Asymptomatic plaque in scalp

Hussein Hassab El-Naby, MD, Sadat Mosbeh, MD

Department of Dermatology, Al-Azhar University, Cairo, Egypt

CLINICAL FINDINGS

A 25 year old male patient presented with asymptomatic mass in scalp for 2 years. The condition started with gradual onset and slowly progressive growth. Sometimes, the patient was complaining of bleeding and oozing from the lesion after minor trauma. There was no history of pain or itching from the lesions. There is no family history of the same condition. On examination, multiple shiny grouped papules forming a plaque having verrucous morphology almost 3 × 2 cm in size were present over the occipital region of the scalp. Pain and tenderness were not observed during examination of the lesion (Fig. 1).

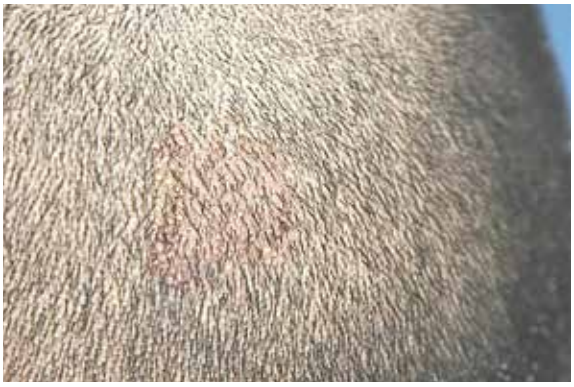

Fig. 1 Showing multiple shiny grouped papules forming a plaque in frontal region of scalp.

What is your clinical differential diagnosis?

Nevus sebaceous
Syringocystadenoma pappilliferum
Syrinogofibroadenoma
Lymphangioma circumscriptum

PATHOLOGICAL FINDINGS

A scalpel biopsy of the lesion was done and histological examination revealed hyperkeratosis, acanthosis and papillomatosis with presence of numerous dilated lymphatics in papillary dermis containing red blood cells and homogenous proteinaceous fluid. Perifollicular and perivascular and perifollicular lymphohistiocytic infiltration admixed with plasma cells and eosinophils. (Fig. 2,3,4)

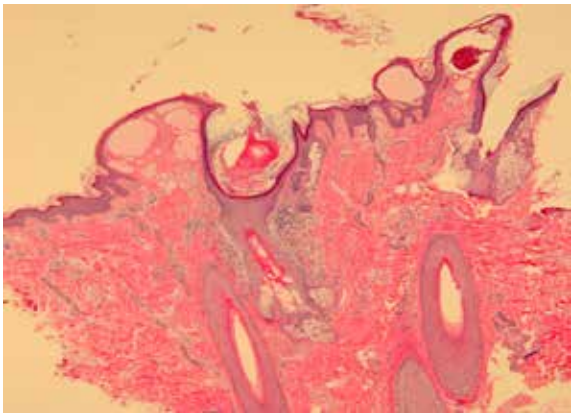

Fig. 2 Hyperkeratosis, acanthosis, papillomatosis and dilated follicle filled with keratin. Dilated lymphatic vascular channels containing lymphatic fluid and RBCS. Perivascular and perifollicular lymphohistiocytic infilterate admixed with plasma cells.

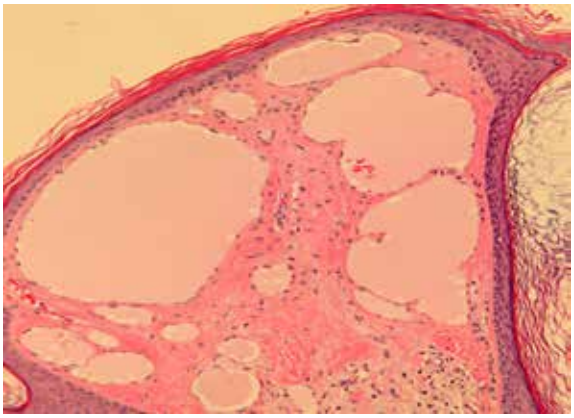

Fig. 3 Numerous lymphatic vessels containing lymphatic fluid in papillary dermis.

Correspondence: *Dr. Hassab El-Naby H, Department of Dermatology, Al-Azhar University, Cairo, Egypt*

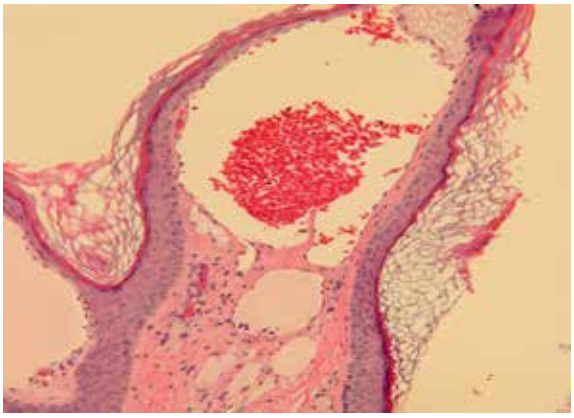

Fig. 4 Papillary dermis containing lymphatic vessels filled with RBCS and lymphatic fliud.

DIAGNOSIS

Lymphangioma circumscriptum

COMMENT

Lymphatic malformations or lymphangiomas are uncommon benign hamartomas composed of dilated lymphatic channels filled with a proteinaceous fluid. They generally do not have connections to the normal lymphatic system and result from maldevelopment of primitive lymphatic sacs.[1] They are commonly seen over chest, mouth, axilla, and tongue. Lymphangioma can occur at any age, but majority are seen in children. Approximately 50% present at birth and 90% are diagnosed before the age of 2 years.[2] Lymphangiomas can be divided into LC, which are superficial cutaneous lesions, and cavernous lymphangioma, which are more deep-seated.[3]

LC, also known as "capillary lymphangioma," "lymphangiectasia," and "dermal lymphangioma," is a rare benign skin disorder involving hamartomatous lymphatic malformation of deep dermal and subcutaneous lymphatic channels.[4]

Peachey et al.[5] classified LC into two main forms: classic and localized. The classic form usually appears at or soon after birth and involves proximal limbs. It is thought to be derived from muscular lymphatic cisterns which failed to form segment during embryonic development. Clinically, lesions appear as vesicular and do not progress into warty plaque. On the contrary, the localized form is seen at any age and has no site predilection.

LC can be classified into congenital and acquired forms. Congenital LC results from local malformation of lymphatics and manifests at birth or before 5 years of age. Whereas, acquired form occurs secondarily due to obstruction of lymphatics commonly in the vulvar region and can manifest at any age secondary to pelvic surgery, radiation therapy, and infection such as tuberculosis, Crohn's disease, and so on.[6] The common sites are axillary folds, shoulder, upper arm, scrotum, penis, rectum, and vulva.

LC can occur as a result of collection of subcutaneous lymph cisterns during embryonic development, which are not connected to the lymphatic system and therefore unable to drain the lymph received from the surrounding tissue. The cisterns are lined with muscle that contracts and, by applying pressure, produces protrusions on the skin. Acquired LC develops due to injury to deep collecting lymphatics, caused by radiotherapy damage or infections such as filariasis, lymphogranuloma venereum, or tuberculosis in advanced age. The exact etiology of LC is unknown, but various growth factors such as vascular endothelial growth factor-C (VEGF-C) and VEGF-D and their receptors on the lymphatic endothelial cells may have a role.[7]

LC is usually asymptomatic. Vesicles that contain lymphatic fluid are compared with frog spawn which are pink to copper-colored while secondary hemorrhage gives it red or black color. Less commonly, it can present as diffuse swelling or with verrucous morphology as was seen in our case.

If there is significant hyperkeratosis, the swelling may clinically resemble warts. It can be complicated by excessive drainage and recurrent cellulitis.

The Clinicopathological challenges of Lymphangioma circumscriptum

Diagnosis	Clinical	PATHOLOGICAL
Nevus sebaceous	• Hairless, mamillated or verrucous lesion since birth or shortly after birth	• Hyperkeratosis,acanthosis and papillomatosis. sebaceous gland attached to hair follicle in upper dermis with apocrine glands beneath it
Syringocystad-enoma Papilliferum	• Also known as papillary syringadenoma • Warty tumor of scalp, neck and face that occurs at any age • Slow growing or a recent change in an apparent birthmark • 1/3 have adjacent nevus sebaceus, 10% have adjacent basal cell carcinoma	• Glandular papillary proliferation connected to skin surface, dense plasma cell infiltrate
Syringofibroad-enoma	• Predilection for extremities, especially acral sites • Verrucous papules, nodules and ulcerative plaques • Gradual and symmetric spread of papular lesions over large areas of body; occasional association with neoplastic or inflammatory conditions	• Thin anastomosing reticulated cords and strands of basaloid monomorphous cuboidal cells extending from the basal layer of epidermis into dermis • Cells are slightly smaller than neighboring keratinocytes • Loose fibrovascular stroma • Cords have scattered ductal structures resembling eccrine ducts

Clinical features of LC may share some skin lesions such as nevus sebaceous, syringocystadenoma papilliferum and syringofibroadenoma of Mascaro.[8] (Table 1) As the clinical presentation of LC may vary from pseudovesicles to nodules or wart-like lesions, it requires histopathological assessment.[9]

The treatment with carbon dioxide laser vaporizes the underlying tissues and seals the lymphatic channels[10] If the laser energy does not penetrate deep enough into the dermis and/or subcutaneous tissue, the patient will only achieve shortterm palliative symptomatic relief and will have lesion recurrence. Furthermore, energy delivered into deeper structures requires large amounts of local anesthesia (injection or tumescent) because of pain during delivery and may be followed by prolonged healing times with the potential for scarring, prolonged erythema, or postinflammatory hyperpigmentation. Pulsed dye laser (PDL) emits high energy laser light in ultrashort pulse durations, allowing for specific targeting of the chromophore hemoglobin (585-595nm) in and around vessels without damaging the surrounding tissues[11] The effectiveness of PDL in the treatment of LC can be limited by the minimal hemoglobin as a chromophore, because the dilated lymphatic channels contain serosanguineous fluid in dilated lymphatic channels.

Sclerotherapy involves injecting detergent sclerosants, chemical irritants, or hyperosmolar agents into the lymphatic malformations to destroy the aberrant vessels. Injectable corticosteroids, tetracycline, 50% dextrose solution, and hyperosmolar saline have all been used in case reports and preliminary trials[12,13] For sclerotherapy, intralesional

injection of 1% sodium tetradecyl sulfate is used with a very good result. Sclerotherapy using sodium tetradecyl sulfate can be considered a successful minimally invasive treatment option for LC. Depending on the study, recurrence rates vary from 58% to 100%. Cryotherapy utilizes very low temperatures to cause immediate vasoconstriction followed by reactive vasodilation, in turn producing cellular necrosis and healing by secondary intention. Recently, imiquimod cream has been used successfully. Imiquimod induces cellular production of endogenous interferons and interleukins[14] Efficacy in these patients is likely related to the ability to inhibit vessel formation and induce endothelial cell apoptosis.

Surgical excision is regarded as the most definitive treatment, giving the highest chance of cure with a recurrence rate of 17% to 23%.[15] By completely excising the subcutaneous cisterns and removing the source of the vesicles, it is possible to eliminate the cutaneous manifestations. Whereas surgical excision offers definitive treatment, it also involves significant risks including scarring, keloid formation, hematoma, wound infection, and nerve injury. Surgery gives the lowest rates of recurrence, but has the highest risk of complications.[16]

REFERENCES

1. Esquivias Gómez JI, Miranda-Romero A, Cuadrado Vallés C, Bajo del Pozo C, Sánchez Sambucety P, Martínez Fernández M, et al. Lymphangioma circumscriptum of the vulva. Cutis 2001; 67:229-32.
2. Morris M. Lymphangioma circumscriptum. In: Unna PG, Morris M, Duhring LA, Leloir H, editors. International Atlas of Rare Skin Diseases. London: Lewis; 1889. p. 1-4.
3. Cahill AM, Nijs ELF. Pediatric vascular malformations: Pathophysiology, diagnosis, and the role of interventional radiology. Cardiovasc Intervent Radiol 2011; 34:691-704
4. Peachey RD, Lim CC, Whimster IW. Lymphangioma of skin. A review of 65 cases. Br J Dermatol 1970; 83:519-27.
5. Mu XC, Tran TA, Dupree M, Carlson JA. Acquired vulvar lymphangioma mimicking genital warts. A case report and review of literature. J Cutan Pathol 1999; 26:150-54.
6. Roy KK, Agarwal R, Agarwal S, Kumar S, Malhotra N, Gopendru N. Recurrent vulvar congenital lymphangioma circumscriptum - A case report and literature review. Int J Gynecol Cancer 2006;16:930.
7. Mehta V, Nayak S, Balachandran C, Monga P, Rao R. Extensive congenital vulvar lymphangioma mimicking genital warts. Indian J Dermatol 2010; 55:121-22.
8. Whimster IW. The pathology of lymphangioma circumscriptum. Br J Dermatol 1976; 94:473-86.
9. Eliezri YD, Sklar JA. Lymphangioma circumscriptum: Review and evaluation of carbon dioxide laser vaporization. J Dermatol Surg Oncol. 1988; 14:357-64.
10. Lai CH, Hanson SG, Mallory SB. Lymphangioma circumscriptum treated with pulsed dye laser. Pediatr Dermatol. 2001; 18:509-10.
11. Bikowski JB, Dumont AM. Lymphangioma circumscriptum: Treatment with hypertonic saline sclerotherapy. J Am Acad Dermatol. 2005; 53:442-44.
12. Niti K, Manish P. Microcystic lymphatic malformation (lymphangioma circumscriptum) treated using a minimally invasive technique of radiofrequency ablation and sclerotherapy. Dermatol Surg. 2010; 36:1711-17.
13. Wang JY, Liu LF, Mao XH. Treatment of lymphangioma circumscriptum with topical imiquimod 5% cream. Dermatol Surg. 2012; 38:1566-69.
14. Browse NL, Whimster I, Stewart G, Helm CW, Wood JJ. Surgical management of lymphangioma circumscriptum. Br J Surg. 1986; 73:585-88.
15. Bond J, Basheer MH, Gordon D. Lymphangioma circumscriptum: Pitfalls and problems in definitive management. Dermatol Surg. 2008; 34:271-75.