CASE REPORTS

LYMPHANGIOMA CIRCUMSCRIPTUM

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Clinical features of six cases of lymphangioma circumscriptum are described.

Key words : Lymphangioma circumscriptum.

Lymphangioma was first described in 1879 by Tidbury Fox and Calcott Fox under the name lymphangiectodes.[1] Malcom Morris coined the term lymphangioma circumscriptum in 1889.[1] Three types of lymphangioma exist, (1) localized lymphangioma circumscriptum, (2) the classic type of lymphangioma circumscriptum, and (3) cavernous lymphangioma, including cystic hygromas.[2]

Cutaneous lymphangioma show the highest incidence of onset in infancy, the majority are present by the age of five years, but these may appear spontaneously in adolescence or adult life.[3] The lymphangioma can involve any area of the skin, but are commonly seen on the anterior and lateral parts of the chest including the breast, thighs, buttocks, neck and axilla in the descending order.[3] Female preponderance in lymphangioma was reported by Peachy[4] and Flangan,[3] but both attribute this to cosmetic reasons.

In the classic type of lymphangioma circumscriptum, one or several large patches with translucent vesicles are present. The area involved in the classic variety of lymphangioma circumscriptum is several sq cm; while in the localized variety it is less than one sq cm.[4] In many instances, there is a diffuse swelling of the subcutaneous tissue beneath the lesions and the adjacent area. In very rare instances, there is associated enlargement of that part of the limb. Some of the vesicles contain an admixture of blood. The skin between and even on top of some of the vesicles may be verrucous in appearance.[2]

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A 14-year-old male had skin lesions on his right thigh with a history of secondary infection and thin, purulent discharge off and on since childhood. The lesions present over the antero-lateral side of upper half of right thigh (Fig. 1) were hyperpigmented or skin-coloured papules, nodules and vesicles, of different sizes ranging from a few mm to one cm. A few lesions had a verrucous surface and a few were compressible on diascopy. There was diffuse thickening on the lateral aspect of the right thigh. Right side inguinal lymph glands, both vertical and horizontal group, were enlarged, non-tender and discrete. General and systemic examination did not reveal any positive findings.

The other five patients had similar lesions at different sites. The lesions were both grouped and solitary. There were three female patients aged 12, 22 and 35. The involved sites were right side chest, left axilla and left lower back respectively. The remaining two patients were males aged 13 and 25. The younger male patient had lesions on the pubis and perinium while the older one had lesions on the scrotum and shaft of the penis.

Fig. 1. Papular and verrucous lesions of various sizes on right thigh.

Enlargement of the draining lymph glands was seen in three cases who gave history of repeated sepsis. Diffuse thickening below the superficial skin lesions was observed in three cases. In four cases, onset was from early childhood while the patient with scrotal lesions gave only eight months history.

Routine investigations in all cases were normal. Histopathological study was carried out on 5 cases. In 4 cases, patchy hyperkeratosis and parakeratosis with marked irregular acanthosis were seen. Papillary dermis had multiple dilated spaces containing lymph, and lined by a single layer of flat endothelial cells. Few of the spaces contained RBCs (Fig. 2). In one case, papillary dermis was normal but a solitary dilated vesicle enclosed in the epidermis was seen. We were not able to see large cisterns with well-developed muscular coat in the deeper layers as described by Lever and Lever.[2]

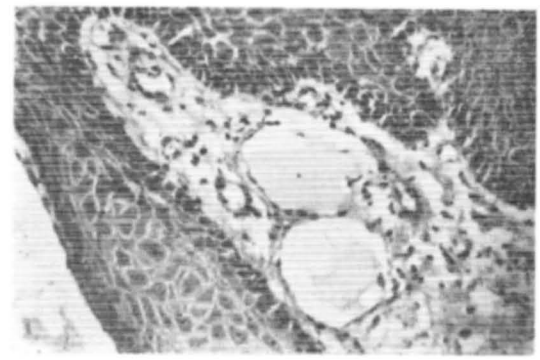

Fig. 2. Lymph spaces lined by a single layer of flat endothelial cells.

Comments

The diagnosis of lymphangioma circumscriptum may be confused with (1) tuberculosis of the skin, (2) verruca vulgaris, and (3) verrucous nevus. Two of our cases had prolonged antitubercular treatment and one case had undergone electro-cauterization.

Clinical features which help the diagnosis include, (1) history of watery discharge starting spontaneously or after unrecognized trauma, (2) repeated bacterial infection with lymphangitis, and (3) compressibility of some lesions, along with diffuse thickening of the subcutaneous tissue underlying the superficial lesions.

Surgery was performed in three of our cases and there was no recurrence during a follow up period of six months to two years. Flangan and Helwig[3] had reported that a single surgical excision cured 78% of cutaneous lymphangioma and re-excision cured additional 12%. Peachy et al[4] attribute recurrence to the difficulty in obtaining adequate excision in the area and depth.

References

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