Case report

LYMPHANGIOMA CIRCUMSCRIPTUM

M. Penko, I. Bartenjev, B. Žgavec and M. Potočnik

ABSTRACT

Diagnosis of lymphangioma circumscriptum was made in a 21-year-old patient with several, clearly demarcated skin lesions, with the diameter of 2 - 8 mm, on scrotum, penis and pubic area as well as on upper medial parts of thighs. Skin alterations appeared clinically as warty lesions and vesicles. The first skin changes appeared when he was 17. When he was 5, he was submitted to the surgery for bilateral cryptorchidism. There was a sporadic outflow of transparent liquid in the affected areas. Histopathological examination showed dilated lymph vessels in papillary and reticular dermis. In some segments the dilated lymphatics extended to the epidermis that was atrophic in these areas.

Radioisotopic lymphography of the affected area revealed a superficial lymphedema where the outflow was extremely slowed down. A communication between skin lesions and lymphatic vessels of pelvic and inguinal area was noticed.

Lymphangioma circumscriptum shows highest incidence in infancy, usually it is present by the age of 5, but it may appear in adolescence and adult life. However, acquired lymphangiomas appear years after the occurrence of predisposing factors, such as surgical procedures, keloids, infections, scleroderma and radiotherapy.

The question to which extent the operation of cryptorchidism played a role in the pathogenesis of lymphangioma in our patient remains open. The patient left the hospital immediately after the diagnostic procedures were done.

KEY WORDS

lymphangioma circumscriptum, acquired lymphangioma, case report

INTRODUCTION

Lymphangioma circumscriptum of the skin is a relatively rare disorder of lymphatic vessels in which the main features are sacculary dilated superficial lymphatics lined by a single layer of endothelial cells. Clinically it appears as clusters of thin-walled translucent vesicles, usually described as "frog spawn" but sometimes there are only a few scattered vesicles within a circumscribed area. They are mostly filled with a clear colorless fluid. Occasionally, because of an admixture of blood, their color ranges from pink through red to black, depending on amount of erythrocytes and their degradation products they contain. (1)

Lesions can affect any area of skin showing a predilection for the neck, axilla, breasts, chest, buttocks and thighs (2). Cases of tongue and vulva involvement were also reported. (10,7)

Classification of lymphangioma circumscriptum remains rather complicated. The reason is probably the fact, that there are no certain specific histologic (2), ultrastructural (3) and clinical (7,8) criteria to distinguish lymphangioma from acquired lymphangiectasias due to lymph stasis secondary to an external cause.

Lever (4) classified lymphangioma circumscriptum into two types: lymphangioma circumscriptum defined by Whimster (1) as the "classical form" that may also be considered as congenital and localized type of lymphangioma circumscriptum. He mentioned lymphangiectasias separately. However localized type was described by other authors also as acquired lymphangioma or lymphangiectasias (11,3) and because of their similarity they do not take them as separate entities.

The classical type according to Whimster (1) has a congenital underlying cause and is characterized by large lymphatic cisterns a few centimeters in diameter which are situated in deep subcutaneous tissues. The cisterns are surrounded by thick muscular walls and their regular contractions apply an increased pressure via the large lymphatic vessels running mainly vertically through the dermis to the superficial, apparently previous normally appearing, lymphatics which subsequently sacculary dilate.

The whole pathological system does not communicate with the general lymphatic system but represents a sequestrated part of it.

This kind of lymphangioma circumscriptum shows highest incidence in infancy, usually it is present by the age of 5, but may appear in adolescence and adult life. (1,2,7,10)

Acquired lymphangiomas show no pre-existing lymphatic abnormalities and vesicles develop as saccular dilatation of superficial lymphatics as a result of compromised lymphatic drainage from affected area. The known extrinsic factors which may cause such conditions include scarring from surgical procedures (12-14), keloids (15), infections (16), scleroderma (17) and radiotherapy. (11) Clinically evident lesions appear years after the occurrence of predisposing factors, in most cases between 8 and 25, but can be evident earlier. (7,8) In such lesions lymphangiography usually reveals a communication with the main lymphatic drainage system.

CASE REPORT

The 21-year-old patient was admitted to the Department of Dermatology because of multiple warty lesions and some small vesicles in the pubic area and on the scrotum. Periodically a clear, water-like fluid was discharged from the mentioned area. According to his statement the troubles had started approximately five years earlier. The patient was operated for bilateral cryptorchidism at the age of 5 and for stenosis arteriae pulmonalis at the age of 8. (Fig. 1 and Fig. 2)

Dermatologic examination revealed numerous whitish warty lesions, a few millimeters in diameter, in the pubic area and on the scrotum. The lesions extended to the proximal part of the penis and two of them were expressed on the inner, proximal side of both thighs. On palpation a water like fluid was occasionally oozing from certain lesions. At general examination a number of carious teeth and pectus infundibuliforme were noted. The routine laboratory tests were within the normal limits.

The biopsy taken from the pubic area revealed a hyperkeratosis with small keratotic plugs in certain follicles as well as an irregular acanthosis and papillomatosis. In the papillae dilated lymph-vessels of different parameters, lined up with a single layer of endothelial cells, were seen. In some of this vessels an admixture of erythrocytes was observed. Certain of them were surrounded by downward proliferating epithelium. One major lymphatic dilatation seemed to be located intraepidermally and the epidermis covering it was atrophic. A few dilated lymph vessels were observed in the reticular dermis. One follicle was surrounded by a modest inflammatory infiltrate. The histopathologic diagnosis was lymphangioma circumscriptum. (Fig. 3)

Radioisotopic lymphography with 99mTc nanokolloid of scrotum and medial parts of the thighs was performed at Department of Nuclear Medicine. The radiolabelled material was injected into one warty lesion on the right side of the scrotum. The reagent penetrated into the inguinal, pelvic and right-side paravertebral lymphnodes and later on also into the left sided lymphnodes. The investigation revealed a superficial lymphedema of the scrotum and medial parts of the thighs, where the outflow was extremely slowed down. It seems that the warty lesions communicate with lymphatics in the inguinal and pelvic areas.

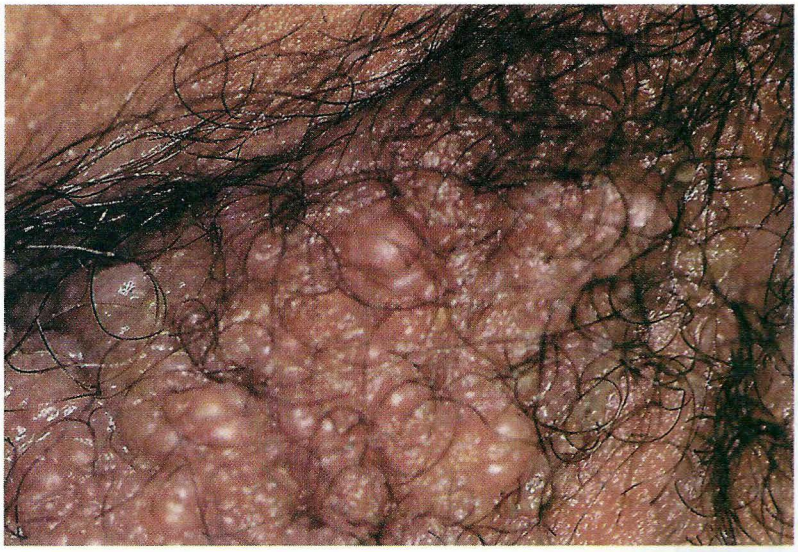

Fig. 1. Numerous whitish, warty lesions in the pubic area.

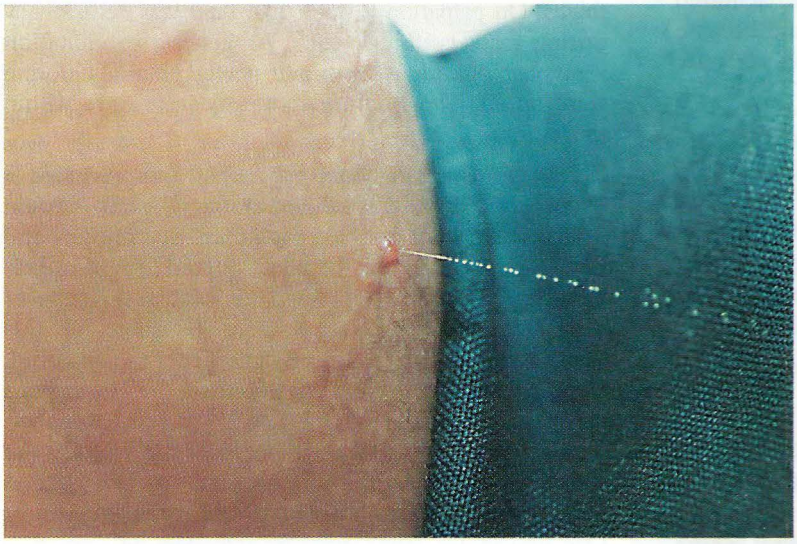

Fig. 2. A jet of water-like fluid from a lesion on the thigh.

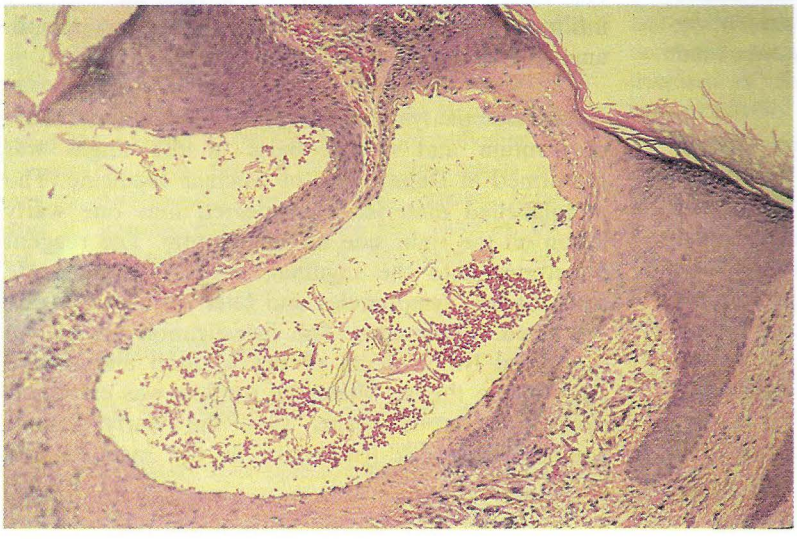

Fig. 3. Histopathology of a lesion from the pubic area:
Cystically dilated lymph vessels containing an admixture of erythrocytes in the papillary dermis. The epidermis displays irregular acanthosis and atrophy in certain areas.

DISCUSSION

It is obvious that in lymphangioma circumscriptum raised intralymphatic pressure plays the main role in pathogenesis, and the subsequent dilatation of apparently normal pre-existing superficial lymphatic vessels represents the pathological substrate for this disorder. Furthermore, it seems that there is no evidence of any benign autonomous pathological overgrowth of any lymphatic components. (3)

The diagnosis of lymphangioma circumscriptum is based on clinical features, history, histopathology, lymphography as well on further functional tests.

The existence of lymphangioma circumscriptum was in the case of our patient unequivocally proved by the mentioned diagnostic procedures. The question to which extent the operation of cryptorchidism played a role in the pathogenesis remains open. The consulted urologist stressed that according to their clinical experiences lymphangiomas were not observed following this procedure.

It is worth to mention that the operation of the stenosis of the pulmonary artery probably indicates a congenital weakness of the circulatory system.

Despite its benign nature, this condition is annoying for the patient, mostly because of frequent discharge of fluid from vesicles which are easily traumatized and may also represent an important entry for infection.

Treatment of lymphangioma circumscriptum which has a tendency for recurrings is usually unsatisfactory. Different therapeutic approaches are used.

Removing or sealing superficial lesions only by surgical excision or further techniques e.g. CO_2 or argon laser, electrocoagulation or cryotherapy, is not promising as vesicles reappear shortly after treatment. In the case of "classical" type of lymphangioma circumscriptum some good results were obtained by surgical excision of the cisterns at the level of the deep fascia leaving the superficial vesicles intact.

A successful control of lymphangioma circumscriptum by superficial X-rays has been reported, but this is still a matter of discussion. Quite often such lesions are best left untreated and perhaps compression through bandaging or hosiery, if the site is appropriate, can be helpful.

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