- ✓ The use of multiple immunotherapies containing second-line drugs may improve prognosis of neuroblastoma-associated PNSs.
- ✓ The 2021 criteria perform well in pediatric PNSs. However, we suggest upgrade antibody-negative rapidly progressive cerebellar ataxia with neuroblastoma to "definite" diagnosis to further improve the diagnostic efficacy of this diagnostic criterion in childhood.