Renal Ewing's Sarcoma/Primitive Neuroectodermal Tumor: Diagnostic and Therapeutic Approaches

Introduction and Objective: To assess the different diagnostic modalities, efficacy of adjuvant therapy and oncological outcome of patients with Ewing's sarcoma/ Primitive neuroectodermal tumor of the kidney (ES/PNET).
Materials and Methods: Data files of seven patients with a diagnosis of renal ES/PNET have been reviewed. Immunohistochemical staining included CD 99, Cytokeratin, FLI-1 and WT-1 gene were utilized to confirm diagnosis. Adjuvant chemotherapy in the form of Cisplatinum and Etoposide was given. Follow up and oncological outcome was recorded.
Results: Loin pain was the main presenting symptom. At time of diagnosis, five patients had locally advanced and/or distant metastasis. Six patients underwent radical nephrectomy; two of them received adjuvant chemotherapy. In one patient, chemotherapy was given without surgery. Mean disease free survival was 4.8 months in 6 patients while one patient still lives free after 24 months. Immuno-histochemical studies showed strong and diffusely positive membranous pattern for CD99 in all patients. The positivity for FLI-1 was detected in four out of five cases (80%), the staining location is nuclear. No tumors expressed pancytokeratin or WT-1.
Conclusions: Combination of CD 99 as a positive marker and WT-1 gene as a negative marker is sufficient immunohistochemical techniques to diagnose ES/PNET. More research is needed to find more effective adjuvant protocols for this aggressive tumor.