Case Report

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Surgery Section

Epitheloid Haemangioma of the Forehead: A Case Report

SURYA RAO RAO VENKATA MAHIPATHY, ALAGAR RAJA DURAIRAJ, VOLGA HARIKRISHNAN

ABSTRACT

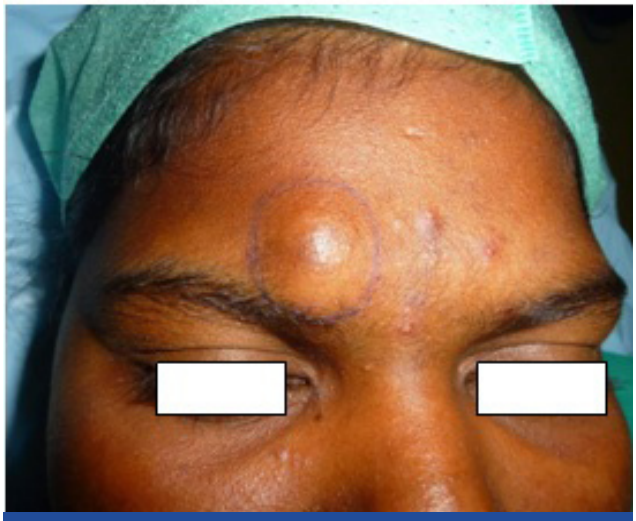
Epitheloid haemangioma is a tumour of soft tissues arising from the vascular endothelium. Here, we present a young female with a swelling of the forehead region mimicking clinically as a sebaceous cyst. Histopathological examination confirmed as epitheloid haemangioma. She was followed up for two years during which time there was no recurrence. This case is being reported as the cutaneous form of this lesion is very rare.

Keywords: Benign, haemangioma, Tumour, Vascular

CASE REPORT

A 22-year-old female presented to the Department of Plastic Surgery, at Saveetha Medical College Hospital, Chennai with a swelling just above right eyebrow for eight months duration. It was spontaneous in onset and gradually progressive. There was no pain, fever or discharge. There was no history of trauma, no history of any comorbid conditions.

On examination, there was a 2 x 1 cm hemispherical swelling forehead just above the medial end of right eyebrow, which was firm, non tender, with well defined margins, not adherent to underlying structures and was adherent to skin at the summit (punctum) [Table/Fig-1]. A diagnosis of a probable sebaceous cyst was made. The plan was excision biopsy & primary closure. A 2 cm incision was made in the eyebrow hairline along the direction of the hair follicles and deepened in layers. A cystic swelling was present, which was excised in toto and sent for histopathological examination. Haemostasis was secured and the incision was closed in layers. Histopathology revealed epitheloid haemangioma [Table/Fig-2,3]. Immediate post-operative picture is shown in [Table/Fig-4].

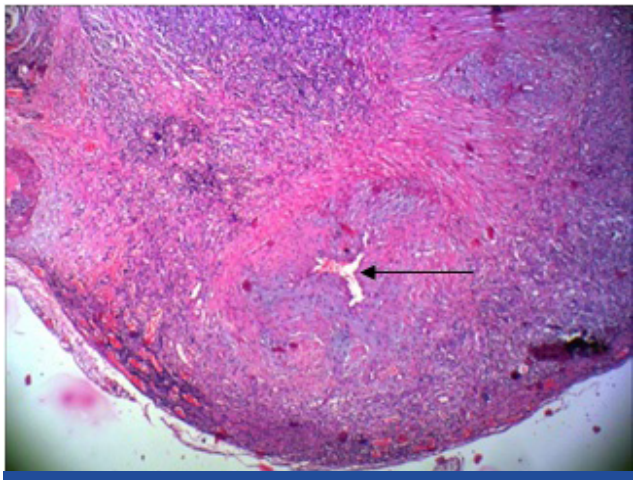


[Table/Fig-1]: Pre-operative photograph showing the lesion on the forehead

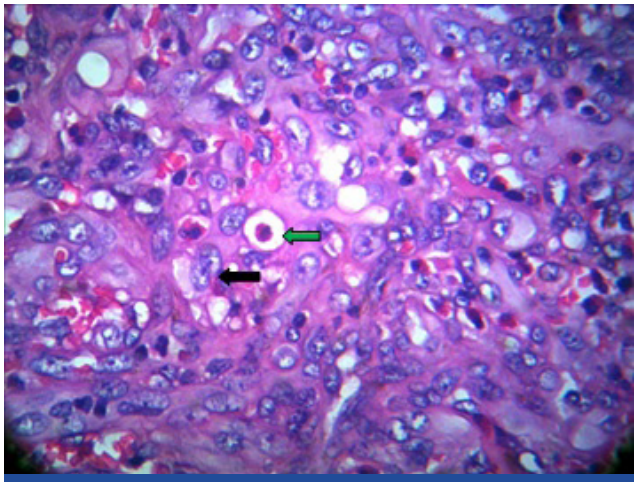
DISCUSSION

Epitheloid haemangioma was first described in 1969 [1]. It is also known as Angiolymphoid Hyperplasia with Eosinophilia (ALHE) or benign angiomatous subcutaneous proliferation. The lesions are usually nodular or papular in the subcutaneous regions of the head and neck of young females [2] generally in the third to fifth decade. It can occur in all races. It usually presents with pruritis and regional lymphadenopathy. There may be eosinophilia in the peripheral smear. It has been associated with trauma, pregnancy, HIV infection and oral contraceptives. Recurrence is uncommon, especially in pregnancy, and no local or distant metastasis has been reported.

The term ALHE is generally used synonymously with Kimura's disease as the features are similar, being common in males with affliction for the regions of head and dermal infiltration by lymphocytes and eosinophils [3] ,but Rosai et al., suggested both to be different clinico pathological entities [4]. In Kimura's disease, lesions present as subcutaneous nodules whereas in ALHE, the lesions are superficial, papulo-nodular with erythema, and are friable. Kimura's disease presents with



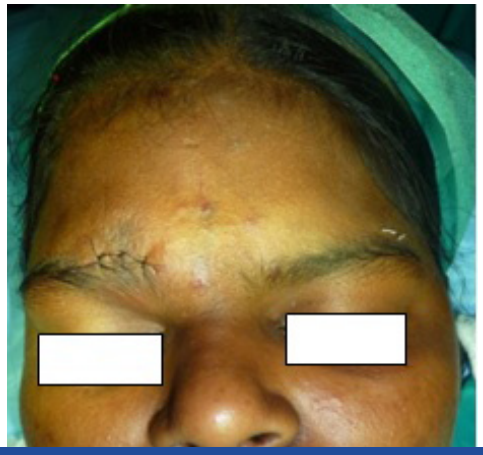
[Table/Fig-2]: The microscopic examination showed a well circumscribed lesion composed of blood vessels predominantly of capillaries (black arrow) admixed with few thick walled blood vessels.



[Table/Fig-3]: They are lined by plump endothelial cells some of them were projecting into the lumen and simulate tombstone (black arrow) while others show single intracytoplasmic vacuole (green arrow). The stroma is densely infiltrated by eosinophils admixed with few lymphocytes and plasma cells

regional lymph node enlargement which is absent in ALHE. On microscopic examination, Kimura's disease features with lymphoid follicles with eosinophilic infiltration whereas capillary proliferations with epitheloid changes are found in ALHE. The other features of Kimura's disease is the presence of peripheral eosinophilia and increased IgE level which is present in only in less than 10% of cases in ALHE [5, 6].

The pathogenesis of ALHE is unclear and there is confusion whether it is a benign vascular lesion or a reaction to trauma [7] that usually occurs in the skin and subcutaneous tissue. It generally presents in adults and it is an indolent lesion, with an average time between appearance and excision of four to five months [8]. Other areas where they are found are bone, liver, pleura, heart and soft tissues of thigh and pelvic cavity.



[Table/Fig-4]: Immediate post-operative picture

On microscopy, the lesions are encapsulated and composed of vessels lined by large endothelial cells protruding into the lumen in a "tombstone fashion" [9], with inflammatory infiltrate around the vessels and eosinophilia clinching the diagnosis of epitheloid haemangioma. The endothelial cells have an epitheloid appearance with large nuclei, nucleoli and plenty of eosinophilic cytoplasm, showing a low mitotic index and absent nuclear atypia [9].

Two varieties of epitheloid haemangioma are present, the typical and the atypical (exuberant) form. The typical form shows mature capillary vessels with a well defined smooth muscle coat. The atypical (exuberant) form shows clumps of endothelial cells having an epitheloid appearance with an indefinite growth pattern and immature vessels [2].

Immunohistochemistry shows positive markers for CD31, CD34 and factor VIII-related antigens and negative for keratin and epithelial membrane antigens [10]. Treatment consists of wide and complete excision of the tumour.

CONCLUSION

Epitheloid haemangioma are rare tumours of the vascular endothelium. They pose a difficulty in diagnosis. Careful examination and discussion with pathologist is of utmost importance. This case is being published for its unusual presentation in the cutaneous form and its rarity.

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AUTHOR(S):
1. Dr. Surya Rao Rao Venkata Mahipathy
2. Dr. Alagar Raja Durairaj
3. Dr. Volga Harikrishnan

PARTICULARS OF CONTRIBUTORS:
1. Associate Professor, Department of Plastic Surgery, Saveetha Medical College Hospital, Thandalam, Kanchipuram, India.
2. Associate Professor, Department of Plastic Surgery, Saveetha Medical College Hospital, Thandalam, Kanchipuram, India.
3. Assistant Professor, Department of Pathology, Saveetha Medical College Hospital, Thandalam, Kanchipuram, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Surya Rao Rao Venkata Mahipathy
Associate Professor, Department of Plastic Surgery,
Saveetha Medical College Hospital, Thandalam,
Kanchipuram-602105, India.
E-mail: surya_3@hotmail.com

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