Hematuria, rectal bleeding and pelvic phleboliths in children with the Klippel-Trenaunay syndrome

E. M. Azouz
Department of Radiology, The Montreal Children's Hospital and McGill University, Montreal, Quebec, Canada

Abstract. Three young patients with Klippel-Trenaunay Syndrome (KTS), pelvic hemangiomatosis and bleeding are presented. Two children had rectal bleeding and the third presented with hematuria. Radiologic investigation of these patients with complicated angiodysplasia is reviewed. In infants and children calcified phleboliths seen on plain radiographs point toward the correct diagnosis. Angiography shows the vascular malformation which is usually on the venous side. Computed tomography delineates the angiomatous masses and identifies upward extension in the pelvis. Urography or renal ultrasound may show an associated kidney anomaly or tumor.

Key words: Klippel-Trenaunay syndrome – Pelvic phleboliths, children – Pelvic angiomatosis – Congenital angiodysplasia

The triad of cutaneous hemangiomas, bone and soft tissue hypertrophy of a lower extremity and congenital superficial venous varicosities is termed Klippel-Trenaunay syndrome (KTS) [8, 15, 20, 32] or congenital angiectatic hypertrophy [22]. Out of a population of 20 children with this rare disease complex, seen at the Asymmetry Clinic of the Montreal Children's Hospital, three had associated pelvic angiomatosis with intermittent bleeding.

Case reports

1. KTS with recurrent, sometimes profuse rectal bleeding

A three-year-old-boy had cutaneous hemangiomas, angiokeratomas and superficial varicosities of the left scrotum, thigh, knee and toes (Fig.1a). He had several episodes of bleeding from an angiokeratotic area on the skin of the lower thigh. He had no significant bone or soft tissue hypertrophy. The mother noticed occasional blood streaking of his stools since the age of one year. This was sometimes associated with the prolapse of an easily reducible purple fleshy mass per rectum. Clinical diagnosis was bleeding hemorrhoids, rectal prolapse and iron deficiency anemia. He had two episodes of profuse rectal bleeding requiring hospitalization. A barium enema was requested to rule out polyps. A preliminary radiography of the abdomen showed seven round small calcific densities in the left side of the true pelvis. The barium enema was normal except for mild mucosal irregularity of the rectum (Fig.1b). An excretory urogram showed normal kidneys but the urinary bladder was somewhat displaced to the right. A left lower extremity venogram showed generalized phlebectasia and gross angiomatosis with a very large vein in the lateral subcutaneous tissues of the thigh (Fig.1c). In the pelvis, this abnormal vein was of a relatively smaller caliber, the change of diameter occurring at the inguinal ligament.

Computed tomography (CT) scanning of the thighs demonstrated the vascular masses (Fig.1d) and in the pelvis, the phleboliths were identified to be anterior: above and to the left of the urinary bladder (Fig.1e). Although no phlebolith was seen posteriorly, sigmoidoscopy revealed purplish bulging mucosa affecting all walls of rectum from anal verge up to ten cm. He also had evacuation of a 3 x 3 cm left scrotal hematoma. At the time of surgery, venous malformation of the gubernaculum was seen and biopsy showed very vascular fibrofatty tissue consistent with angiomatosis. After medical management of his anemia, the only surgical treatment of his rectal bleeding consisted of suturing and removing the larger areas of bulging abnormal rectal mucosa.

Comment. This case represents KTS with left lower extremity and pelvic involvement. Symptoms included episodes of rectal bleeding, most probably of venous origin. The diagnosis of pelvic angiomatosis was strongly suspected by the demonstration of phleboliths on plain radiography. Venography and CT scanning confirmed the diagnosis, the first by directly showing the venous abnormalities and the second by demonstrating the diffuse nature of the pelvic involvement.

2. KTS with one episode of mild rectal bleeding

An 11-year-old girl was followed at the Clinic for four years for a diffuse hemangioma of the right lower extremity involving the buttock, thigh, leg and foot (Fig.2a). She also had superficial varicosities and small angiokeratomas around the knee. Because of
Fig. 1a–e. V.L. Three-year-old boy with KTS and recurrent sometimes profuse rectal bleeding. a Cutaneous hemangioma and angio-keratoma of his left lower extremity, predominantly around the knee and distal thigh. b Barium enema: mucosal irregularity of the rectum (arrow) in the region of the pelvic phleboliths. c Left lower extremity venography: gross venous malformation and phlebectasia in the thigh. d CT distal thighs: cutaneous thickening laterally and posteriorly, large subcutaneous vascular masses posteromedially (V) and absent long saphenous vein on the left side. e CT lower pelvis: phlebolith and mass indents the left lateral wall of the opacified urinary bladder (b). Surprisingly, no phlebolith is seen posteriorly.