



CASE REPORT OF BENIGN VERRUCOUS LESION INFLAMMATORY LINEAR VERRUCOUS EPIDERMAL NEVUS (ILVEN)

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KEYWORDS :

INTRODUCTION

ILVEN; Linear verrucous epidermal nevus; Verrucous epidermal nevus;

An epidermal naevus is due to an overgrowth of the epidermis. Lesions are present at birth (50%) or develop during childhood (mostly in the first year of life). The abnormality arises from a defect in the ectoderm, the outer layer of the embryo that gives rise to epidermis and neural tissue.

An inflammatory linear verrucous epidermal naevus (ILVEN) is a special kind of epidermal naevus. Epidermal naevi are birthmarks due to an overgrowth of the epidermis (upper layers of the skin).

(ILVEN) is a type of skin overgrowth, called epidermal nevus. It is characterized by skin coloured, brown, or reddish, wart-like papules (nevi). The nevi join to form patches or plaques that often follow a pattern on the skin known as the "lines of Blaschko". The affected areas of the skin may be red, itchy, and inflamed. ILVEN typically presents from birth to early childhood and often is limited to one side of the lower half of the body. It affects females more often than males. It usually occurs alone; however, rarely ILVEN can be associated with other symptoms as part of an epidermal nevus syndrome. Rarely, ILVEN can become cancerous (for example, basal cell carcinoma or squamous cell carcinoma).

ILVEN is caused by a genetic change that occurs after conception. How this alteration leads to the signs and symptoms of ILVEN is not well understood. There is no one treatment or cure for ILVEN. Products used to soften the skin (emollient), moisturizers, and topical steroids may be utilized to relieve symptoms of dryness and itching. Recently, several patients have been treated with laser therapy with good results.

CASE REPORT

A 9 Year hindu male child patient came with his parents. Coming from lower socio- economical state, studying in 4th standard. Presented with chief complain of black colour pigmentation growth over the umbilicus since birth.

Patient was relatively asymptomatic at birth than 5 days after his birth, his parents noticed a black colour pigmented growth over the umbilicus. Then his parents took him to a hospital. Then clinicians told his parents the growth was normal. Nothing to worry about it. It will subside, when grows up but when the child grows up, this black colour pigmented growth increases upto the present stat. On general examination, patients was vitally stable. No icterus/pallor/ cynosis/

clubbing/ oedema/ lymphadenopathy/ bilateral air entry was present. No rales/ rhonchi/ crepitation.

Per abdomen soft non tender, no rigidity, no guarding, no rebound tenderness.

Local examination on inspection a multiple discrete brown papules linear pigmented growth present over the umbilicus. They fuse to each other. The surface of the lesion dry and hilly. No redness swelling in duration present surrounding the pigmented growth.

On palpation multiple discrete pigmented papule was non warm, non tender. It was dry, rough margin, fixed to the Superfascial layer of the skin, no impulse on coughing, transillumination was negative.

PHOTOGRAPH



9 year male child



9 year male child

INVESTIGATION

Complete Haemogram- Normal
RFT & LFT - Normal
PT with INR - Normal
HIV/HBSAG were Non reactive

Chest X-ray/ PA View- on basis of history and clinical examination and investigation, decision was taken excision under general anaesthesia and specimen sent to HPE for

further evaluation. The patient was treated by excision under general anaesthesia.

Biopsy was On gross examination greyish, brown, soft tissue structure measuring – 7.5cm* 0.8cm* 0.5 cm.

On microscopic examination, it shows Benign verrucous lesion-verrucous vulgaris. No evidence of malignancy is seen.

POTHOS



Intra-OP

Post-OP

After Discharge

Post Operative Status.

Post operative recovery was uneventful.

DISCUSSION

Like other linear epidermal Naevi, ILVEN is characterized by warty lesions that tend to group together in a linear pattern. The difference is that the lesions are red, inflamed and itchy, sometimes intensely so. The surface of the lesions may look like eczema (dry, red, scratched) or like psoriasis (red and scaly).

The abnormality resulting in ILVEN arises from a defect in the ectoderm. This is the outer layer of the embryo that gives rise to epidermis and neural tissue. The defect causing the skin lesions may also result in disorders of other internal organs such as the brain, eyes and skeleton. This is extremely rare with ILVEN.

ILVEN is diagnosed by careful history and physical examination. A skin biopsy can be helpful and may show the characteristic histological features of ILVEN.

ILVEN is a condition that normally only affects one side of the body (unilateral). Usually the left side of patients is affected. The condition is persistent and forms along characteristic lines. It usually appears on an extremity in infancy or childhood. Altman and Mehregan described six characteristic features of ILVEN: (1) early age of onset, (2) predominance in females (4:1 female-male ratio), (3) frequent involvement of the left leg, (4) pruritus, or "itchiness" (5) marked refractoriness to therapy, and (6) a distinctive psoriasiform and inflammatory histologic appearance.

- The plaques are characterized histologically by hyperkeratosis which is a thickening of the outer layer of skin. Hyperkeratosis is often associated with an abnormal amount of keratin production. Also characteristic is moderate acanthosis a thickening of the stratum spinosum with elongation of rete ridges.
- Characteristic histologic feature is regular alternation of slightly raised parakeratotic areas without a granular layer (hypogranulosis) and slightly depressed orthokeratotic areas with prominent granular layer (hypergranulosis). Orthokeratotic hyperkeratosis is characterised by hyperkeratosis with non-nucleated cells. Parakeratotic hyperkeratosis is characterised by hyperkeratosis with nucleated cells.
- The orthokeratotic area shows a basket-weave-pattern.
- The dermis shows scattering of chronic inflammatory infiltrate (Munro's microabscess) sometimes giving a spongi-

form appearance.

Reported treatments include topical agents, dermabrasion, cryotherapy, laser therapy, and surgical excision. These therapies have a high failure rate because of incomplete relief of symptoms, scarring, or recurrence.

Though similar in appearance, ILVEN will not respond to therapies known to affect psoriasis. ILVEN can be very difficult to live with but can be treated. The most effective method is full-thickness excision of the lesion. CO₂ Laser Surgery can resurface the skin to give a flat, smoother and more normal appearance, but does not remove the lesion.

There is no real effective medical treatment for epidermal naevi. Topical calcipotriol may reduce the thickness of the skin in some cases. If necessary, laser or surgical removal of naevi may be performed

CONCLUSION

A 9 years male child patient came with his parents with chief complaint of black colour pigmentation growth over the umbilicus since birth. ILVEN is diagnosed by careful history and physical examination. A skin biopsy can be helpful and may show the characteristic histological feature of ILVEN. ILVEN most often affects one leg and may extend from buttocks to the foot. There is no real effective medical treatment for epidermal naevi. Topical calcipotriol may reduce the thickness of the skin in some cases. If necessary, laser or surgical removal of naevi may be performed.

Conflicts of Interest

All authors state no conflicts of interest.

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