

KLIPPEL-TRENAUNAY SYNDROME SYMPTOMS IMPROVE ON ANTI-ANGIOGENIC THERAPY- A CASE REPORT

Sheila Nguyen, and Arkadiusz Dudek, MD, PhD

Klippel-Trenaunay Syndrome (KTS) is a rare congenital vascular disease characterized by malformation of capillary, venous, and lymphatic vessels and bony and soft tissue hypertrophy of involved an extremity. The exact cause of KTS is unknown and traditional treatment has been largely conservative and symptomatic. Recently a novel angiogenic factor VG5Q has been identified as a susceptibility gene for KTS, and increased angiogenesis may be pathogenic mechanism of KTS.

We present a case of a 37 year old female patient with multiple lymphangiomas, keratoangiomas and hemangiomas forming, especially in the right lower extremity due to KTS. She was unsuccessfully treated with multiple surgeries and vincristine chemotherapy, left with non-healing open wounds. Antiangiogenic therapy with sunitinib, a novel oral vascular endothelial growth factor receptor (VEGFR), platelet-derived growth factor receptor (PDGFR), and fibroblast growth factor receptor (FGFR) tyrosine kinase inhibitor was initiated. During the course of treatment, patient experienced notable improvement in the affected area, with decrease in size of angiomas and healing of open wounds.

We propose that antiangiogenic therapy maybe an effective strategy for KTS management due to the fact that vascular malformation plays a major role in the pathology of KTS.