

Primary Extraosseous CNS Ewing Sarcoma in Children: A Rare Case Series from a Major Public Neuro-Oncology Center in Pakistan

Think of CNS Ewing sarcoma in aggressive pediatric CNS tumors

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1 Background & Objective

🦴 Rare: primary CNS-EES, rarer in children

🌐 Gap: pediatric CNS-EES rarely studied

Objective: document the center experience with pediatric CNS-EES in a resource-limited setting.

2 Methods

■ **Design:** ambidirectional case series, 2023–24

■ **Setting:** Children's Hospital Lahore, age <16 y

■ **Diagnosis:** round blue cells + NKX2.2 + MDT

■ **Outcome:** event-free survival (EFS), Wilson CI

3 Patient Profile

4/139

CNS tumors

2.9%

incidence

8.5 y

median age

1:1

male : female

5.75 wk

symptoms

75%

consanguinity

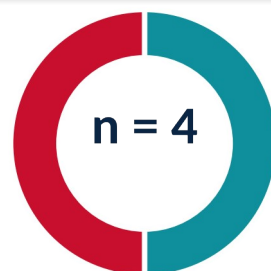
50%

café-au-lait

2/4

adjuvant Rx

4 Tumor Site



50%

Cerebral
parietal lobe (2)

50%

Spinal
L4–S2; cervical (2)

8 Discussion

■ **Parietal lobe** in both cerebral cases (2/2)

■ **Consanguinity** 75%; café-au-lait macules 50%

■ **Only 2/4** received adjuvant therapy; LMIC delays

■ **NKX2.2** is a fair substitute for translocation tests

5 Diagnostic Clues

NKX2.2 positive

4/4

Round blue cell morphology

4/4

CD99 positive

1/2 tested

Translocation testing

unavailable

Suspected syndrome

2/4

KEY RESULTS

4 of 139 pediatric CNS tumors were primary CNS Ewing sarcoma

2.9%

incidence (95% CI 1.1–7.2)

4.5 wk

median EFS (range 4–40 weeks)

25%

survival: 1 of 4 in remission

4/4

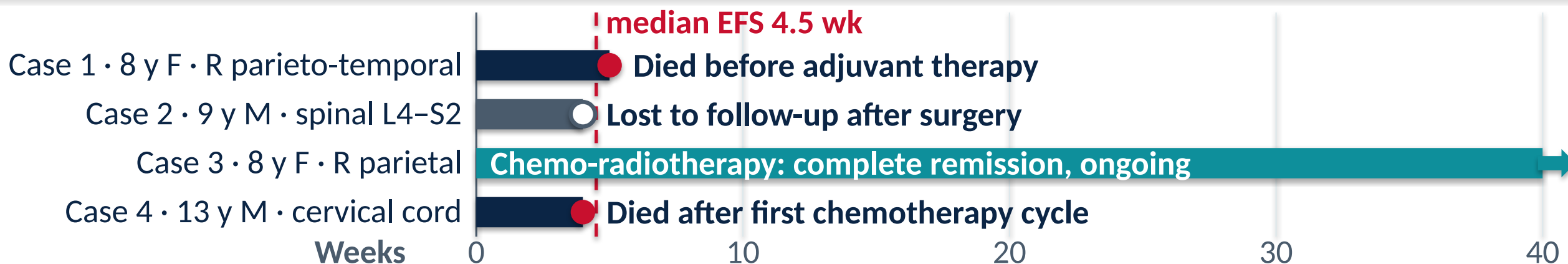
NKX2.2 positive (IHC)

7 Event-Free Survival by Case

● Died

○ Lost to follow-up

➡ Ongoing remission



Redrawn from Table I. EFS = diagnosis to death, treatment abandonment or data cut-off (Feb 2025).

Limitations: n = 4 • 25% lost to follow-up • 50% died on treatment • no translocation testing

10 Conclusion

CNS-EES is rare but not infrequent here. Round blue cells with NKX2.2 are a fair diagnostic substitute without translocation testing; consider it before surgery in aggressive pediatric CNS tumors.

11 References

1. Haveman 2020, Cancers
2. Jiang 2020, Front Oncol
3. Pasricha 2023, Indian J Pathol

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