The Journal of Laryngology & Otology (2011), ***125***, *1083–1086.*

doi:10.1017/S0022215111001277

CLINICAL RECORD

Large, solitary angiokeratoma in the posterior third and base of the tongue: case report

M DUTTA, S GHATAK, G BISWAS, R SINHA

Department of Otorhinolaryngology and Head Neck Surgery, R G Kar Medical College and Hospital, Kolkata, West Bengal, India

Abstract

Objective: We present an extremely rare case of isolated angiokeratoma of the tongue.

Method: Case report and review of related literature.

Results: An 18-year-old, male adolescent presented with a fleshy, intermittently bleeding mass in the posterior third and base of the tongue. The lesion was initially suspected to be a lingual thyroid or haemangioma, but histopathological features were consistent with angiokeratoma. Magnetic resonance imaging revealed that the lesion extended up to the vallecula and involved the lamina propria and superficial tongue musculature. No similar lesions were found elsewhere in the body. No metabolic derangements were identified in the patient or his family. The $2.6 \times 1.5 \times 0.5$ cm mass was excised under general anaesthesia.

Conclusion: We present the 1st case of isolated lingual angiokeratoma in a male, the 4th such case overally, the largest ever documented. The lesion was situated in the posterior third and base of the tongue, a position not previously described.

Key words: Angiokeratoma; Oral Cavity; Tongue

Introduction

Angiokeratomas are mucocutaneous lesions characterised by hyperkeratosis, parakeratosis and acanthosis of the epidermis, together with dilated dermal vessels containing red blood cells. They present mostly in localised form; systemic forms occur only in rare disorders such as Fabry's disease. However, isolated mucosal angiokeratoma involving the oral cavity is extremely rare.

Case report

An 18-year-old, male adolescent attended the otorhinolaryngology clinic complaining of an uncomfortable, slowly progressing mass in his tongue, present for approximately eight months, which bled occasionally due to friction.

Examination revealed a fleshy, exophytic, reddish, granulomatous lesion with a rough, corrugated surface in the posterior third of the tongue, a few millimetres to the left of the midline (Figure 1). It was painless, firm, non-compressible and did not bleed on touch. Its posterior extent could not be assessed on direct vision, but indirect laryngoscopy showed that it extended up to the vallecula. No cervical lymph nodes were palpable. The patient was otherwise asymptomatic, his only complaints being the associated discomfort and occasional bleeding. There was no history of local trauma.

Macroscopically, the lingual mass closely resembled a haemangioma, but was non-compressible: rather, it felt firm and appeared solid. It also resembled a lingual thyroid due to its position in the posterior third of the tongue, behind the foramen caecum. However, its surface was rough and granular, and closer inspection revealed it to be situated in a slightly paramedian position.

A subsequent thyroid scan failed to show any thyroid tissue in the location of the lesion. Normal radiotracer uptake was evident at the normal anatomical position of the thyroid gland.

A punch biopsy was taken under local anaesthesia. The procedure was almost bloodless, making a diagnosis of haemangioma even less probable. Histopathological analysis showed dilated dermal vessels containing red blood cells, underneath a hyperplastic epidermis. In places, the vascular channels were almost obliterated by rete-ridges (Figure 2). These features are pathognomonic of angiokeratoma.

This diagnosis prompted a search for similar lesions elsewhere in the body. Despite a meticulous survey, none were found. The patient's presenting complaints and subsequent examination findings were inconsistent with any systemic or biochemical abnormalities, nor did any of his family members suffer from symptomatology suggestive of a definite syndrome.

Thus, a diagnosis of isolated angiokeratoma of the tongue was made.

The lesion was scheduled for excision under general anaesthesia.

Pre-operatively, a magnetic resonance imaging (MRI) scan of the oropharynx and tongue was performed to determine the exact extent and, more importantly, depth of the lesion. The T1-weighted images were isointense. However, the T2-weighted images showed a hyperintense lesion in

Accepted for publication 31 December 2010 First published online 21 June 2011

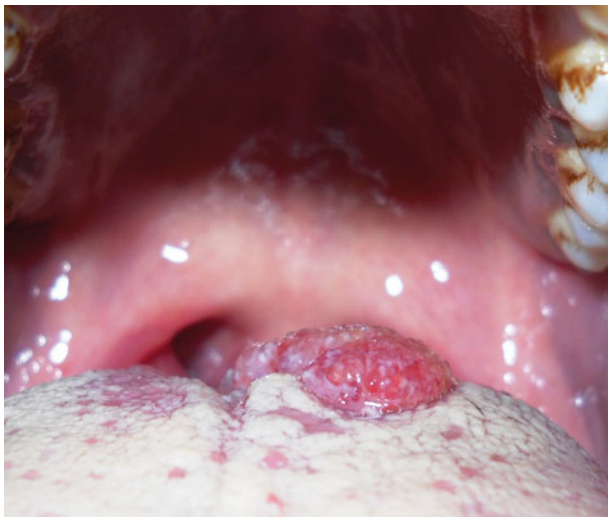

FIG. 1
Clinical photograph showing the fleshy mass in the posterior third of the tongue, with a rough surface, situated a few millimetres lateral to the midline.

the posterior third of the tongue extending up to the vallecula for a length of 2.6 cm, and involving the lamina propria and superficial tongue musculature (Figure 3). The clinico-radiological profile indicated a size of approximately $2.6 \times 1.5 \times 0.5$ cm.

The mass was excised under general anaesthesia.

The post-operative period was uneventful. The patient recuperated well, and no recurrence was seen at three months' follow up.

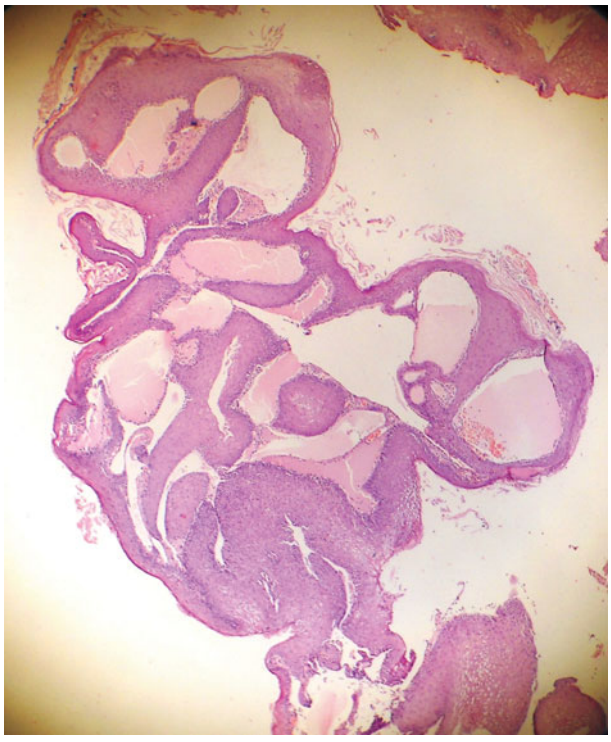

FIG. 2
Photomicrograph of punch biopsy specimen, showing hyperplasia of the epidermal layer, with dilated dermal blood vessels containing red blood cells. In places, the vascular lumen is almost obliterated by rete-ridges. (H&E; ×100)

Discussion

Angiokeratomas are benign, mucocutaneous lesions characterised histologically by hyperkeratosis, parakeratosis and acanthosis of the epidermis, together with dilated dermal vessels surrounded by rete-ridges containing blood or fibrin thrombi.[1] Grossly, they appear as dark brown, irregular, papular lesions which may erode and bleed.[2] Angiokeratomas present as one of five forms, either solitary or localised; alternatively, they can form part of a systemic disorder (Table I). Mucosal angiokeratoma of the oral cavity is generally associated with rare syndromic disorders with biochemical derangements, such as Fabry's disease and fucosidosis.[1] Additional histological features of this type of angiokeratoma include swollen, vacuolated vascular endothelial cells.[1]

However, isolated angiokeratoma of the oral cavity, without evidence of similar lesions elsewhere, or features of metabolic or biochemical derangement, is extremely rare. An extensive search in the Pubmed and Medline databases yielded only five previously reported cases of isolated

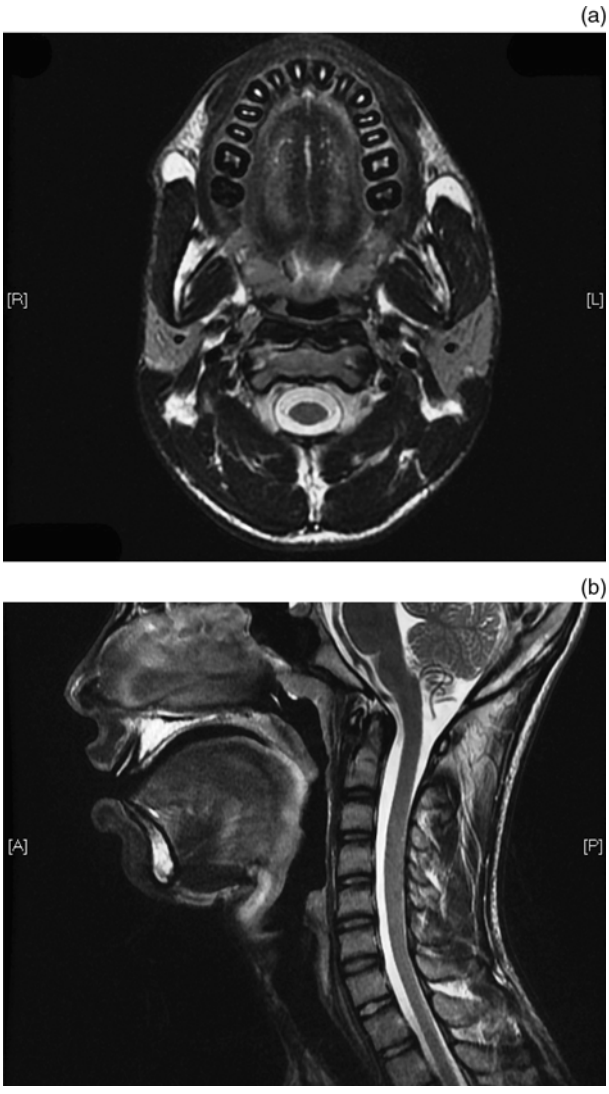

FIG. 3
T2-weighted axial and sagittal magnetic resonance imaging scans showing respectively (a) a hyperintense lesion in the posterior third of the tongue extending up to the vallecula, and (b) involvement of the lamina propria and the superficial musculature of the tongue. R = right; L = left; A = anterior; P = posterior

https://doi.org/10.1017/S0022215111001277 Published online by Cambridge University Press

TABLE I
TYPES OF ANGIOKERATOMA[1,3–5]

Type	Extent	Location & characteristics	Syndromic association
Angiokeratoma of Mibelli	Localised	Bilateral Multiple Mostly in female adolescents History of trauma or chilblains Dark red, warty papules Digits & dorsum of fingers, toes, elbows & knees at bony prominences	No
Angiokeratoma of Fordyce	Localised	Dark red or black papules Scrotum, vulva, clitoris ± oral cavity (including tongue) Multiple Seen in elderly persons	No
Angiokeratoma circumscriptum (angiokeratoma corporis naeviform*)	Localised	Multiple Unilateral Presents at birth or early childhood Mostly in females Deep blue to black, extensive patches in lower extremities May involve oral cavity	No
Solitary angiokeratoma	Localised	Solitary lesion in skin or (rarely) oral cavity Mostly in lower extremities May be associated with trauma Red-blue or black warty papules	No
Angiokeratoma corpora diffusum	Systemic	Lower extremities (umbilicus to knees) Oral cavity Dark red or punctate papules Multiple	Fabry's disease† Fucosidosis‡ Amyloidosis Vasculitis

*Congenital form. †α-galactosidase deficiency; ‡β-mannosidase deficiency.

angiokeratoma of the oral cavity, three of which affected the tongue (Table II).

To the best of our knowledge, our patient is only the fourth reported case of solitary angiokeratoma of the tongue. Unlike the other reported cases, in which the lesion was papular, our patient's lesion measured $2.6 \times 1.5 \times 0.5$ cm. Thus, it represents the largest angiokeratoma of the tongue reported to date. In addition, involvement of the posterior third and base of the tongue has not previously been reported, making our case unique. Furthermore, all previously reported solitary angiokeratomas of the tongue have been found in females, whereas our patient was male, the first such reported instance.

Angiokeratoma has been known as a pathological entity for more than a century, since Mibelli coined the term in 1891.[6] However, it was only in 1967 that solitary angiokeratoma was described as a distinct entity, by Imperial and Helwing.[7] The first case of isolated angiokeratoma of the oral cavity was reported as late as 1997, by Leung and Jordan.[2] Since then, four similar cases have been reported.[1,3,4,8] This raises suspicion that such cases have until recently been under-reported; a view proposed in a review of 14 cases of angiokeratoma of the tongue (not all cases were isolated) encountered over a 10-year period.[9]

The pathogenesis of angiokeratoma is unclear. It has been postulated that the primary inciting events are vascular ectasia in the papillary dermis (immediately inferior to the basement membrane) and associated increased proliferative capacity surrounding these vascular malformations, resulting in secondary involvement of the adjacent epidermis (in the form of the hyperplasia and hyperkeratosis which characterise angiokeratomatous lesions).[10]

The appearance and rarity of angiokeratomas invite diagnostic dilemma, and the lesion can be confused with melanocytic naevus, malignant melanoma, warts, focal epithelial hyperplasia, and vascular lesions such as haemangioma and capillary aneurysm.[10,11]

Our patient presented with a fleshy, exophytic lesion in the tongue, with occasional bleeding, which was confused with haemangioma and lingual thyroid. The final diagnosis was reached only on histopathological analysis. Both the punch biopsy and subsequent excisional biopsy revealed features

TABLE II
REPORTED CASES OF SOLITARY ANGIOKERATOMA OF TONGUE

Study	Title	Year	Citation	Pt age (y), sex
Siponem *et al.*[3]	Solitary angiokeratoma of the tongue	2006	*J Oral Pathol Med* 2006;**35**;252–3	54, female
Sion-Vardy *et al.*[1]	Solitary angiokeratoma of the tongue	2008	*Med Oral Patol Oral Circ Bucal* 2008;**13**:E12–14	45, female
Ergun *et al.*[4]	Solitary angiokeratoma of the tongue treated with diode laser	2009	*Lasers Med Sci* 2009;**24**:123–5	16, female

Pt = patient; y = years

https://doi.org/10.1017/S0022215111001277 Published online by Cambridge University Press

characteristic of angiokeratoma, with dilated dermal vessels containing red blood cells, rete-ridges almost occluding the lumen, and hyperplasia of the epidermis. A pre-operative MRI of the tongue, performed to determine the extent and texture of the lesion, showed posterior encroachment up to the vallecula, together with involvement of the subepithelial tissue (lamina propria) and the superficial tongue musculature. No similar lesions were found elsewhere in the body; the patient was completely asymptomatic except for his lingual mass. In addition, there was no history of symptoms suggestive of biochemical derangement, in the patient or his family.

- **Angiokeratomas are benign, mucocutaneous lesions characterised by epithelial hyperplasia and dermal vascular dilations, often surrounded by rete-ridges**
- **Mucosal angiokeratoma of the oral cavity is usually seen as part of a generalised syndromic disorder (e.g. Fabry's disease or fucosidosis)**
- **It may coexist with similar lesions in other sites (e.g. in Fordyce's or congenital forms)**
- **Isolated angiokeratoma of the oral cavity (including the tongue) without systemic involvement is exceptionally rare**
- **This paper presents the fourth case of isolated angiokeratoma of the tongue, and the first such case: (1) in a male, (2) in the posterior third and base of the tongue, and (3) of such large size**

The treatment of choice for a symptomatic angiokeratoma is 'cold steel' surgical excision.[1] Electrocoagulation, cryotherapy and laser ablation are viable alternatives.[4,12,13]

References

1 Sion-Vardy N, Manor E, Puterman M, Bodner L. Solitary angiokeratoma of the tongue. *Med Oral Pathol Oral Cir Bucal* 2008; **13**:E12–4

2 Leung CS, Jordan RCK. Solitary angiokeratoma of the oral cavity. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1997;**84**:51–3

3 Siponem M, Penna T, Apaja-Sarkkinem M, Palatsi R, Salo T. Solitary angiokeratoma of the tongue. *J Oral Pathol Med* 2006;**35**:252–3

4 Ergun S, Mete Ö, Yeşel S, Tanyeri H. Solitary angiokeratoma of the tongue treated with diode laser. *Lasers Med Sci* 2009;**24**: 123–5

5 Bhargava P, Bhargava S, Mathur D, Agarwal DS, Bhargava R. Angiokeratoma of tongue. *Indian J Dermatol Venereol Leprol* 2001;**67**:270

6 Mibelli V. A new form of de-keratosis, angiokeratoma [in Italian]. *G Ital Mal Vener* 1891;**30**:285

7 Imperial R, Helwig EB. Angiokeratoma. *Arch Dermatol* 1967; **95**:166–75

8 Fernandez-Flores A, Sanroman J. Solitary angiokeratoma of the tonsillar pillar of the oral cavity. *Rom J Morphol Embryol* 2009; **50**:115–17

9 Nausheen Y, Aamir A, Zubair A, Akhter H, Rashid A, Naila K *et al*. Angiokeratoma of tongue – a series of 14 cases. *J Pak Med Assoc* 2006;**56**:285–7

10 Schiller PI, Itin PH. Angiokeratomas: an update. *Dermatology* 1996;**193**:275–82

11 Segura-Saint-Gerons R, Toro-Rojas M, Ceballos-Salobreña A, Aparicio-Soria JL, Fuentes-Vaamonde H. Focal epithelial hyperplasia. A rare disease in our area. *Med Oral Patol Oral Cir Bucal* 2005;**10**:128–31

12 del Pozo J, Fonseca E. Angiokeratoma circumscriptum naeviforme: successful treatment with carbon-dioxide laser vaporization. *Dermatol Surg* 2005;**2**:232–6

13 Gorse SJ, James W, Murison MS. Successful treatment of angiokeratoma with potassium tritanyl phosphate laser. *Br J Dermatol* 2004;**3**:620–2

Address for correspondence:
Dr (Prof) Ramanuj Sinha,
4RA, 2/1, Purbachal, Salt Lake,
Kolkata 700097,
West Bengal, India

Fax: +91 33 28281551
E-mail: sinha_ramanuj@yahoo.co.in

Dr R Sinha takes responsibility for the integrity of the content of the paper
Competing interests: None declared