

CHYLOUS METRORRHEA

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ABSTRACT

A nine-year-old girl with primary (idiopathic) chylous metrorrhea is described. Extravasation from retroperitoneal lymphatics into the vaginal vault was seen on dorsal pedal lymphangiography. Bony abnormalities were also present in the lumbosacral vertebrae. Nineteen months after transperitoneal lymphangiectomy she is asymptomatic.

Disturbances of lymph flow and mesenteric lacteals are associated with a number of syndromes characterized by chylous reflux. Among these are chylous ascites, chylopericardium, chyloarthrosis and chyluria. Chylous metrorrhea or milky vaginal discharge is another rare variant. We recently observed a young girl with this abnormality that responded to excision of dilated lymphatics in the retroperitoneum.

CASE REPORT

A nine-year-old girl had an intermittent milk-like vaginal discharge since the age of four years which on occasion persisted for three to four months. Four months before hospital admission, fluid began to leak through a skin vesicle adjacent to the left groin with the gradual onset of swelling of the left labia majoris. She was afebrile, had no sign of infection but the milky vaginal discharge was aggravated while standing.

Overall, body development was normal and the family history unremarkable. Physical

examination disclosed a small vesicle covered by a thin crust on the upper part of the left groin. The right leg was longer than the left and plain x-rays of the pelvis and extremities confirmed a pelvic tilt with a 4 cm leg length discrepancy of which 3 cm extra length was attributable to the femur and 1 cm to the tibia. Other bony abnormalities in the lumbosacrum included lytic lesions (tomography), a rounding of the anterior sacral promontory, and an abnormal trapezoidal shape to the fifth lumbar vertebra with posterior arch diastasis.

Dorsal pedal lymphangiography revealed normal peripheral lymphatics. However, ilioaortic lymphatic trunks showed saccular dilations with delayed emptying of contrast medium (Fig. 1A). Abnormal lymph channels were also found emanating from femoroinguinal nodes, draining into the prepubic region and reaching the midline from where contrast extravasated into the vaginal wall (Fig. 1B). On the left, hypogastric lymphatics filled retrograde with reflux into lumbar channels. The thoracic duct was normal. Delayed films demonstrated irregular and partially opacified periaortic lymph nodes. At laparotomy two weeks later, a large number of dilated lymphatic saccules at the pelvic brim were resected and obliterated with hemo-clips. Intraoperative visualization of these lymphatics was facilitated by administration of whipped butter colored with Sudan III a few hours before operation. There were no post-operative complications. More than two

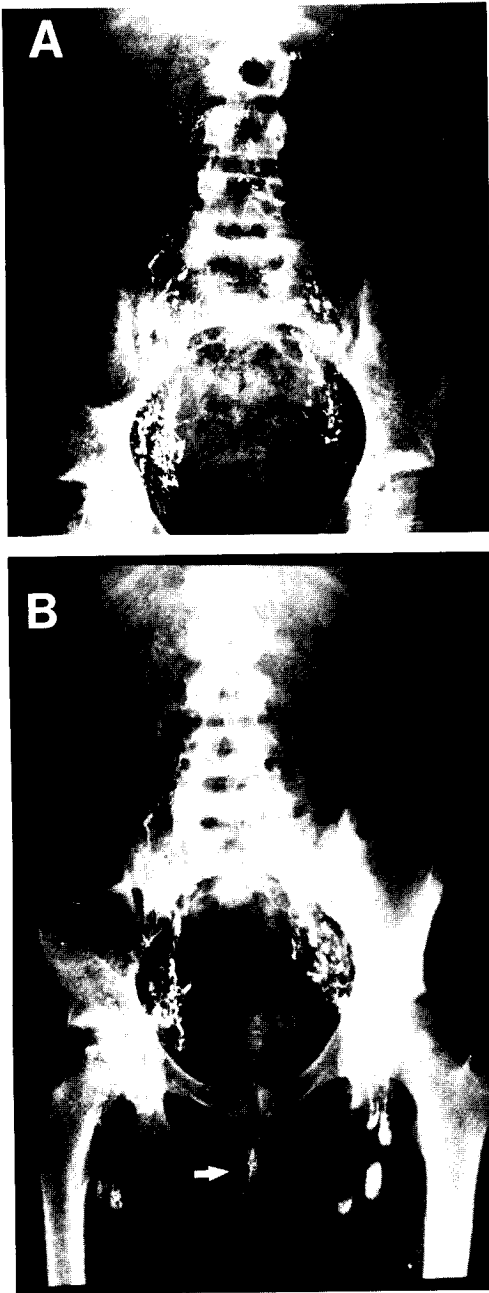


Fig. 1: Dorsal pedal lymphangiography demonstrating dilated, lymphatic sacculles of the aortic iliac trunks (A). Abnormal lymphatics are also seen draining femoral-inguinal lymph nodes (B) and extravasating into the vaginal wall (arrow). Abnormal retrograde filling of hypogastric lymphatics with reflux into lumbar channels are also seen.

years later she is asymptomatic. Specifically, there has been no recurrence of metrorrhea nor of lymph drainage from the groin.

DISCUSSION

Pathophysiology:

Intestinal absorption of long-chain triglycerides involves conversion into chylomicrons and transport by mesenteric lymphatics (lacteals). Interference with this process may result in retrograde drainage of intestinal lymph into the lower limbs (chylous edema); peritoneal cavity (chylous ascites); bowels (steatorrhea); pleural space (chylothorax); urinary tract (chyluria); pericardium (chylopericardium) and rarely the uterus, fallopian tubes or vagina (chylous metrorrhea) (1). Kinmonth and Taylor (2,3) suggest two patterns of chylous reflux. The first type shows a preference for accumulation of chylous edema in the legs accompanied by fistulization into the genitalia. Lymphangiogram typically shows megalymphatics or lymphatic varicosities. The second type occurs primarily in the pleural and peritoneal cavities where lymphatics are characteristically hypoplastic. Pathogenetically, Kinmonth maintains there is functional insufficiency of abdominal lymphatics but others favor high-grade obstruction of the thoracic duct. In most patients, however, lymphangiography fails to confirm thoracic duct obstruction and in the patient reported the thoracic duct was in fact normal.

Clinical Features:

The etiology of chylous metrorrhea is either primary (idiopathic) or secondary (e.g. filariasis, trauma, irradiation or neoplasia). Our patient corresponds to primary or unknown (Kinmonth Type I). Clinical manifestations usually first appear before puberty and commonly are misdiagnosed or confused with urachal or urinary fistulas, gonorrhea or aberrant apocrine-mammary glands. Chylous discharge may relate to positional changes (as confirmed in our patient) with exacerbation in the erect position. Discharge also in-

Table 1
Treatment of Chylous Metrorrhea by Pelvic Lymphangiectomy

Author Ref.	Year	Age of Onset (years)	Peripheral Lymphedema	Lower Limb Growth Increase	Skin Vesicles	Lymphangiogram	Operation	Follow-Up Year (y) Month (m)
Martorell (9)	1963	18	+	+	-	-	Pelvic lymphangiectomy (retroperitoneal)	20 Y.
Kinmonth (7)	1964	16	+ chyluria	+	-	+	Lymphangiectomy (transperitoneal) Charles Operation Hysterectomy	?
Kinmonth (7)	1964	20	+	+	-	+	Lymphangiectomy	1 Y
Beau (11)	1965	9	-	-	-	+	Lymphangiectomy (transperitoneal)	6 M.
Burnett McCaffrey (6)	1967	31	+	-	-	+	Lymphangiectomy (transperitoneal)	10 M.
Ma'Luf (5)	1971	18	+	+	+	+	Lymphangiectomy	15 Y.
Mitchell (10)	1978	11	- Chylous Ascites	-	-	+	Salpingectomy pelvic lymphangiectomy	3 Y.
Jiminez-Cossio	1985	9	-	+	+	+	Pelvic Lymphangiectomy (transperitoneal)	19 M.

creases with menstruation, heightened intraabdominal pressure and after sexual intercourse (4). Commonly small skin vesicles appear and rupture with accompanying lymph discharge (5). Commonly chylous metrorrhea is associated with leg lymphedema (5,9).

Treatment:

Because the syndrome of chylous metrorrhea is rare, operative and non-operative experience are limited. Indeed, only eight patients including this report were subject to definitive operation (Table 1). The outcomes varied from "complete cure" to partial remission. Early attempts to resect the fistula site were unsatisfactory with high recurrence (12-14). Mikulicz (14) described successful obliteration in a seventeen-year-old girl using irradiation to the vulva and lower abdomen. During an earlier laparotomy in this patient resection was not feasible as there were no dilated lymphatics.

Conclusions:

1. Primary chylous metrorrhea is a rare variant of the chylous reflux syndromes and the cause is unknown.
2. Symptoms often appear at school age with intermittent episodes of variable amounts of chylous discharge.
3. Symptoms may be confused with venereal diseases (e.g. gonorrhoea) or urinary fistulization.
4. Often, there is a nearby cutaneous lymphatic fistula and lymphedema of the leg(s).
5. Lymphangiography is essential for early accurate diagnosis.
6. Definitive treatment by transperitoneal lymphangiectomy is recommended.

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