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PRIMARY CHYLOCOLPORRHEA SUCCESSFULLY MANAGED BY DIVISION AND LIGATION OF RETROPERITONEAL LYMPHATICS

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ABSTRACT

Chylocolporrhea or chylous vaginal discharge is a rare manifestation of the primary chylous reflux syndrome. We describe its occurrence in a young child successfully treated by ligation of retroperitoneal, groin, and vaginal megalymphatics.

Chylocolporrhea or chylous vaginal discharge is a rare manifestation of the primary chylous syndrome. We report a young girl with this phenomenon successfully managed by ligation of feeding retroperitoneal, groin, and vaginal lymphatics.

Case Report

A 6-year-old girl had milky vaginal discharge since the age of 3 years. Otherwise she was asymptomatic. Physical examination revealed an underdeveloped child with otherwise normal external genitalia. Of note, there was no peripheral lymphedema, no ascites, and no dermal lymphangiomata. Blood tests revealed mild hypoalbuminemia (3gm/dl), mild hypoimmunoglobulinemia (IgG 326mg/dl, IgA 67mg/dl, IgM 41mg/dl), and a normal red and white blood cell count. The milky vaginal discharge revealed a protein content of 800mg/dl, cholesterol 37mg/dl, triglycerides 707mg/dl, glucose 60mg/dl, and a small number of leukocytes. The quantity of the vaginal discharge was approximately 300 to 800ml/24h.

Abdominal ultrasonography was unremarkable. Cystourethrography revealed mild ureteral reflux. Intravenous pyelography was normal. Computerized tomography showed tortuous structures around the uterus and in the perirectal region extending into the paraaortic area, which were interpreted as either distended, tortuous veins, and/or megalolymphatics *(Fig. 1A)*. Peripheral phlebography was normal. Conventional lymphography revealed megalolymphatics around the vulva, vagina, uterus, and parametrium. The thoracic duct was patent and normal appearing. Based on these findings, we opted to interrupt if possible the aberrant functioning lymphatics responsible for the chylous vaginal discharge.

On the operating table after induction of general anesthesia, patent blue dye was injected between the toes of both feet. A sequential intermittent pneumatic device (Lympha-press sleeves) was placed on the patient's legs to facilitate milking and rapid transport of the blue dye into the retroperitoneum (1). At laparotomy, huge megalolymphatics were noted along the iliac vessels bilaterally extending around the uterus and the pararectal region *(Fig. 1B)*. A methodical division and ligation of these megalolymphatics was carried out. By the next day the vaginal discharge stopped completely and the patient was

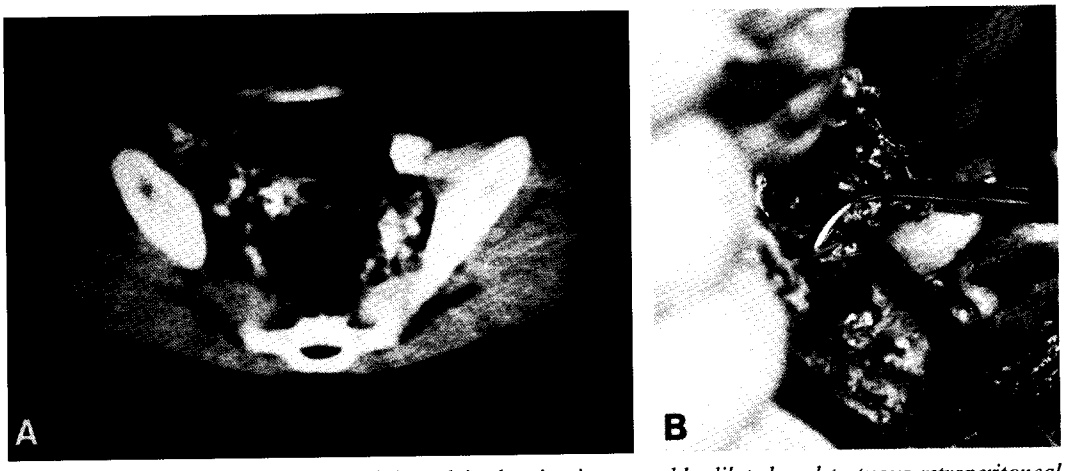

Fig. 1. (A) Computed tomography of the pelvis showing innumerable dilated and tortuous retroperitoneal lymphatics filled with oil contrast (direct lymphography). (B) Racemose megalolymphatics in the retroperitoneum which are undergoing ligation and division.

discharged seven days after operation. Three months later, however, the chylous vaginal discharge reappeared although in a lesser amount (100-200ml/24h). On the premise that chylous reflux was finding its way retrograde through groin lymphatics into the genitalia, the patient was operated on again and lymphatics in both groins draining toward the vagina were divided while maintaining their continuity to the retroperitoneum. After the second procedure, the vaginal discharge again stopped although sporadically small amounts (10-20ml) of lymphatic vaginal leakage reappeared. Four months later, however, chylous vaginal discharge recurred with leakage volumes rising to 500-800ml/h. Accordingly, the patient was operated on for the third time. Again Lymphapress sleeves were placed on the patient's legs and patent blue dye again injected into the web space between the toes. While the sequential pneumatic device was in operation, a careful vaginal exploration revealed four small openings through which lymphatic "blue" fluid were seen to escape. A submucosal oversewing of these vaginal "pores" was done and its "sealing" effectiveness tested for 30 minutes by the lack of continued vaginal staining as the Lymphapress continued to squeeze the legs and the position of the operating table was changed. After this third operation, chylous vaginal discharge stopped completely and one year later the patient is asymptomatic.

DISCUSSION

Lymphatic insufficiency (analogous to venous insufficiency) can arise primarily or secondarily to external trauma, irradiation, operations, or neoplasia. Because of progressive lymphatic valvular incompetence, lymph from the cisterna chyli begins to flow retrograde into distal lymphatics which in turn become unable to propel lymph antegrade. The increasing hydrostatic pressure causes these distal lymphatics to enlarge and become tortuous. Intestinal chyle unable to drain cephalad, "backs up" into distal tissues or may rupture into areas with little or no resistance such as the abdominal cavity (chylous ascites), urinary bladder (chyluria), the uterus (chylometrorrhea), or into the skin of the abdominal wall or thighs (chylous edema).

Primary chylous reflux draining through the vagina (chylocolporrhea) is extremely rare. In 1981, Adashi et al., reviewed the gynecological features of the primary chylous reflux syndrome (2), and described only 16 patients over the last 100 years of whom only 7 were operated upon. The only successful treatment has

been by division and ligation of megalolymphatics in the retroperitoneum in one or more stages. Indeed, a similar case history in a 9-year-old girl was reported by Jimenez-Cossio (3) where ligation and interruption of retroperitoneal lymphatics also was successful in arresting chylous vaginal discharge.

The technique of milking the patent blue dye instilled intraoperatively into the toes by the Lymphapress (sequential intermittent pneumatic device) helps to dilate the megalolymphatics in the retroperitoneum and facilitates their visualization thereby allowing rapid and efficient identification, division and ligation of the "feeding" lymphatics, and assures further, that lymphatic leakage after operative repair has ceased.

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