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Original Research Paper

Pathology

EXTENSIVE ECCRINE ANGIOMATOUS HAMARTOMA SUCCESSFULLY TREATED IN A 7 YEARS GIRL CHILD.

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ABSTRACT Eccrine angiomatous hamartoma (EAH) is a rare, benign cutaneous hamartomatous lesion characterized by the proliferation of the eccrine glands and thin-walled vascular channels in the middle and deep dermis along with other dermal elements, such as adipose tissue, hair, and nerve bundles. Most of the lesions are reported in the extremities, mostly the lower limb. We report an extensive case of eccrine angiomatous hamartoma in a seven years old female child presented with multiple, hyperpigmented nodules over the right thigh lateral aspect and leg present since birth. The lesions are gradually increasing and associated with pain, hypertrichosis, and right knee joint flexion deformity. Complete excision of the lesion in the leg and thigh was performed in two stages and raw areas covered with thick split skin grafts harvested from the left thigh.

KEYWORDS : Cutaneous Hamartomas, Angiomatous lesion, Hypertrichosis, Hyperhidrosis.

A 7 years old female child presented with hyperpigmented nodules over the lateral aspect of the right thigh and leg since birth which is gradually increasing in size. She had pain, flexion deformity of the right knee joint resulting in limb length discrepancy. She also had hyperhidrosis, sebaceous secretions, pigmentation, hypertrichosis, firm nodularity, or pigmented plaque. Lesions are not associated with itching and no lesions were observed at other sites. No significant past and family history. On local examination, multiple hyperpigmented patches over the lateral aspect of the right thigh near the knee joint and leg are seen [Figure 1A, 1B]. Systemic examination reveals bilateral symmetrical vitiligo of the labia [Figure 1C].

The radiograph shows multiple opacified lesions in the dermis and subcutaneous area over the lateral aspect of the right thigh and leg [Figure 2A, 2B]. Ultrasound scanning revealed multiple prominent vascular channels noted along the subcutaneous plane from mid-thigh to proximal leg predominantly showing venous flow pattern with a few of them showing arterial flow wave pattern. Few of the vessels are thrombosed and the impression was multiple arteriovenous malformations [Figure 2C]. All the lesions are confined only to the skin and subcutaneous tissue. A 4mm skin biopsy was taken and it shows numerous thin-walled dilated and collapsed fine venules throughout the dermis which are in close association with prominent hyperplastic eccrine units. Features are consistent with eccrine angiomatous hamartoma.

While most of the reported cases are solitary or very few, in our case the total nodules or plaques are 42 in number in the right leg and thigh. They are all situated in the skin and subcutaneous tissues. Even though the child is having the lesion since birth and the parents took the child to different doctors including tertiary care, no proper advice was given; it only shows the rarity of the lesion.

We operated on the patient in two stages. Initially, the lesion over the lateral aspect of the right leg was excised and the raw area is covered by split skin grafting in 2019 [Figure 3- A, B, and C]. The flexion deformity of the knee was corrected. Because of Covid -19 problems, we could not perform the 2nd surgery early and we operated in 2021. For the lesions over the lower part of the right thigh excision and split skin grafting was done and splinting of the right knee was done to prevent flexion deformity [Figure 3a- A, B, C, and D].

The excised specimen shows multiple hyperpigmented nodules over the skin surface and grey-brown cut surface [Figure 4B]. Microscopy shows unremarkable epidermis, dermis shows hyperplastic eccrine glands with surrounding mucinous stroma, hair follicles, many thin-walled congested and dilated venous channels, thick-walled muscular arteries, lymphatic channels, lobules of mature adipose tissue, and nerve bundles [Figure 4-A, C, and D].

After two successful surgeries, the child was relieved of her suffering. The wound healed well and the graft was healthy [Figure 5B]. Flexion deformity of the knee joint was reduced comparatively [Figure 5A and 5B]. An extensive review of the literature could not show a parallel case and we are reporting this case because of its extreme rarity and procedure adopted for successful management. There is also associated bilateral symmetrical vitiligo of the labia.

DISCUSSION

The term EAH was coined by Hyman et al. in 1968, but actually, it was first described by Lotzbeck in 1859 as an angiomatous-appearing lesion on the cheek of a child [1]. In 1960s, Vilanova et al. described as sweating angiomatous hamartoma and Domonkos & Suarez as sudoriparous angioma, for similar lesions with hyperhidrosis [2].

EAH is a rare, benign cutaneous hamartomatous lesion characterized by the proliferation of both eccrine glands and thin-walled vascular channels in the middle and deep dermis [1]. EAH may occur as a result of abnormal interaction between the differentiated epithelium and the mesenchyme due to biochemical alterations, leading to abnormal proliferation of adnexal structures and vascular elements in congenital cases [3], [4]. EAH is primarily a disease of the young, usually appearing at birth or arising during childhood [1]. Similar findings are seen in our case as the disease was present since birth. Few cases of adult-onset [5], familial [2] and, late-onset of EAH were reported in the literature [6, 7, 8, and 9]. It shows no gender or racial preponderance.

EAH commonly presents as a solitary bluish/brown nodule or papule/plaque; multiple lesions are uncommon which are seen in our case with multiple hyperpigmented nodules associated with pain on the lateral aspect of the right thigh

and leg causing limb length discrepancy. Sometimes it can mimic a capillary haemangioma in appearance [10]. The predilections sites are the distal extremities mostly palms and soles followed by the trunk and rarely face [6]. Some of the lesions may be exclusively accompanied by hyperhidrosis or hypertrichosis which is noted in our case [5]. It was postulated that infiltration of small nerves may be responsible for the pain, as seen in our case, and a local increase in the temperature within the angioma may produce sweating, which is seen in the eccrine component of the hamartoma in cases with hyperhidrosis [1].

Histologically, Pelle et al [11] proposed the diagnostic criteria for EAH which include hyperplasia of normal or dilated eccrine glands, the close association of the eccrine structures with capillary angiomatous foci, and variable presence of pilar, lipomata's, mucinous, and/or lymphatic structures. The histological variants of EAH include follicular, lipomata's and mucinous and anecdotal case of EAH with verrucous angioma was reported [3].

Immunohistochemical staining in EAH delineates normal eccrine structures and the vascular elements. The vascular elements are identified by using Factor VIII-related antigen and anti-Ulex europaeus-1. The secretory portions of the eccrine glands are identified by S100, CAM 2.5, anti-EMA, and anti-CEA, whereas the ductal portions are stained with anti-CEA, cytokeratin, and anti-EMA [12]. Lymphatic vessels are identified using D2-40 staining [11]. The main clinical differential diagnoses for EAH include vascular malformations, tufted angioma, smooth muscle hamartoma, glomus tumor, eccrine nevus, blue rubber bleb nevus, and macular telangiectatic mastocytosis [12]. Doppler ultrasonography may help in confirming the clinical suspicion of an angiomatous lesion, but accurate diagnosis of EAH remains with histology [1].

EAH is a benign hamartomatous lesion without spontaneous regression or malignant transformation. The lesion may enlarge in proportion with the patient's growth. The definite therapeutic management is surgical excision and repair of the defect by split skin grafting. In case of refusal for surgical management, Botulinum toxin injection gives excellent results for lesions accompanied by hyperhidrosis [13]. Hypertrichosis can be treated by Laser depilation [3]. Few cases of EAH are successfully treated with pulsed dual-wavelength sequential 595- and 1,064-nm laser [14]. A case of late-onset extensive eccrine angiomatous hamartoma was treated with four injections of intralesional sclerosant (Aethoxysclerol) and it regressed successfully [15].

CONCLUSION

We are presenting this unique case of an extensive eccrine angiomatous hamartoma in a 7 years old girl affecting the right lower limb which is present since birth. Because of the extensive nature of the lesion and associated pain and tendency for flexion deformity of the right knee, we have to excise the lesion and since the defect is large we performed split skin grafting. It is pertinent to note that even though multiple lesions are present in the right lower limb, no other lesions were seen elsewhere in the body.

Legends For Photographs

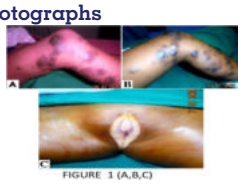

Figure 1A, 1B - Preoperative photograph of right lower limb showing flexion deformity of knee and multiple hyperpigmented patches over the lateral aspect of right thigh and leg.

Figure 1C - Showing bilateral symmetrical vitiligo of the vulva.

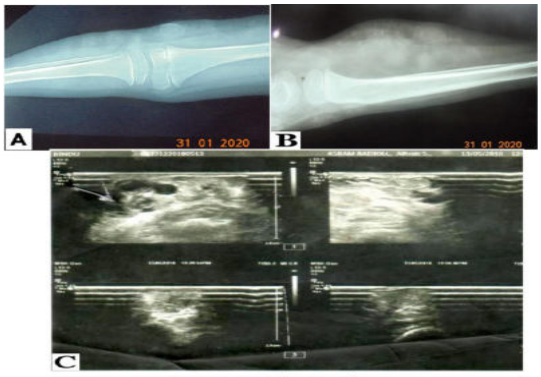

FIGURE 2(A,B,C)

Figure 2a, 2b - Radiograph Of Right Lower Extremity And Knee Anteroposterior And Lateral View Showing Multiple Opacified Lesions In The Dermis And Subcutaneous Area Over The Lateral Aspect Of Thigh And Leg.
Figure 2c - Ultrasound Reveals Multiple Prominent Vascular Channels Noted Along A Subcutaneous Plane From Mid-thigh To Proximal Leg Predominantly Showing Venous Flow Pattern And Few Of Them Showing Arterial Flow Wave Pattern.

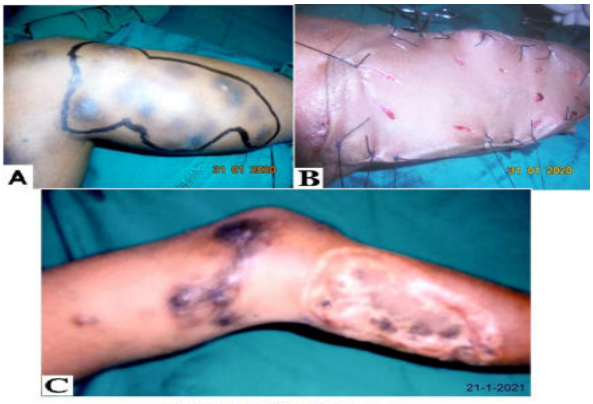

FIGURE 3(A,B,C)

Figure 3a – First Stage Of Operation For The Leg Lesion, Up To Marked Area Was Completely Excised.
Figure 3b – Split Skin Grafting Of The Raw Area Was Performed.
Figure 3c – Follow-up Photograph 3 Weeks After The First Surgery.

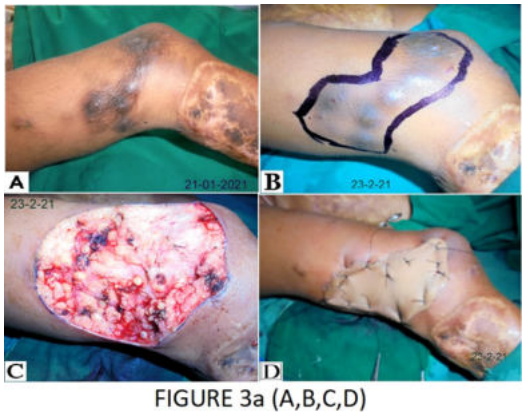

FIGURE 3a (A,B,C,D)

Figure 3a – A-Second stage of operation, preoperative photograph.
Figure 3a – B- Extent of excision.
Figure 3a – C- Defect after complete excision.
Figure 3a – D- Split skin grafting of raw area and correction of flexion knee deformity.

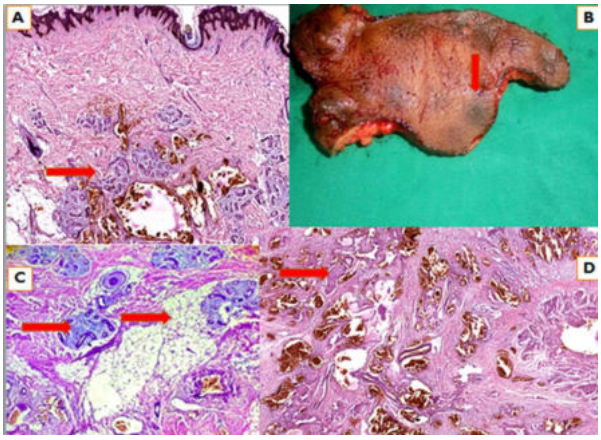

Figure 4- A- Microphotograph showing the unremarkable epidermis, hyperplastic eccrine glands and terminal hair follicles in the dermis, abnormally dilated blood vessels in the deep dermis, and subcutaneous tissue (H&EX100).
Figure 4- B-gross Photograph Of The Lesion Excised On The Lateral Aspect Of The Right Thigh.
Figure 4- C- Microphotograph Showing Increased Dermal Mucin And Fatty Infiltration (h&ex100).
Figure 4- D- Microphotograph Showing Many Abnormally Dilated And Congested Blood Vessels, Lymphatic Channels, Thick-walled Muscular Artery, And Entrapped Nerve Bundles In The Deep Dermis (h&ex100).

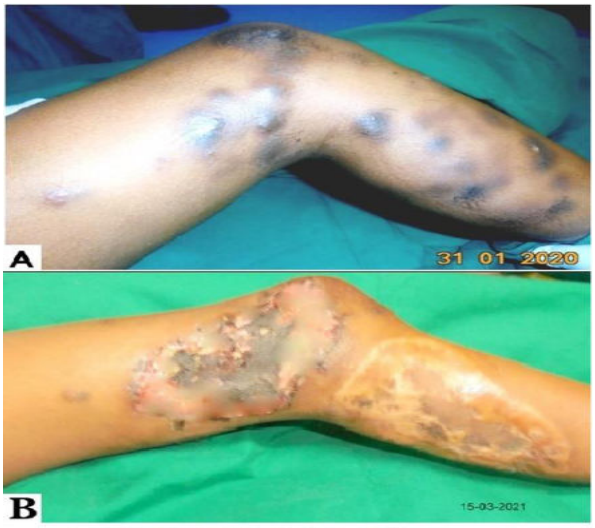

FIGURE 5 (A,B)

Figure 5a- Preoperative Photograph With Flexion Deformity.
Figure 5b- Postoperative Photograph After Complete Excision Of Both The Lesions And Correction Of Flexion Deformity Of The Knee.

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