Case Report

Congenital Solitary Angiokeratoma of Tongue: A Rare Case Report
Manupriya Nain, Shilpi Agarwal, Gautam bir Singh, Raghuram Devenga

Abstract
Angiokeratoma is a rare cutaneous vascular disorder that can occur in several clinically distinct conditions. Oral mucosal angiokeratomas are rare and commonly seen as a component of angiokeratoma corporis diffusum. An 11 year old female child presented with swelling on dorsal surface of tongue since birth. Swelling was erythematous, sessile, non tender. There were no other lesions on oral mucosa or anywhere on skin. Surgical excision was done and on histopathological analysis a diagnosis of angiokeratoma was made. Isolated oral Angiokeratomas are rare. Previous literature search revealed only a single case of Angiokeratoma of tongue with congenital etiology. We are describing a case of angiokeratoma of tongue which is solitary and congenital.

Keywords: Angiokeratoma; Congenital; Tongue; Oral; Mucosal; Solitary; CD34.

Introduction
Angiokeratomas are capillary vascular malformations characterized clinically by asymptomatic solitary or multiple keratotic papules or plaques and histologically by epidermal acanthosis, hyperkeratosis and vascular ectasia of papillary dermis. On the basis of historic aspects, clinical appearance and distribution they have been divided into localized and systemic forms. The localized angiokeratoma include, bilateral form occurring on dorsa of fingers and toes (Angiokeratoma of Mibelli), localized scrotal form (Angiokeratoma of Fordyce), multiple papular and plaque like (Angiokeratoma circumscriptum), solitary papular angiokeratoma and localized vulval form (Angiokeratoma of vulva).5-8 Angiokeratoma corporis diffusum belongs to widespread variety. The generalized systemic form is usually associated with a metabolic disorder, the most common being Fabry’s disease and fucosidosis.6-8

As all varieties of angiokeratoma have similar histologic features, diagnosis is usually made on basis of clinical presentation. Isolated oral mucosal angiokeratomas are rare and only few case reports have been reported in literature.5 Oral mucosal Angiokeratomas are most commonly seen as a component of angiokeratoma corporis diffusum. They may be associated uncommonly with angiokeratomas of scrotum, gastrointestinal mucosa or both.7,8 However, isolated oral mucosal angiokeratomas has not been placed anywhere in the classification of angiokeratoma. In this report we describe a congenital solitary angiokeratoma of tongue in a child. Only one case of solitary Angiokeratoma with congenital etiology has been found on extensive literature search.9

Case Report
An eleven year old female child presented with swelling on dorsal surface of tongue since birth. The swelling was associated with burning sensation. On examination of oral cavity, the patient was found to have 3 x 2 cm raised lesion on midline of tongue extending to right side. The mass was erythematous, sessile, non-tender and do not bleed on touch (Figure 1a). There were no other lesions on oral mucosa or anywhere on skin. On medical examination, no metabolic abnormality was detected. Complete blood count, Liver function tests, renal function tests and Urine examination was within normal limits. Lesion was excised under local anesthesia. Histopathological findings included stratified squamous epithelium showing acanthosis, parakeratosis and papillomatosis. Underlying papillary dermis showed large dilated vascular spaces lined by normal appearing endothelial cells, filled with blood and surrounded by elongated rete ridges. On Immunohistochemistry these endothelial cells were positive for CD34. Based on these findings, a histopathological diagnosis of angiokeratoma was made (Fig 1b).
Discussion
Mibelli reported the first case of Angiokeratoma more than a century ago in 1889.\(^1\) Isolated oral angiokeratomas appear to be a rare and relatively new subset, described for the first time in 1997 by Leung and Jordan.\(^10\) To date, only ten case reports (Table 1)\(^ {9,17} \) mostly of solitary lesions and a single case with congenital etiology have been reported.\(^9\)

Figure 1: Erythematous lesion on dorsal surface of tongue (a) and histopathology showing dilated blood vessels in papillary dermis, surrounded by elongated rete ridges (b). On Immunohistochemistry these endothelial cells were positive for CD34 (c, d & e).

<table>
<thead>
<tr>
<th>Reference</th>
<th>Age/Sex</th>
<th>Sites involved</th>
<th>Solitary / Multiple</th>
<th>Nomenclature as per proposed classification</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leung et al.(^{10})</td>
<td>82 / Male</td>
<td>Buccal mucosa</td>
<td>Solitary</td>
<td>Type 1A(^5)</td>
</tr>
<tr>
<td>Kumar et al. (^{11})</td>
<td>-</td>
<td>Tongue</td>
<td>Solitary</td>
<td>Type 1A(^5)</td>
</tr>
<tr>
<td>Bhargava et al.(^{12})</td>
<td>5 / Male</td>
<td>Tongue (dorsum and lateral)</td>
<td>Multiple</td>
<td>Type 1A(^m)</td>
</tr>
<tr>
<td>Vijaykumar et al.(^{13})</td>
<td>12 / Male</td>
<td>Tongue (ventral, lateral, dorsum)</td>
<td>Multiple</td>
<td>Type 1A(^m)</td>
</tr>
<tr>
<td>Varshney (^{14})</td>
<td>12 / Female</td>
<td>Tongue (dorsum)</td>
<td>Solitary</td>
<td>Type 1A(^5)</td>
</tr>
<tr>
<td>Siponene et al(^{15})</td>
<td>54 / Female</td>
<td>Tongue</td>
<td>Solitary</td>
<td>Type 1A(^5)</td>
</tr>
<tr>
<td>Yildirim et al.(^9)</td>
<td>9 / Female</td>
<td>Tongue (dorsum)</td>
<td>Solitary</td>
<td>Type 1A(^5)</td>
</tr>
<tr>
<td>Ergun et al.(^{16})</td>
<td>16 / Female</td>
<td>Tongue</td>
<td>Solitary</td>
<td>Type 1A(^5)</td>
</tr>
<tr>
<td>Sion-Vardy et al.(^{17})</td>
<td>45 / Female</td>
<td>Tongue (lateral)</td>
<td>Solitary</td>
<td>Type 1A(^5)</td>
</tr>
</tbody>
</table>

Table 1: List of isolated oral angiokeratoma reported till date.
Tongue appears to be most common site for isolated oral angiokeratomas. Interestingly isolated oral mucosal angiokeratomas doesn’t fit in classification and new classification systems have been proposed by Ranjan et al. as follows:

**Type 1:** Primary (purely mucocutaneous and not associated with systemic disorders)
- Type 1A, Isolated angiokeratomas of the oral cavity
  - Type 1A solitary (s)
  - Type 1A multiple (m)
- Type 1B, Mucocutaneous Angiokeratomas, i.e. oral angiokeratomas associated with cutaneous angiokeratomas (e.g. angiokeratoma of vulva/scrotum)
  - Type 1B regular
  - Type 1B irregular
- Type 1C, Angiokeratomas occurring simultaneously in oral cavity, skin (e.g. vulva/scrotum), and gastrointestinal mucosa
  - Type 1C regular
  - Type 1C irregular

**Type 2:** secondary (As component of a generalized systemic disorder)
- Type 2A, As a component of Fabry’s disease
  - Type 2A regular
  - Type 2A irregular
- Type 2B, As a component of fucosidosis
  - Type 2B regular
  - Type 2B irregular

On the basis of this classification, the present case was classified as type IA. Only one case of a congenital angiokeratoma in oral cavity has been reported previously in a child without metabolic disease.

To the best of our knowledge this case is second reported case of congenital solitary angiokeratoma on tongue without metabolic disease. Angiokeratoma is a dark irregular lesion which bleeds occasionally. Therefore it can be mistaken clinically for melanocytic nevus, malignant melanoma, verruca vulgaris, hemangioma, capillary aneurysms, spitz nevus or focal epithelial hyperplasia. Excisional biopsy with histologic examination is important to confirm the diagnosis. The pathogenesis of angiokeratoma is still uncertain. Vascular ectasia within the papillary dermis appears to be a primary event while epidermal changes are a secondary reaction. Increased proliferative capacity on surface of vascular malformations and close proximity of vascular spaces to epidermis in angiokeratoma could explain the reactive epidermal growth. Treatment of oral angiokeratoma is surgical excision. Prognosis of Angiokeratoma is good, although recurrences have been noted in few cases.

**Author Affiliations**
1. Dr.Manupriya Nain, Senior Resident, 2. Dr.Shilpi Agarwal, Professor, Department of Pathology, 3. Dr.Gautam Bir Singh, Assistant Professor, 4. Dr.Raghuram Devenga, Post Graduate Student, Department of ENT, Lady Hardinge Medical College, New Delhi, India.

**Acknowledgments**
We would like to thank Dr. Nain M., for looking out references and writing manuscript. Dr. Agarwal S., for final diagnosis and finalizing manuscript and Dr. Singh GB, Dr. Raghuram for operating the case and work up of patient.

**References**
10. Leung CS, Jordan RC. Solitary angiokeratoma of the oral cavity. Oral

Corresponding Author
Dr. Manupriya Nain, Senior Resident, Department of Pathology, Lady Hardinge Medical College, New Delhi, India. Email: drmanupriya@yahoo.com

Source of Support: Nil, Conflict of Interest: None Declared.