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LONG PROGRESSION FREE SURVIVAL WITH PAZOPANIB IN EWING SARCOMA OF PELVIS

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Introduction - Purpose : Ewing Sarcoma (ES), Peripheral Neuroectodermal Tumor (PNET), Malignant Small Cell Tumor of Torakopulmonary Region (Askin Tumor) and atypic ES are all classified as ES family tumors. Standard treatment modality of ES is vincristine, doxorubicine,cyclophosphamide (VAC) and IE. VAC/IE 4-6 cycle preoperatively or before local treatment then VAC/IE continued until totally 14-17 cycles in locally presented patients.

Findings : In 2014 , a 23 years old woman admitted to our clinic with complaints of leg pain, abdominal pain and an inguinal mass. Andominal ultrasonography imaging revelaed a mass in the pelvis. 18 FDG-PET/CT imaging revelaed a mass oin the pelvis of 16x13x17,5 cm size and destructing right iliac bone, sacrum and descending to right acetabulum. Pathology of tructbiopsy of the mass revelaed small round cells with scant cytoplasm, immunohistochemical staining showed CD 99 positivity and negative for Pan CK, EMA, Vimentin,Chromogranin, Synaptophysin, CD 56, and LCA. Genetic analysis by FISH showed t (11,22) (q24,p12) which forms a fusion transcriptor gene, EWSR 1- FLI1. VAC/IE chemoterapy regimen was planned preoperatively for 4 cycles. PET/CT imaging revealed stable disease pattern after 4 cycles of chemotherapy. Multidisciplinary tumor board decided to operate patient. But the patient refused operation. Definitive radiotherapy had given to pelvis.After definitive radiotherapy pazopanib 800 mg/day started to patient since she refused further chemotherapy. After three months PET/CT imaging revealed complete metabolic response of mass and no new lesions. Figure 1. She is using pazopanib 800 mg/day for 35 months and last PET/CT imaging revealed stable disease, complete metabolic response of mass. Figure 2.

Discussion : The specific cell of origin in ES is believed to be neuroectodermal origin. Nearly all cases demonstrate reciprocal translocation including EWSR 1 gene on chromosome 22. ES is accepted as systemic dişsease ewen if in localized disease pattern since majority of patients who dont take systemic intensive chemotherapy will develope metastases in one year. Our patient refuses systemic chemotherapy after localized treatment of radiotherapy and used pazopanib 800 mg/day . Pazopanib an oral multitargeted tyrosine kinase inhibitör approved fort he treatment of soft tissue sarcoma other then gastrointestinal stromal sarcoma and liposarcoma. The exact mechanism of pazopanib in soft tissue sarcoma is unclear. There are just few case reports about the effect of pazopanib in ES patients in literature and the progression free survivals reported in that studies are limited to justa a few months. In our case the patient is responsive to pazopanib for nearly 3 years and the affect is ongoing. We dont know in which patients pazopanib works better, we need a marker or clue for the patient selection. We need controlled randomized clinical trials with pazopanib for further proof of clinical benefit in soft tissue sarcoma patients.

Keywords: ewing sarcoma, pazopanib, pelvis