Asymptomatic linear papules and vesicles on lower abdomen

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CLINICAL FINDINGS

A 12-year-old female patient presented with multiple progressively enlarging, irregularly shaped papules with flesh and pigmented colors as well as persistent translucent vesicles that contained clear fluid, variable in size located on the right side of lower abdomen, of 9 years duration. It had not responded to topical steroids, salicylic acid. Cryotherapy was tried but with minimal and transient improvement. According to the patient it started as small papules that gradually increased in size with appearance of vesicles that contain clear fluid. The patient did not complain of any systemic illness, and there was no family history of similar lesions.

Clinical examination of lower abdomen revealed multiple clusters of translucent vesicles that contained clear fluid in some lesions and pink to dark red fluid with hemorrhage in other lesions. The lesions ranged from minute vesicles to small bullae, with scattered hyperkeratotic warty like papules. There was no tenderness or pain on palpation (Fig. 1-4). Rest of the dermatological and systemic examination of the patient was normal. Nails and mucous membranes were not affected and showed no significant abnormalities. Routine laboratory investigations including CBC, blood sugar, hepatic and renal profile revealed no abnormal findings. X-ray of abdominopelvic region showed no involvement of underlying bones.

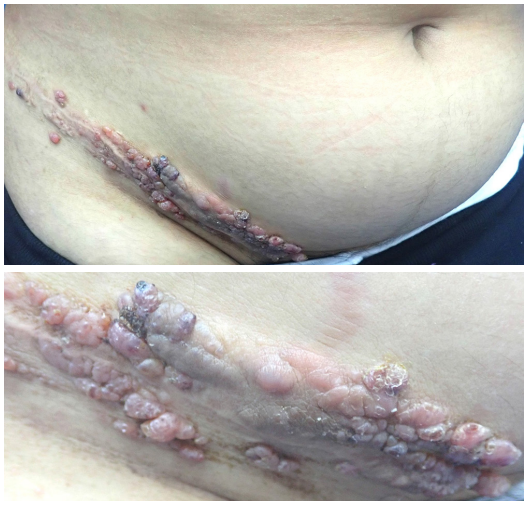

Fig. 1&2 Grouped shiny papules and vesicles with clear fluid affecting lower abdomen.

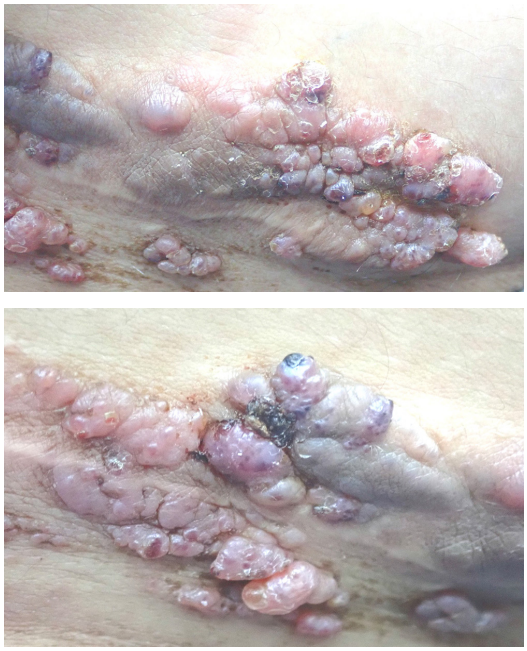

Fig. 3&4 Multiple clusters of translucent vesicles containing clear fluid in some lesions and pink to dark red fluid with hemorrhage in other lesions.

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What are your clinical differential diagnoses?

- Warts
- Lymphangioma circumscriptum
- Angiokeratoma
- Herpes zoster
- Metastatic carcinoma of the skin

Incisional biopsy was performed and histological analysis revealed hyperkeratosis, acanthosis and elongated rete ridges. The papillary and upper reticular dermis showed multiple dilated lymphatic space lined with plump endothelial cells and containing lymphatic fluid (Fig. 5,6).

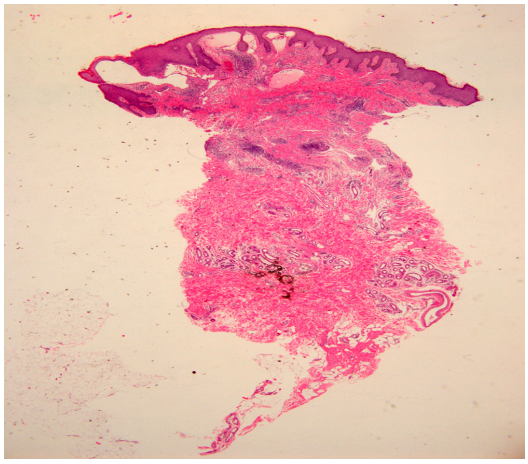

Fig. 5 Dilated vascular spaces in upper and mid dermis.

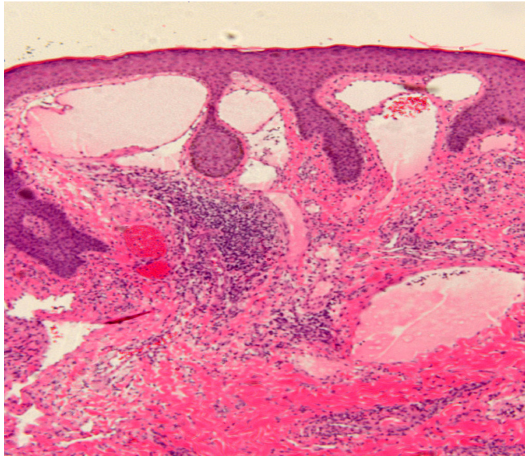

Fig. 6 Dilated lymphatic spaces lined with plump endothelial cells and containing eosinophilic fluid.

DIAGNOSIS:

Lymphangioma circumscriptum

DISCUSSION

Lymphangiomas are rare malformations of the lymphatic ducts and may affect any cutaneous or mucous surface.[1-3] They can be classified into localized or generalized malformations and have a congenital or acquired origin.[2,3] Lymphangiomas may also be classified as simple, cavernous, and cystic[4] that is called simple cutaneous lymphangioma,[5] which is renamed circumscribed lymphangioma.[6] By time lymphangiomas were grouped according to both the depth and the size of the abnormal lymphatic vessels into two categories: superficial, in the form of cutaneous lymphangioma circumscriptum (CLC), and deep, that include cavernous lymphangioma and cystic hygroma.[2]

Lymphangioma can be evident at any age, but the greatest incidence occurs at birth or early in life. About 50% of lymphangiomas can be diagnosed at birth, and most lymphangiomas are evident by the 5th year of life.[11]

Lymphangiomas are rare vascular neoplasm that represent about 4% of the vascular tumors and around 25% of benign vascular tumors in childhood.[2,3,7,8] This congenital malformation may be dating since birth in approximately 50% of cases.[1,2] On the other side, the tumour arises until two years of age. In 90% of cases and it is not a gender-related disease.[2,7]

Cutaneous lymphangioma circumscriptum, that is recently called superficial lymphatic malformation (SLM), is a rare benign tumor but it is the most common type of lymphangioma.[1,3] It affects both the skin and mucous surfaces, mainly head and neck, proximal extremities, buttocks, chest,

armpits, and oral cavity.[1,3] The most common presentation in the form of a group of translucent vesicles varying in size from 2-4 mm, similar to frog spawn, but with variable colors as a result of hemoglobin degradation.[2,3,7] Lesions may also present with hyperkeratotic and/or verrucous plaques.[7]

Although the lesion may appear localized to the dermis, this neoplasm in most of the cases extends deeply and laterally. The lesions may have a warty appearance on their surface, and this may be the cause that they are often confused with warts. Shah et al report a lymphangioma presenting on the penis.[12]

The case report corroborated the literature regarding the presentation, location, and most common clinical findings of the superficial lymphangioma on the abdomen with appearance of "frog spawn." The most relevant diagnoses in such cases were as follows: Herpes zoster, viral warts, angiosarcoma, hemangioma, epidermal nevus, angiokeratoma, and leiomyoma.[2,3,7,8]

Dermoscopy shows a picture of lacunar or saccular pattern, because of the presence of lymphatic fluid varying in color from light brown to violet, a result of the volume of blood wrapped by pale septa, which shows the hypopyon-like, characterized by blood sedimentation.[3,8,9]

Imaging tests should be done to determine the extent of the lesion, the involvement of internal organs, or compression of adjacent structures and also help in planning of the surgical excision.[10]

Magnetic Resonance Imaging (MRI) is the test of choice, in which the diagnosis is confirmed by the evidence of serpiginous-like structures in the subcutaneous tissue.[10]

Histopathological examination is considered golden clue for the diagnosis of CLC[1-3,9] in which vesicles appear as dilated lymphatic vessels in the papillary dermis, with the possibility to extend up to the reticular dermis and subcutaneous tissue. Hyperkeratosis and irregular acanthosis may be present in some cases but not all the cases.[2,7]

The lymphatic lumen is filled with finely granular hyaline material and red blood cells, that may have originated from small capillary shunts.[1]

The correlation of the clinical, dermoscopic, radiological, and histopathological findings of the lymphatic system is helpful for accurate diagnosis.

No medical therapy has been proven effective for lymphangiomas. Lymphangioma does not responsend to radiation therapy or steroids. However, propranolol represents a potential option, which may be of benefit even for intractable diffuse lymphangiomatosis.[13] Sodium tetradecyl sulfate may be used with care for off-label treatment of lymphangioma circumscriptum.[14]

Antibiotics are given for secondary cellulitis. In the case of lymphangioma circumscriptum, severe recurrent cellulitis may happen especially in immunocompromised patients.[14]

The ideal treatment for lymphangiomas is complete surgical excision.[15] In which, there is about 75% of cure and also there is low rates of recurrence.[1,2,10]

Local recurrences are common in lymphangiomas. Adequate excision of lymphangiomas can be difficult and unfeasible. This problem is the main reason for the high rate of recurrence. Tumors that are confined to the superficial dermis are more suitable for surgical excision, which is associated with a high rate of success.

There are other therapeutic modalities that include cryotherapy, sclerotherapy, cautery, CO_2 laser, thermoablation and radiofrequency therapy.[10,16,17] Bleomycin sclerotherapy has been advocated for

head and neck lymphangiomas.[18]

Lymphangioma circumscriptum may also be treated with simple electrodessication. The tumour does not always respond to use of the pulsed-dye laser.[19]

Vaporization with a carbon dioxide laser has been tried with good results.[20] Other lasers may also be used.[21] Intra-oral lymphangioma may be treated using fractional carbon dioxide laser.[22]

Another therapeutic option for treatment of lymphangioma circumscriptum is 23.4% hypertonic saline sclerotherapy.[23]

The use of intralesional OK-432 (Picibanil) is a new and effective treatment for macrocystic lesions,[24] but the response of microcystic or cavernous lesions to OK-432 has been disappointing and surgery remains the most effective treatment for these microcystic and cavernous lesions.[25,26]

Postoperative vacuum-assisted closure devices may be used to decrease the risks of recurrence and infection.[27]

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