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Clinical Presentation and Management of Klippel Trenaunay Syndrome: A Review

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Klippel Trenaunay Syndrome (KTS) is a rare congenital, developmental, multisystemic syndrome primarily affecting the extremities. Neonatal and early childhood are the usual periods for clinical presentation, which is characterized by a triad of nevus flammeus, venous varicosities, and osseous or soft-tissue hypertrophy. There is an equal occurrence among males and females, with a low incidence rate, and no relation to a specific race or ethnic group. In addition to the clinical triad the syndrome can be limited to the skin or it can extend deeply involving other systems including GIT, GU and hematologic. Color Doppler sonography nowadays is considered as the most accurate diagnosing method. A multidisciplinary approach to manage KTS is warranted. Depending on the presentation it can be managed either conservatively or surgically. Compression is the hallmark of conservative management; laser can be used to treat port wine stains. Techniques for ablation of superficial veins and malformations are individualized and may include sclerotherapy. Surgical intervention may be minimal invasive as epiphysiodesis or more aggressive surgery like amputation. Furthermore, treating clinical manifestations of KTS is crucial since it has a negative impact on the patients' quality of life (QOL), from pain, functional limitations, and cosmetic effects requiring a psychological support for the patients and their families. Our aim in this review is to highlight the important points about KTS syndrome in order to raise the level of knowledge and enable differentiation from other vascular anomalies. Another aim is to discuss the diverse management plans based on clinical presentation and the patient's main concerns.

Biography

Alqahtani NN is currently working as a Medical Intern at Princess Nourah Bint Abdulrahman University Riyadh, Saudi Arabia

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