A Case of Nonvenereal Sclerosing Lymphangitis of the Penis

Hyohyun Ahn, M.D., Aeri Kim*, M.D., Il Hwan Kim, M.D.

*Department of Dermatology, Department of Pathology**,
College of Medicine, Korea University, Seoul, Korea

The patient was a 41-year-old healthy man, who developed a tender, cord-like serpiginous mass just proximal to the coronal sulcus for two weeks. He was a sexually active, non-promiscuous, married man. We had taken a biopsy, and noticed the subsiding of the lesion without further treatment.

Nonvenereal sclerosing lymphangitis of the penis is a rare self-limiting peculiar disorder involving the lymphatics of the penile sulcus. Clinically, it presents as a cord-like nodular penile lesion with characteristic cartilaginous firmness. Histologically, it is described as hypertrophy and sclerosis of the lymphatic vessel walls with mild inflammatory cellular infiltration, and occasional obstruction of the lymphatic vessel. But, because such features including sclerosis varies according to the time when the biopsy was taken, they are not attributable to all cases. Our case shows the same clinical and pathological features of 'benign transient lymphangiectasis'. Except for the painful cases, no specific treatments are usually warranted.

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Key Words : Sclerosing lymphangitis, Penis, Factor VIII related antigen, CD-34

Nonvenereal sclerosing lymphangitis(NVSL) of the penis is a rare asymptomatic and benign disorder of men 20 to 40 years of age[1]. Clinically, it occurs as a peculiar penile lesion just proximal to the coronal sulcus, and abates spontaneously even without treatment, rarely bringing to the attention of the physician. Because of the self-limiting and benign nature of the disease and patients' unawareness, there could be more cases unreported.

CASE REPORT

A 41-year-old man presented a mildly tender, skin-colored, cord-like, serpiginous and nodular mass just proximal to the coronal sulcus lasting for less than two weeks (Fig. 1-left). It was cartilaginous, firm and freely movable on palpation. He was sexually active but a non-promiscuous married man. He had no history of major medical illness and viral infections. And the physical examination showed nothing unusual except above findings. There was no inguinal lymphadenopathy. The routine laboratory examinations including VDRL were negative or within normal limits. We had taken a biopsy of the lesion on his first visit. The lesion gradually improved spontaneously after the biopsy was taken (Fig. 1-right).

The histopathologic examination showed a few dilated lymphangiectatic spaces, lined by flat endothelium. The surrounding stroma showed sparse inflammatory infiltrate. There was no red blood cell in the lumina (Fig. 2). Immunohistochemically, Ulex Europaeus Agglutinin-I (UEA-I) stained slide showed negatively stained endothelial cells of luminal surface. The CD-34 (Fig. 3) and factor VIII-related antigen (Fig. 4) stained slide also showed negatively stained luminal surface, contrasting positively stained vascular walls located in the vicinity.

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Reprint request to : Hyo Hyun Ahn, M.D., Department of Dermatology, College of Medicine, Korea University, Seoul, Korea

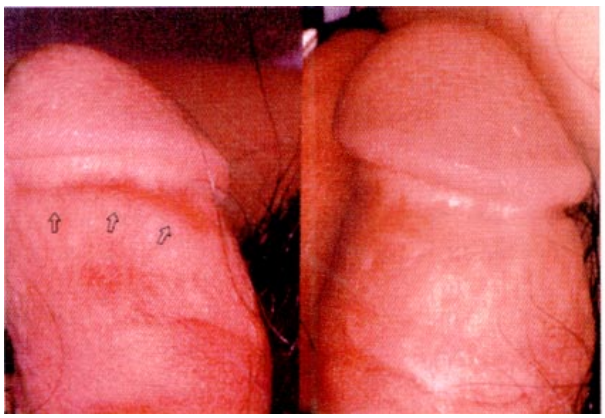

Fig. 1. A clinical photograph showing a cord-like mass on penile shaft, just proximal to coronal sulcus (left - pretreatment, lesion indicated by arrows, right - follow-up)

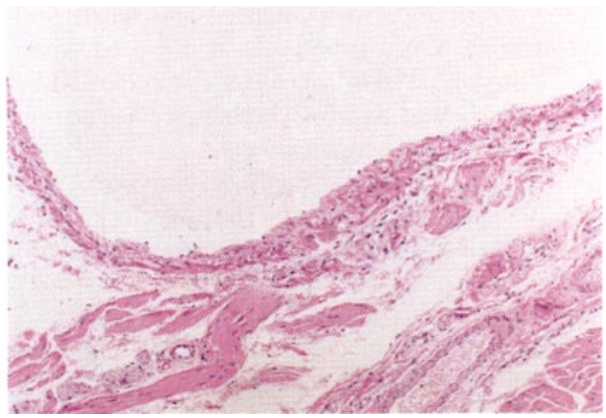

Fig. 2. A photomicrograph showing a few dilated spaces lined by flat endothelium. The surrounding stroma showed increased collagen and sparse inflammatory infiltrate(H&E, ×100).

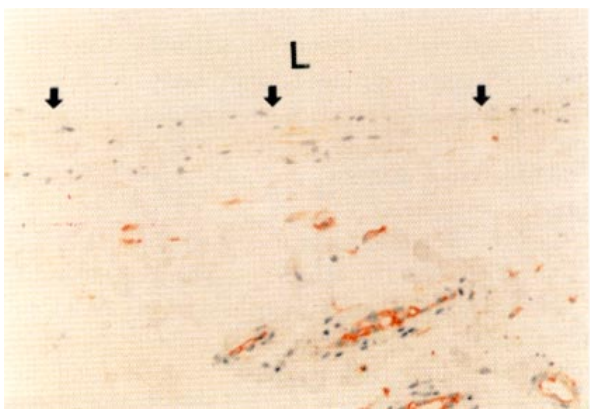

Fig. 3. A immunohistochemical stain for CD-34, endothelial cells of luminal surface stained negative (CD-34, ×200, L: lumen indicated by arrows).

Fig 4. A immunohistochemical stain for factor Ⅷ-related antigen, endothelial cells of luminal surface stained negative (factor VIII, ×200).

DISCUSSION

NVSL of the penis is a very rarely reported disorder, which occurs only in sexually active men. Hoffman originally reported this disease as 'simulation of primary syphilis by gonorrheal lymphangitis' in 1923, and later named it 'nonvenereal plastic lymphangitis of the penis' recognizing the nonvenereal nature of the disorder[2]. In 1962, Nickel and Plumb coined the term 'sclerosing lymphangitis of penis'[3], and this is the term most commonly used. But, Hutchins[4] and McMillan[5] preferred to call it as 'benign transient lymphagiectasis of the penis' and 'localized lymphoedema' respectively, because of the minimal inflammatory reaction seen on histologic sections, not justifying the specific term 'sclerosing' in every case. And, the reviews and observation by Hutchins showed lymphangiectisis in early period, sclerosing lymphangitis in more advanced period (more than 2 weeks) and occasional thrombotic occlusions in persistent lesions[4]. So, it may be inferred that the same disease process could be described as benign transient lymphangiectasis, sclerosing lymphangitis, and lymphangifibrosis thrombotica occlusiva by many authors as to the time when the biopsies were taken. Other synonyms such as 'circular indurated lymphangitis of the penis' are still found in the literature[6] and our textbooks[7,8], by the similar reason. In Korea, there have been only a few cases reported in dermatologic[9,10] and radiologic[11] literature. In Korean dermatologic literature, all cases reconciled with

classic descriptions, with emphasis on sclerosis and hypertrophy. Only two weeks had passed when the biopsy was taken in our case, and accordingly the specimen showed mainly lymphangiectasia rather than sclerosis and lymphangitis.

Clinically, it begins with the sudden onset of a cord-like thickening of tissue that develops at the corona or just proximal to the coronal sulcus with characteristic cartilaginous consistency[1], usually developing within a short time of sexual intercourse[12]. Occasionally, the lesion may extend proximally to the penile shaft[8]. The overlying skin is not attached to the lesion, making them freely movable[3,9]. Our case also showed the penile lesion just proximal to the coronal sulcus. From a clinical point of view, this location is important for diagnosis.

Even though most patients complaint of neither tenderness nor discomfort, some patients reported including our case, feeling irritation, tenderness or painful erection before the appearance of the lesion[1,9,12,13]. Occasionally, it can be associated with asymptomatic edema of sulcus and glans penis[14], and rarely with inguinal lymphadenopathy[5].

The cause of this disease is controversial. Since it tends to develop in men who are hyperactive sexually, it is likely that trauma plays an important role. Other possible causes are irritation from menstrual blood, and infective causes such as tuberculosis, viral infection, and Chlamydia[1,12,15]. Some authors suggested the association with genital herpes[15], and Chlamydia trachomatis[16]. But, generally, most cases, including ours, did not have the associated history of venereal disease[1,9,10,12,13]. Because of the rarity of this disorder and some patients reportedly with recurring symptoms, the anatomical variation was proposed as a predisposing factor[17]. Overall, most cases, including our case, are probably primarily caused by trauma.

Controversy exists as to the nature of the vessel involved[8]; a recent study presented data suggesting that the vessels were lymphatic in origin[18]. Histologic features are hypertrophy and sclerosis of the lymphatic vessel walls with mild inflammatory cellular infiltration and occasional obstruction of the lymphatic vessel[1,12]. Older lesions may show evidence of recannalization[8]. But, these features are not attributable to every case, because of the spectral nature of the extent of them, especially sclerosis. As shown in Fig 2, there is not much sclerosis around the vessels but the lymphangiectatic space and sparse inflammatory infiltrate in histologic section of our case, supporting the view of some authors in insisting on not using the term 'sclerosis'[4-8].

Immunohistochemically, UEA-1 stained slides show positively stained luminal surface, and factor VIII-related antigen stained slides reveal negatively stained luminal surface, as a supporting evidence of lymphatic origin[19]. The lymphatic vessels are differentiated based on 1) radial shaped lumen, 2) absence of blood cells in lumen, 3) lack of uniformity in the thickness of the vessel wall, 4) the particular arrangement of components comprising vessel wall, and 5) negative staining for factor VIII-related antigen on luminal surface and wall[19]. But, it is not always possible to differentiate between vascular and lymphatic origin histologically, even when assisted immunohistochemically.

The differential diagnosis includes lymphangioma circumscriptum, foreign body granuloma, lymphogranuloma venereum, syphilitic chancre, pyogenic infection and most importantly superficial phlebitis (Mondor's disease), which has bluish hue[1,20].

Cessation or reduction in sexual activity is recommended, and the lesion resolves spontaneously over a period of 1 or 2 months[1]. So, to avoid unwarranted surgical procedures and antibiotic or antiviral therapy to this benign self-limiting disease, the proper diagnosis is needed. But, there were recurrent cases, warranting long-term follow up[10]. Surgical removal of the indurated tissue can be carried out in the rare instance in which tenderness is troublesome and persistent[13].

This is a case of NVSL of the penis occurring on a healthy middle-aged man, which showed another end of the spectrum of histologic features. And, we reported with a brief review on the disease.

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