Case Report

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A Rare Case of Cutaneous Lymphangiectasia of the Vulva Secondary to Tuberculosis

Kirana Pailoor*, Muktha R. Pai, Hilda Fernandes***, Rohan Chandra Gatty****, Jayaprakash C.S.*****, Nisha J. Marla***

*Associate Professor ***Professor *****Professor & Head, Dept. of Pathology ****Associate Professor, Dept. of Onco. Surgery, Father Muller Medical College, Kankanady, Mangaluru, Karnataka 575002, India. **Professor, Dept of Pathology, A.J. Institute of Medical Sciences and Research Centre, Mangaluru, Karnataka 575004, India.

Abstract

We present an interesting case of acquired cutaneous lymphangiectasia of vulva following tubercular lymphadenitis. It was a 35 year old lady who initially had cervical lymphadenitis proven positive for Tuberculosis and received Anti- Tubercular treatment about thirty years ago. She developed multiple raised lesions over the vulva and left upper thigh of one year duration. On examination, the patient had a large polypoidal growth involving both sides of the vulva, left upper thigh and over pubic area. She underwent simple vulvectomy and left thigh growth excision. Histopathological examination of the vulvectomy specimen confirmed the clinical diagnosis of vulval lymphangiectasia.

Keywords: Acquired Vulval Lymphangiectasia; Vulval Lymphangiectasia; Tuberculosis.

Introduction

Cutaneous Lymphangiectasia, also called as acquired lymphangioma, is a benign cutaneous disorder involving the dermal and subcutaneous lymphatic channels.It is characterised by the presence of a circumscribed eruption of thin-walled translucent vesicles.When it arises on the vulva, diagnostic and treatment difficulties pose a challenge. It has to be differentiated from lymphangioma circumscriptum which is a developmental defect of the deep dermal and subcutaneous lymphatics, although clinically and histologically they have similarities. However, they can be differentiated based on the history because lymphangiomas are present since birth or early childhood, while acquired cutaneous lymphangiectasia develop later and are associated with various causes. The aim of this report is to bring forth unusual clinical manifestations following childhood tuberculosis, the prevalence of which is high in our country.

Corresponding Author: Kirana Pailoor, Associate Professor, Department of Pathology, Father Muller Medical College, Kankanady, Mangaluru, Karnataka 575002, India.
E-mail: dockirana@yahoo.co.uk

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Case Report

A 35 year old woman came with multiple raised lesions over the vulva and left upper thigh of one year duration. She gave history of getting treated for multiple swellings that developed over right side of the neck and inguinal regions on both sides three decades ago. On local genital examination , the patient had a large polypoidal growth involving both the sides of the vulva, left upper thigh and over pubic area. Multiple linear scars were present over upper thigh and groin bilaterally. The patient underwent simple vulvectomy and left thigh growth excision.The skin surface of the vulvectomy specimen showed a multinodular warty growth measuring 11x4x2 cm [Figure 1]. The cut surface was grey white, firm with focal hemmorhagic areas [Figure 2]. The left thigh growth showed a raised nodular lesion measuring 1.5x0.5x0.3 cm. The cut surface was grey white and firm in consistency. Histopathological examination of the vulvectomy specimen revealed thin-walled and ectatic lymphatic channels in the superficial dermis. The dermal papillae and the dermis showed proliferating , congested capillary sized blood vessels, melanophages and diffuse as well as perivascular mixed inflammatory infiltrate [Figure3, 4]. Section

from the growth over left medial thigh showed a similar picture. With the above findings, we came to a diagnosis of lymphangiectasia of vulva.

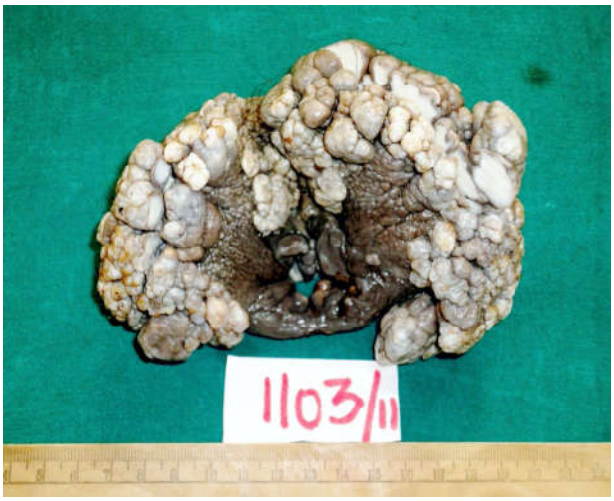

Fig. 1: Shows multinodular warty growth

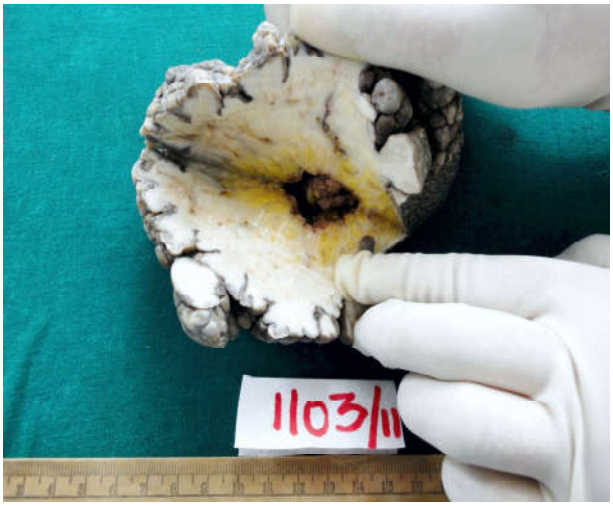

Fig. 2: Cut surface showing grey white areas

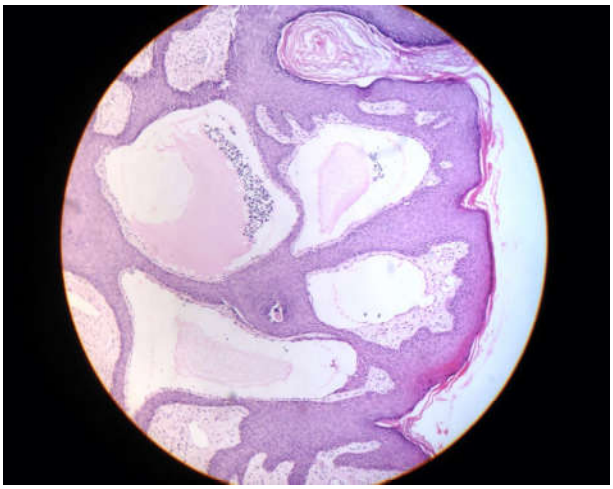

Fig. 3: Showing thin walled, ectatic lymphatic channels (H & E x 4X)

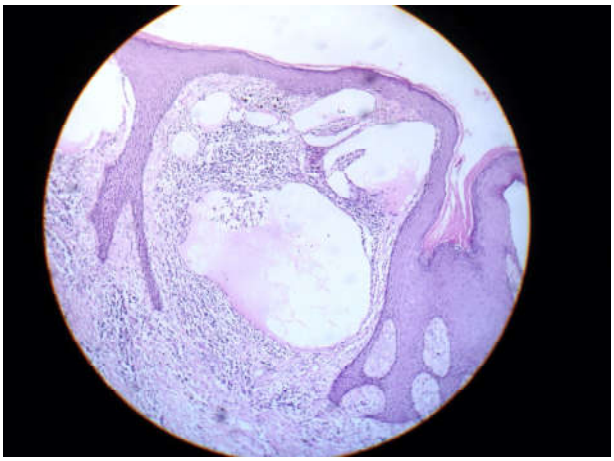

Fig. 4: Showing melanophages, proliferating blood vessels and perivascular mixed inflammatory infiltrate (H & E x 10X)

Discussion

Cutaneous lymphangiectasia is a term applied to lesions consisting of one or multiple groups of translucent vesicles, which represent an acquired or congenital dilatation of lymphatic channels [1-4]. They are not true neoplasms or hamartomas. They arise due to the obstruction of the lymphatics secondary to radiation, surgery, trauma, keloid, scrofuloderma, neoplasia or infections such as filariasis, tuberculosis, recurrent erysipelas and lymphogranuloma venereum [1-7].

Vulval lymphangiectasia is a rare disease and is usually reported following surgery/ radiotherapy for carcinoma of the cervix or vulva, tubercular inguinal lymphadenitis or Crohn's disease of the vulva [1-6]. The lymphatic vessels of the superficial dermal plexus drain a fixed area of skin through the vertical collecting lymphatics to the deep plexus. Damage to the deep lymphatic vessels leads to back-pressure and dermal backflow, with subsequent dilatation of the upper dermal lymphatics [4,6]. The lesions are typically known to arise approximately 7 to 15 years after lymph node dissection and radiotherapy of the genitalia.

It affects females aged 20 to 75 years. Heuvel et al described a case of vulval lymphangiectasia secondary to lymphnode tuberculosis , exactly similar to our patient [2]. Vulval lymphangiectasia can be asymptomatic, pruritic, burning or painful. It is an unpleasant but benign condition. Our patient was asymptomatic. Clinically, lymphangiectasia is characterized by thin-walled translucent vesicles filled with clear colourless fluid. Our patient had diffuse polypoidal mass with hyperkeratotic surface on the genitalia and vesicles on the extragenital sites.

Histologically, dilated lymphatic channels are present in the superficial and mid-dermis and a few dilated lymphatics are seen in the deep dermis. The overlying epidermis may display varying degrees of hyperkeratosis, acanthosis and papillomatosis and it may appear to enclose the ectatic lymphatic channels. The dilated lymphatic channels may contain scattered lymphocytes and red blood cells. This entity has to be distinguished from lymphangioma circumscriptum (LC) which is a congenitally derived hamartoma with early onset of the lesions. Histologically, LC tends to have more extensive involvement of the deep dermis and subcutis. Other conditions such as mucin secreting metastatic adenocarcinoma mimicking acquired lymphangioma, benign lymphangio-endothelioma, syringoma and condyloma should be ruled out [2-7].

Lymphangiectasia of the vulva can be often misdiagnosed as herpes, genital warts , molluscum contagiosum or sometimes as malignancy due to the huge growth [1-5]. However, oozing vesicles or papules and recurrent infections should bring into mind the possibility of lymphangiectasia. Diagnosis and treatment of this lesion is important as it may be associated with pain, chronic oozing and infection, occasionally leading to cellulitis. The diagnosis is mainly clinical, aided by histopathological finding of dilated lymphatics in the dermis [2-7].

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