

Eccrine Angiomatous Hamartoma Masquerading as Hemangioma in a Neonate

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ABSTRACT

Introduction: Eccrine Angiomatous Hamartoma (EAH) is a rare benign tumor-like lesion seen in infancy and childhood and it involves mainly the extremities. It is very rare in neonatal age and more so on the scalp where the hemangiomas are more common.

Case Report: We are reporting a rare case of EAH in a female neonate who presented with a single lesion on the scalp. Physical examination revealed a reddish soft tissue mass of 5x4 cm size on the scalp. Ultrasonography and C T Scan suggested it to be a vascular lesion. It was excised and histopathological examination revealed proliferation of eccrine glands and thin walled blood vessels making a diagnosis of EAH.

Conclusion: Eccrine Angiomatous Hamartoma (EAH) can mimic a capillary haemangioma and surgical excision followed by histopathological examination is only the final word in making the diagnosis of this condition.

Keywords: Eccrine Angiomatous Hamartoma, Hemangioma, Scalp

T scan revealed a well defined soft tissue density mass arising in the scalp region adjacent to coronal suture on left side with underlying bony defect in left parietal bone and without any intracranial extension. After the hematological investigation the patient was operated under general anaesthesia and excision biopsy was done. The histopathology examination found it to be a case of Eccrine Angiomatous Hamartoma (Figure-2). The baby was discharged on 5th post operative day in good condition.

DISCUSSION

Eccrine Angiomatous Hamartoma (EAH) is a rare benign cutaneous hamartoma, which is composed of proliferating eccrine secretory coils and ducts intimately associated with capillary angiomatous channels and sometimes also other minor elements, such as fatty lobules and hair. It was first described by Lotzbeck in 1859 as an angiomatous appearing lesion on the cheek of a child and the present term EAH was coined by Hymann and co-workers in 1968.⁴

Eccrine Angiomatous Hamartoma usually occurs as a solitary lesion, but cases with multiple lesions have been described.⁵ The exact pathogenesis is not known but various theories like abnormal induction of heterotypic dependency with resultant malformation of adnexal as well as mesenchymal elements have been proposed. Clinically, EAH presents as an angiomatous lesion, usually solitary, although several cases with multiple lesions have been described.⁶ It is generally asymptomatic, occasionally presents with pain and focal hyperhidrosis in older children. The EAH lesions are commonly seen on the palm and sole, but may also be present at other parts of the feet and even on the face, neck and on the trunk.¹ In the present case the lesion was in the parietal area of the scalp (Figure-1).

On histopathological examination, EAH is an unencapsulated lesion with dermal proliferation of mature appearing eccrine secretory and ductal structures which are intimately associated with thin-walled angiomatous channels, typically of a capillary nature but of variable size.⁷ This lesion has a lobular appearance but ill-defined lesions have also been described on rare situations. Moreover it has also been associated with a number of other histological elements like adipose tissue, epidermis, mucin, apocrine glands, neural elements, and neurovascular glomic bodies. Immunohistochemical studies on EAH have

INTRODUCTION

Eccrine Angiomatous Hamartoma (EAH) is a rare, benign cutaneous tumor usually present at birth or during early infancy and childhood whereas capillary hemangioma (CH) is the most common tumor occurring of skin in this age group.^{1,2} EAH typically occurs as a flesh or reducible coloured solitary nodule or plaque on the extremities but it is rare on the scalp. It grows slowly and becomes symptomatic whereas CH typically regresses over time. Patients of EAH usually present with a solitary, congenital nodule or a plaque that may be painful.³ In a rare clinical situation like the present case it becomes very difficult to differentiate between EAH and CH and then histological evaluation becomes necessary because of the diverse clinical behavior of the these two skin lesions. As a result excision biopsy of the lesion is essential for a definitive line of management. The present case is being reported to stress the need for surgical excision biopsy of this type of rare skin lesion when there is diagnostic dilemma especially when the lesion is on the scalp as in the present case where EAH is rare but the CH is commonly seen.

CASE REPORT

A five days old female baby was admitted in paediatric surgery ward of PGIMS Rohtak with complain of swelling on the scalp since birth. The baby was born by a full term normal vaginal delivery in a district hospital and was referred for the above mentioned complaint. On examination there was a 5x4 cms reddish swelling on the parietal region of the scalp and there was no other associated anomaly (Figure-1). Ultrasonography of the swelling revealed a heterogenous mass of size 4.5x 3.4 cm showing flow on colour Doppler at vertex appearing to arise from scalp and a possibility of hemangioma was kept. C

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Figure-1: The clinical photograph showing the soft tissue lesion (approx 5x4cm) on the scalp of the neonate.

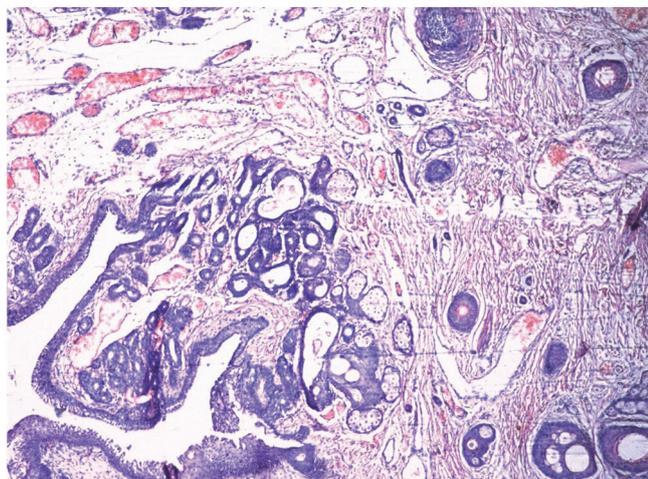


Figure-2: Microphotograph revealing skin covered tissue showing proliferation of eccrine glands and thin walled blood vessels (H and E 40X).

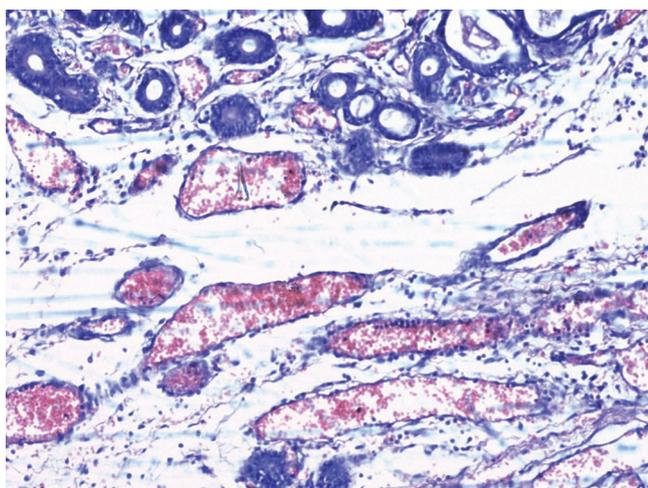


Figure-3: Microphotograph showing proliferation of eccrine glands and thin walled blood vessels (H and E 100X).

been done demonstrating the presence of antigens commonly found in eccrine glands, such as carcinoembryonic antigen (CEA) and S-100.⁸ In the present case the histopathology showed proliferation of eccrine glands and thin walled blood vessels deep to the epidermis (Figure-2 and Figure-3) but immunohistochemical examination was not done.

The natural history of EAH is benign and there have been no reported complications.⁹ But when the presentation is like a hemangioma as the present case, simple excision is usually necessary followed by the histopathological examination as it can only be the final word to differentiate between the two skin lesions.

CONCLUSION

Eccrine Angiomatous Hamartoma (EAH) is a rare benign lesion and it can mimic a capillary hemangioma (CH) in appearance. The surgical excision is the ideal line of management in a situation where it is necessary to confirm the diagnosis which can only be done after histopathological examination.

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