Primary Chylous Lymphangiectasia of Female Genitalia

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Abstract
We report a case of a 50-year-old female who presented with multiple thick-walled vesicles involving the labia majora and minora which started developing at the age of 10 year. The vesicles extruded milky white fluid when punctured. Biochemical analysis of the fluid was consistent with chyle. Lymphoscintigraphy showed reflux of the radioactive tracer into the genitalia, thus confirming the diagnosis of primary chylous lymphangiectasia. The patient was not willing to undergo definitive surgical treatment. We report the case considering the rarity and to depict the usage of biochemical analysis of the vesicular content and lymphoscintigraphy to detect this entity.

Key Words: Chylous lymphangiectasia, congenital, female genitalia

Introduction
Primary chylous lymphangiectasia is characterized by congenitally dilated and incompetent lymphatics, resulting in the backflow (reflux) of the chyle into the abdomen, pelvis, or lower limbs.[1] We report a case of primary chylous lymphangiectasia of female genitalia.

Case Report
A 50-year-old female presented with multiple tiny fluid-filled lesions in the genitalia since 10 year of age, with a history of milky white discharge per vaginum. There were recurrent infections and pain which got resolved with antibiotics. She underwent multiple sittings of electrodessication elsewhere which were soon followed by recurrences. On examination, there were multiple thick-walled vesicles involving the labia majora and minora, which extruded milky white fluid when punctured under sterile aseptic precautions [Figure 1]. On speculum examination, the vagina was normal. Inguinal nodes were not enlarged. Per-abdominal examination did not reveal any palpable mass. Thus, the diagnosis of chylous lymphangiectasia was considered based on the clinical findings.

The vesicular fluid was odorless, and the specific gravity was more than 1.012. Triglyceride content of the aspirated vesicular fluid was 3402.9 mg/dL (blood triglyceride level of the patient being 261 mg/dL). These features are suggestive of chyle contained in the vesicles. Transperineal, transvaginal, and transabdominal ultrasonographies were unremarkable. Lymphoscintigraphy using Tc-99m and

Figure 1: Multiple thick-walled vesicles involving the labia majora and minora, with edema of the labia majora (more on the left side). One such lesion on pricking extruded milky white fluid seen on the left labia majora

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single-photon emission computed tomography revealed reflux of tracer into the genitalia, thus confirming the diagnosis [Figure 2]. Definitive surgical modalities were suggested but could not be carried out due to the patient’s financial constraints.

**Discussion**

Chyle is a milky fluid which is a mixture of lymph (from the genitalia and lower limb) and fat (absorbed from the small intestine). Lacteals of the small intestine and the lymphatics from the lower limbs and genitalia drain into the cisterna chyli.

Primary developmental abnormalities of the lymph vessels (lymphangiectasia) or secondary causes (tumor and trauma) lead to accumulation of the chyle and disruption of the lymphatics, causing chylous lymphangiectasia in the genitalia (scrotum in male and labia in female), perineum, or legs. The other presentations of chylous reflux include chylothorax, chylous ascites, chyluria, intestinal lymphangiectasia (manifesting as protein-losing enteropathy), and edema of the lower extremities. These features are postulated to be due to the valvular incompetence of the lymphatic channels or atresia of the lymphatics at or above the level of the cisterna chyli.

Primary chylous lymphangiectasia of the genitalia presents as edema of the genitalia, persistent vaginal discharge of chylous fluid, or lymphangiectatic vesicles which when ruptured extrude milky fluid mixed with chyle and lymph, similar to the present case.

Although this entity is congenital, onset occurs commonly during teenage (age of onset ranging between 1 day and 81 years). The discharge of chyle from cutaneous vesicles interferes greatly with quality of life. They also serve as entry point of bacteria causing cellulitis or lymphangitis. Malnutrition due to depletion of lipids and proteins and impaired immunity due to loss of immunoglobulins may occur.

The characteristic features of the chylous fluid from the lymphangiectatic vesicles are as follows:
- Milky appearance
- Fat (milky portion) floats and lymph (clear fluid) settles on standing
- Odorless
- Specific gravity > 1.012
- Triglyceride levels > 110 mg/dL

In our patient, the fluid from vesicles was milky and odorless, had specific gravity more than 1.012, and had elevated triglycerides. The milky white color distinguishes this entity from the mere lymphangiectasia where clear fluid will be contained in the vesicles.

Lymphoscintigraphy is an initial diagnostic test. Lymphangiography is more confirmatory as it exactly depicts the dilated retroperitoneal lymphatics and localizes the site of chyle reflux. Magnetic resonance imaging also delineates the dilated lymphatics. In the present case, lymphoscintigraphy showed backflow of the chyle into the genitalia. However, lymphangiography could not be done due to patient’s financial constraints.

The aim of definitive surgical treatment is to stop the chylous reflux by diminishing retrograde flow of chyle and diverting the incompetent lymphatics into the venous system. The surgical options also include excision, ligation, or sclerotherapy of the incompetent lymphatics, which can be combined with lymphovenous anastomosis or lymphatic bypass grafting. Since the pathology of this disorder lies deeper, the mere ablation of the vesicles will lead to frequent recurrences as witnessed in our patient.

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**Conflicts of interest**

There are no conflicts of interest.

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