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CLINICAL RECORD

Solitary fibrous tumour of the tongue: a series of four cases

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Abstract

Background: Solitary fibrous tumour is a soft tissue tumour of mesenchymal origin. It was first described in the pleura and has since been reported in many anatomical locations. Thirteen cases in the tongue have hitherto been reported. A positive CD34 result has traditionally been used to confirm the diagnosis, although this is often non-specific to solitary fibrous tumour. To date, nuclear STAT6 expression has not been reported in solitary fibrous tumour of the tongue.

Method: This paper presents a further four cases of solitary fibrous tumour of the tongue, the largest series to date. Clinical, histopathological and immunohistochemical findings are detailed, including nuclear STAT6 expression.

Results: All four cases were positive for CD34; two cases showed nuclear expression of STAT6. The tumours were excised completely and there have been no recurrences in at least one year.

Conclusion: Solitary fibrous tumour should be considered as a differential diagnosis for tongue swellings, with the potential to recur.

Key words: Solitary Fibrous Tumour; Signal Transducer And Activator Of Transcription 6; CD34 Antigen

Introduction

Solitary fibrous tumour is a soft tissue tumour of mesenchymal origin. It was first described in the pleura,[1] and has since been reported in many anatomical locations including the skin and, rarely, the oral cavity.[2–5] Solitary fibrous tumours of the oral cavity were first described by Suster *et al*. in 1995.[6] In the largest series of oral solitary fibrous tumours to date,[7] only 2 of the 21 cases affected the tongue, whereas 11 affected the buccal mucosa or vestibules.

In its latest classification of soft tissue and bone tumours,[8] the World Health Organization categorised solitary fibrous tumour as a fibroblastic or myofibroblastic tumour of 'intermediate' malignancy; that is, a tumour which, though usually benign, may on occasion metastasise. Histologically, solitary fibrous tumour is a spindle cell neoplasm with a prominent vascular pattern. However, its microscopic features are not specific, and immunohistochemistry is required to confirm the diagnosis.

Until recently, cluster of differentiation (CD) 34 antigen, for which solitary fibrous tumour is strongly positive,[3,4,9,10] was the usual marker used. However, CD34 is itself non-specific. Recently, nuclear (rather than membranous or cytoplasmic) expression of signal transducer and activator of transcription 6 (STAT6) has been the 'gold standard'.[11] STAT6 expression has not previously been reported in solitary fibrous tumour affecting the tongue. In this report, we describe four cases of lingual solitary fibrous tumour. All four tumours were surgically excised and none have recurred in at least one year.

Clinical presentations

Case one

A 46-year-old man was urgently referred with a swelling on the left lateral border of the tongue. The swelling had been gradually enlarging over a period of one year, causing aesthetic and functional problems. His past medical history included asthma, which was well controlled through the use of inhalers. He smoked 20 cigarettes per day and denied consumption of any alcohol. Clinical examination revealed a swelling that was pedunculated and lobulated, with a maximum dimension of 2.5 cm (Figure 1). The overlying mucosa appeared thin, but not ulcerated. There were no complaints of paraesthesia or bleeding from the tongue, nor were there any palpable regional lymph nodes. A benign process was favoured, with differential diagnoses of fibroepithelial polyp, granular cell tumour and lipoma. The mass was excised under general anaesthesia using carbon dioxide laser. Healing was uneventful and there has been no recurrence in one year.

Case two

A 20-year-old man was urgently referred with a swelling on the dorsum of the tongue, just to the left of the midline, which had been gradually enlarging over a period of 6 months. There was no history of trauma to the area in question. The patient was medically healthy, and did not consume tobacco and declared negligible alcohol consumption. On examination, there was a well-circumscribed

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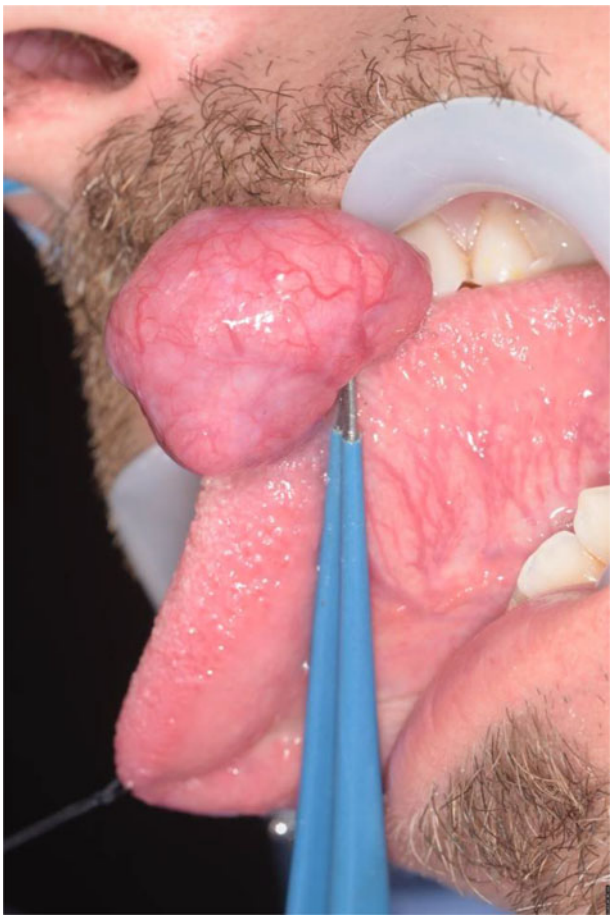

FIG. 1
Clinical presentation of case one.

polypoid growth with a sessile base 4 mm in maximum dimension. The overlying mucosa was normal and there was no lymphadenopathy. A provisional diagnosis of fibro-epithelial polyp was made. Excisional biopsy of the mass was performed under local anaesthesia. Healing was uneventful and there has been no recurrence in one year.

Case three

A 49-year-old woman was referred for a swelling on the dorsum of the tongue of 4 months' duration. Although symptomless, she noticed it was increasing in size. The patient was medically fit and well, did not consume tobacco, and consumed alcohol within the recommended weekly limit. Clinical examination of the tongue revealed a firm, sessile polyp approximately 4 mm in diameter. The overlying mucosa was normal. Examination findings of the head and neck were otherwise unremarkable. A provisional diagnosis of fibro-epithelial polyp was made. Excisional biopsy of the mass was performed under local anaesthesia. Healing was uneventful and there has been no recurrence in 18 months.

Case four

A 46-year-old woman was referred for a swelling on the left side of her tongue of 1 year's duration. The patient recalled no traumatic episode prior to the development of the lump, which was symptomless and of constant size. The patient had a history of asthma and psoriatic arthritis. She was a non-smoker and consumed moderate amounts of alcohol. Clinical examination of the tongue revealed a defined nodule 7 mm in diameter on the lateral aspect of the left anterior tongue. The overlying mucosa was normal. Examination findings of the head and neck were otherwise unremarkable. A provisional diagnosis of a lipoma, fibroma or mucocoele was made. Excisional biopsy of the lesion was performed under local anaesthesia. Healing proceeded normally and there has been no recurrence in five years.

Histopathology and immunohistochemistry

Microscopically, all cases involved a circumscribed tumour consisting of cytologically bland, densely packed spindle-shaped cells with minimal cytoplasm lying in a fibromyxoid stroma (Figure 2). Thin-walled blood vessels were present throughout and assumed a 'staghorn' appearance. Mitoses were scarce.

Immunohistochemistry showed strongly positive membranous and cytoplasmic expression of CD34 in all cases (Figures 3 and 4) and nuclear expression of STAT6 in two cases (cases one and four) (Figure 5). The cell proliferation index (as determined by nuclear expression of Ki67) was

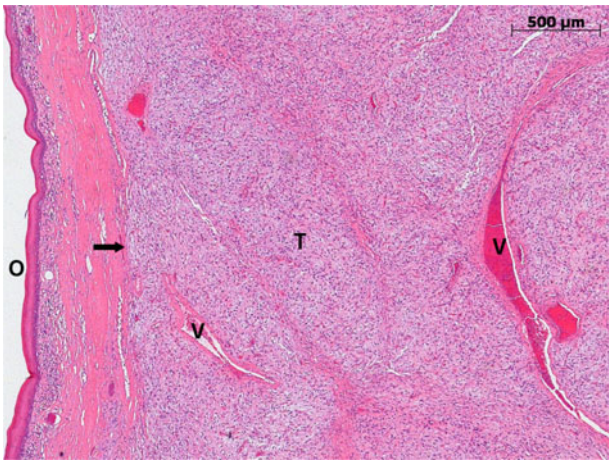

FIG. 2
Low power view of solitary fibrous tumour of the tongue, showing densely packed spindle cells with prominent blood vessels (V). The tumour (T) has a defined margin (arrow). (H&E stain.) O = oral mucosal surface

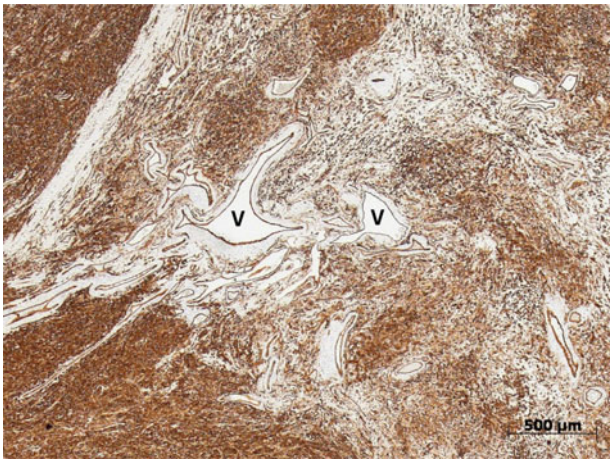

FIG. 3
Tumour is diffusely positive for CD34 (golden brown reaction product). (Immunoperoxidase stain.) V = 'staghorn' blood vessels

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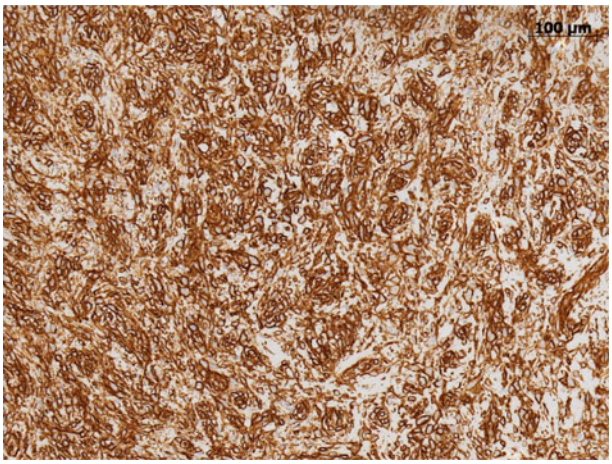

FIG. 4
High power view showing the membranous staining pattern of CD34, which highlights the outline of the tumour cells. (Immunoperoxidase stain)

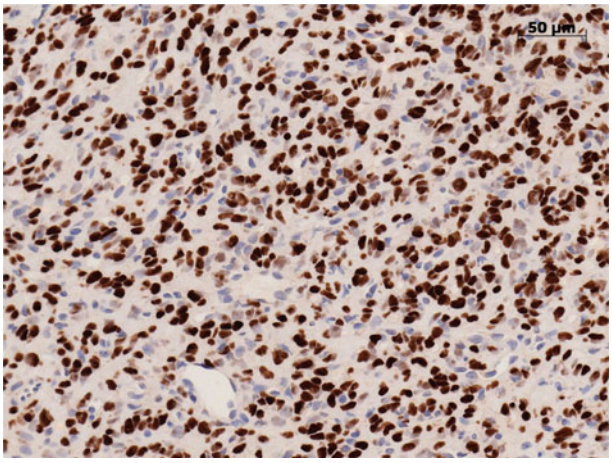

FIG. 5
High power view showing the nuclear staining pattern of STAT6 in the tumour cells. (Immunoperoxidase stain)

low (less than 5 per cent) in all four cases. These morphological and phenotypic features are consistent with solitary fibrous tumour.

All tumours were treated as benign. Complete excision of the tumour was confirmed histologically in all cases. Table I summarises the details of the four cases.

Discussion

Solitary fibrous tumour was first described in 1992 by Klemperer and Rabin in the parietal or visceral pleura.[1] However, the name betrays the fact that the tissue of origin is uncertain. Many reports have since shown it is not a tumour specific to this location.[6,8,10]

Solitary fibrous tumour usually presents in the fifth decade of life, as was the case in three of the patients presented here, and represents less than 2 per cent of all soft tissue tumours.[12] They are usually indolent and painless but if traumatised, may become symptomatic (as in case one).

Chan proposed a set of criteria to assist in histopathological diagnosis.[10] Specifically, solitary fibrous tumour is characterised by: short, spindled or ovoid cells, with scanty and poorly defined cytoplasm; alternating hypercellular and hypocellular foci, with sclerosis in the latter; circumscription; scanty mitotic figures (less than 4 mitoses per 10 high power fields); and intimate intertwining of collagen fibrils with spindle cells.[10] Nonetheless, in sections stained with haematoxylin and eosin (Figure 2), solitary fibrous tumour has a number of differential diagnoses,[12] and immunohistochemistry for STAT6 and CD34 is necessary to clinch the diagnosis.[10,13]

Of the 13 lingual solitary fibrous tumour cases already described in the literature, all were positive for CD34.[2–5,7,11,14–19] However, our series is the first to demonstrate nuclear STAT6 reactivity in lingual solitary fibrous tumour (Figure 5). STAT6 is more specific for solitary fibrous tumour than CD34, and, although expressed in the majority of solitary fibrous tumours, STAT6 is not expressed by them all.[20] It is currently unknown whether lack of STAT6 expression in solitary fibrous tumour has any prognostic significance.

Although benign, solitary fibrous tumour may be locally infiltrative, and either recur or, occasionally, metastasise.

TABLE I
SUMMARY OF CLINICAL PRESENTATIONS, INVESTIGATIONS, TREATMENTS AND OUTCOMES

Parameter	Case 1	Case 2	Case 3	Case 4
Patient details	46-year-old male	20-year-old male	49-year-old female	46-year-old female
Presentation	Lobulated mass on left lateral border of tongue, approximately 2.5 cm in diameter	Sessile, enlarging lump on tongue dorsum, approximately 4 mm in diameter	Firm, sessile polyp on tongue dorsum, approximately 4 mm in diameter	Firm, defined nodule on tongue lateral border, 7 mm in diameter
Duration of presenting complaint (months)	12	6	4	12
Other complaints	Occasional pain	No other complaints	No other complaints	No other complaints
Treatment modality	Excision with carbon dioxide laser	Excision by conventional scalpel biopsy	Excision by conventional scalpel biopsy	Excision by conventional scalpel biopsy
Immunohistochemistry	– Strong expression of CD34 & STAT 6 – Negative for cytokeratins, S100, CD31, desmin & EMA – Ki67 index <5%	– Strong expression of CD34 – Negative for cytokeratins, S100, SMA, CD31, desmin & EMA – Ki67 index <5%	– Strong expression of CD34 – Negative for cytokeratins, S100, SMA, desmin & EMA – Ki67 index almost zero	– Strong expression of CD34 & STAT6 – Negative for cytokeratins, S100, SMA & EMA – Ki67 index <5%
Outcome	No recurrence or metastasis in 1 year	No recurrence or metastasis in 1 year	No recurrence or metastasis in 18 months	No recurrence or metastasis in 5 years

CD = cluster of differentiation; EMA = epithelial membrane antigen; SMA = smooth muscle actin

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A review by Cox *et al*. showed that of 142 cases of solitary fibrous tumour in the head and neck region, 10 had atypical or malignant features, and recurrence was largely dictated by the presence of positive margins at the time of excision, rather than the microscopic grade of the tumour itself.[21]

A high proliferation index is cited to be a risk factor in dictating aggressive tumour behaviour. Künzel *et al*. demonstrated that tumours could progress from a low to high proliferative index, and hence warranted long-term follow up.[22]

- **Solitary fibrous tumours are of mesenchymal origin; several extra-pleural locations have been reported including the head and neck**
- **There are 13 reported cases of solitary fibrous tumour of the tongue in the literature**
- **All 13 cases were positive for CD34, but none reported nuclear STAT6 expression**
- **In our series of four cases involving the tongue, all were positive for CD34 and two demonstrated nuclear STAT6 expression**
- **Nuclear STAT6 expression is more specific than CD34 positivity in solitary fibrous tumour diagnosis**
- **Surgical excision is the 'gold standard' treatment; to date, there have been no recurrences following excision**

There is only one report of a frankly malignant solitary fibrous tumour of the tongue.[5] In addition to infiltration, this tumour showed severe nuclear atypia, a high mitotic count and areas of necrosis. However, neither this nor any of the previously reported cases of solitary fibrous tumour in the tongue have recurred or metastasised; all have successfully been treated with complete surgical excision. In all four of the cases reported here, there has been no recurrence or metastasis in at least one year.

Conclusion

Although rare, solitary fibrous tumour should be considered in the differential diagnosis of tongue swellings. All of our cases were initially diagnosed clinically as fibro-epithelial polyps. Solitary fibrous tumour is usually benign and can be treated with simple surgical excision. However, review is necessary, as solitary fibrous tumour has the potential to recur and on occasion metastasise.

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