Photoclinic: Klippel-Trenaunay-Weber Syndrome

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The parents of this 2-year-old boy brought their son for evaluation of swelling of the right leg and excoriation and serosanguineous discharge from the ankle region of 3 days' duration. The child had had a hemangioma of the right ankle since birth. Subsequently, there was gradual spread of the lesion along the leg to the buttocks.

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Sairah Chachad, MD, of Morgantown, WVa, and Atiya Khan, MD, of Fort Wayne, Ind, write that this patient had Klippel-Trenaunay-Weber syndrome with secondary cellulitis of the leg. Hemangiomas are common in infancy and are often benign. Klippel-Trenaunay-Weber syndrome is characterized by the triad of capillary and venous malformations, venous varicosity, and hyperplasia of soft tissue in the affected area. Parkes Weber syndrome is diagnosed when this triad is accompanied by an arteriovenous fistula. The vascular anomaly of Klippel-Trenaunay-Weber syndrome is apparent at birth and usually involves a lower limb; however, more than one extremity, as well as the trunk or face, may be affected. Most cases are unilateral. Enlargement of the soft tissues may be gradual and may involve the entire extremity. Edema of the involved leg can occur and may lead to lymphatic abnormalities. Complications may include bleeding, secondary infection (as in this patient), thrombophlebitis, gangrene of the affected extremity, and congestive heart failure. Hematuria secondary to angiomatous involvement of the urinary tract is an infrequent complication. Management is generally supportive, although surgical intervention may be an option for some patients. Because discrepancies in leg length are common in Klippel disorders, close orthopedic follow-up is required.

This patient was given a 10-day course of oral cephalexin for the cellulitis.

References: REFERENCES:

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