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abstract

Clinical Features and Treatment Outcomes in Children with Ewing Sarcoma

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Introduction: Five-year survival rates in children with Ewing Sarcoma (ES) have increased to 70-80%. We aimed to investigate the clinical features and treatment outcomes in children with Ewing sarcoma.

Methodology: 175 patients with ES diagnosed between 2000 and 2022 were included. Clinical features and treatment outcomes were recorded retrospectively.

Results: Median age at diagnosis was 10.6 years (0.4-17.6), and the female/male ratio was 1.3. The most common tumor locations were the extremities, trunk, and pelvic region in 39.4%, 29.7%, and 22.3% of patients, respectively. Metastatic disease was present in 34.8% of children. Treatment regimens were MMR (cisplatin, cyclophosphamide, adriamycin, vincristine) in 26, PIAV (cisplatin, ifosfamide, adriamycin, vincristine) in 63, and Euro-Ewing 99 in 86 children. Radiotherapy was administered to 102 and major resection was performed in 118 patients. Five-year overall survival (OS) and event-free survival (EFS) rates for the entire group were 57.4%

and 45.8%The five-year OS and EFS rates in children who received MMR, PIAV, and Euro-Ewing 99 were 57.7% & 46.2%; 49.2% & 38.1%; 63.4% & 52.3% (p=0.1).

The five-year OS and EFS rates for metastatic versus localized disease were 36.1% & 25%, compared to 69.6% & 57.5% (p<0.05). Children with tumor volumes ≤8 cm (n=95) had five-year OS and EFS rates of 66.8% & 55%, while those with tumors >8 cm (n=76) had 46.6% & 34.1% (p<0.05). The five-year OS and EFS rates for the upper extremity, lower extremity, trunk, pelvic, and cranial locations were 71.9% & 56.1%; 59.5% & 48.3%; 53.8% & 51.5%; 41.7% & 24.4%; 85.7% & 75% (p=0.04). The five-year OS for children with resistant and relapsed disease was 35.2% and 24.5%.

Conclusion: The treatment outcomes demonstrate progress in achieving better survival rates. The tumor's location, volume, and extent of disease have prognostic significance for survival rates. New and more aggressive treatments are needed in risk groups with trunk-pelvic location, big tumors, metastatic, refractory/relapsed disease.