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CASE REPORT

Solitary fibrous tumour of the tongue: a case report

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Abstract

Solitary fibrous tumours (SFTs) are uncommon benign mesenchymal neoplasms that occur relatively rarely in the oral cavity. An SFT in the tongue of a 67-year-old woman is presented. A firm, asymptomatic, dome-shaped mass was found on the left ventral surface of the tongue. Complete surgical resection of the mass was performed. Microscopically, the tumour was well-circumscribed and composed of variably cellular and patternless distributions of bland spindle and ovoid cells within variably collagenous stroma, with interspersed large branching or "staghorn"-shaped vessels. Immunohistochemically, tumour cells were positive for CD34 and STAT6, but negative for CD68, S-100 protein, epithelial membrane antigen and α-smooth muscle actin.

Introduction

Solitary fibrous tumours (SFTs) are fibroblastic mesenchymal neoplasms that were first described by Klemperer and Rabin in 1931. SFTs were initially described in the pleura[1], but they are now found at almost every anatomic site[2]. The occurrence of SFTs in the oral cavity is relatively rare[3–5].

Microscopically, SFTs show a broad spectrum, with appearances often varying from field-to-field within one tumour. This diversity sometimes makes it difficult to distinguish SFTs from other lesions that show similar histopathological findings[2,3].

Immunohistochemically, most SFTs show diffuse moderate to strong reactivity for CD34, and CD34 immunoreactivity is considered to be one of the effective tools for the diagnosis of SFTs[2–5]. Recently, *NAB2-STAT6* gene fusion has been reported to be a genetic hallmark of SFTs[6–8], and immunostaining for STAT6 has been proposed as a useful tool for the diagnosis of SFTs[9,10]. In the oral region, reports using immunohistochemical staining for STAT6 are rare, except for one study of Kao *et al.*[3]. In this report, a rare case of SFT on the ventral surface of the tongue that occurred in a 67-year-old woman is presented, along with the immunohistochemical findings for STAT6.

Case report

A 67-year-old woman was referred to our clinic for the evaluation of a painless nodule on the ventral surface of her tongue. She noticed the nodule one year earlier, and it had gradually increased in size. She had a history of hyperlipidaemia that was well-controlled by medication. On local examination, a relatively firm, well-circumscribed, dome-shaped mass with normal colour and measuring 6.0 mm in diameter was found on the left ventral surface of the tongue (Fig. 1). There were no other abnormalities in the oral region and no obvious submandibular lymphadenopathy. Based on the initial clinical diagnosis of a benign soft tissue neoplasm, excisional biopsy of the mass was performed (Fig. 2).

Microscopically, the specimen was well-circumscribed, and the tumour was composed of variably

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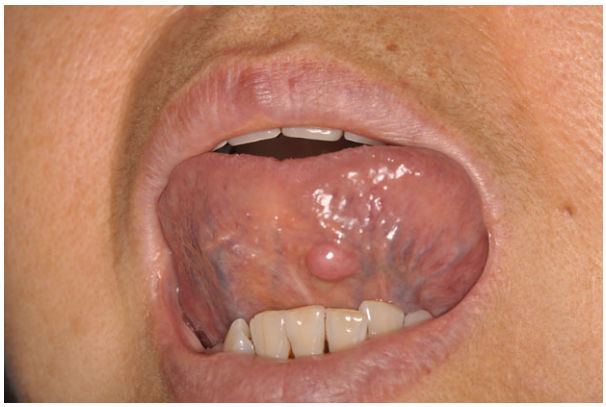

Figure 1 Intraoral photograph showing a firm, dome-shaped mass on the left ventral surface of the tongue.

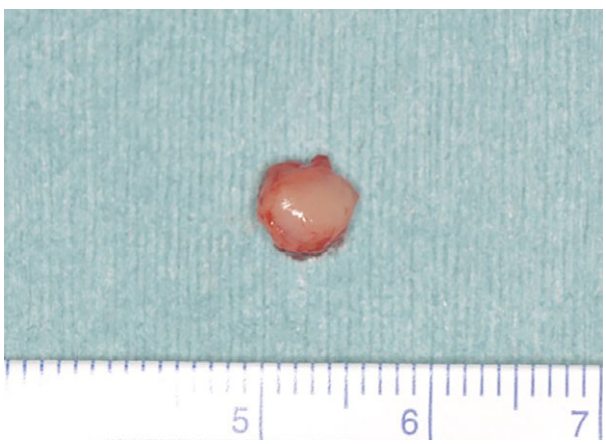

Figure 2 Gross appearance of the resected mass.

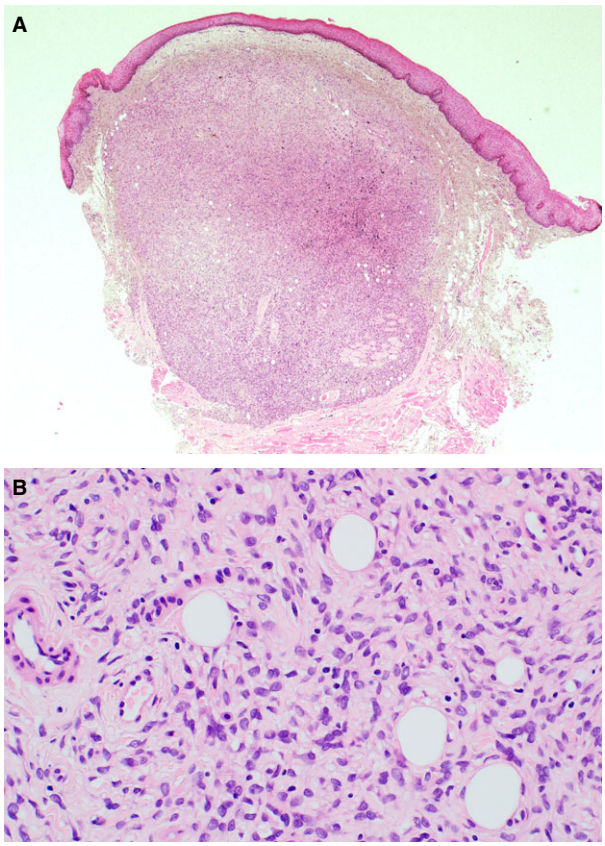

Figure 3 Histopathological findings of haematoxylin and eosin staining. (A) The tumour is well-circumscribed and hypercellular, and hypocellular areas with variably collagenous stroma and "staghorn"-shaped dilated vessels are found (Original magnification × 20). (B) Patternless proliferation of bland spindle and ovoid cells within collagenous stroma is found (original magnification × 200).

cellular and patternless distributions of bland spindle and ovoid cells within variably collagenous stroma that frequently showed areas of dense hyalinisation, as well as interspersed large branching or "staghorn"-shaped thin-walled vessels. Mature adipocytic cells and nuclear atypia and mitotic activity were not found (Figs. 3A and 3B).

Immunohistochemically, the tumour cells were diffusely moderately to strongly immunoreactive for CD34 (Fig. 4) and negative for CD68, S-100 protein, epithelial membrane antigen (EMA), and α-smooth muscle actin (SMA). Less than 1% of tumour cells were positive for Ki67. SFT was highly suspected, and additional immunohistochemical staining with STAT6 was performed. The tumour cells were diffusely moderately positive for STAT6 (Fig. 5).

The patient's post-operative course was uneventful, and there was no recurrence during the 2-year follow-up period.

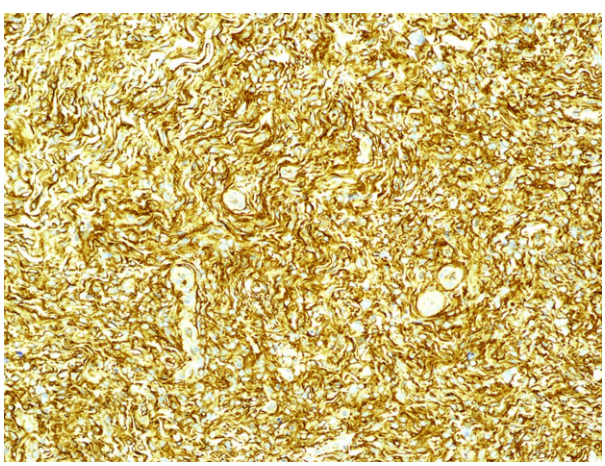

Figure 4 Immunohistochemical staining with CD34 showing diffuse moderate to strong staining of tumour cells (Original magnification × 100).

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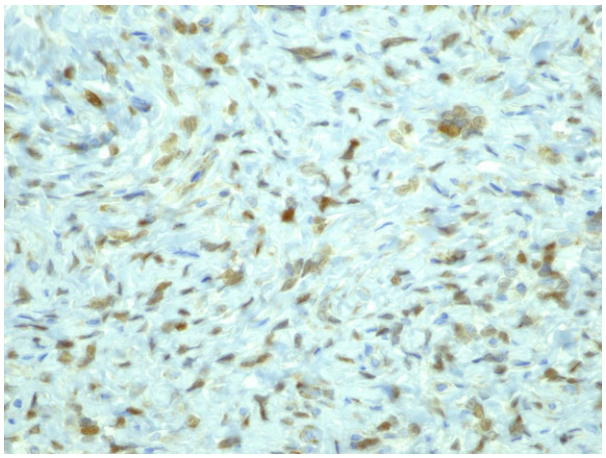

Figure 5 Immunohistochemical staining with STAT6 showing diffuse moderate staining of tumour cells (Original magnification × 200).

Discussion

SFTs are fibroblastic mesenchymal neoplasms that occur ubiquitously in various anatomical locations, whereas occurrence in the oral region is uncommon[3]. SFTs account for 3% of all mesenchymal tumours of the oral region[5]. There have been approximately 90 cases of oral SFTs reported[11]. Oral SFTs most commonly affect the buccal mucosa and tongue and predominantly affect women in their sixth decade of life[3,5]. To the best of our knowledge, 15 cases of SFTs in the tongue have been reported, including the present case[12]. Clinically, oral SFTs present as submucosal, slow growing, asymptomatic masses of various sizes[5]. Most of the tongue cases have a dome-shaped appearance and are less than 30 mm in size[3,12]. The differential diagnosis of oral SFTs includes mucocele, salivary gland tumours, lipoma, vascular malformations and leiomyoma, and the present case had a mucocele-like appearance, although it was slightly harder[13].

Microscopically, an SFT is characteristically a circumscribed neoplasm composed of variably cellular and patternless distributions of bland spindle and ovoid cells within variably collagenous stroma that frequently shows areas of dense hyalinisation, as well as interspersed large branching or "staghorn"-shaped thin-walled haemangiopericytic vessels. Nuclear atypia and mitotic activity are generally scarce, and mature adipocytic and multinucleated cells may be found[2,12]. SFTs, which have a wide histological spectrum, can sometimes be difficult to distinguish from other benign and malignant tumours that have similar histological features[9]. Immunohistochemical staining is very effective to distinguish SFTs from other fibroblastic tumours. SFTs show positive reactivity for CD34, CD99, Bcl-2 and EMA, while desmin, cytokeratin and S-100 protein are usually negative[13]. On immunohistochemical staining, CD34 has been considered the most reliable marker for the diagnosis of SFTs[2–5,13]. However, CD34 expression is also common in other tumours such as soft tissue perineuroma, dermatofibrosarcoma protuberans and spindle cell lipoma, which are included in the differential diagnosis of SFT[9].

Recently, SFT has been recognised as a translocation-associated neoplasm, with the *NAB2-STAT6* gene fusion derived from inv 12 (q13q13), and the fusion arises from recurrent intrachromosomal rearrangements on the chromosome, resulting in nuclear expression of the C-terminal portion of STAT6[6–8]. Doyle *et al.*[9] investigated STAT6 expression by immunohistochemistry in SFTs and other soft tissue tumours arising outside the central nervous system to validate the diagnostic utility of this novel marker. They reported that 59 of 60 SFT cases (98%) showed nuclear expression of STAT6, and non-SFT cases were negative except for three dedifferentiated liposarcomas and one deep fibrous histiocytoma, which showed weak staining. Yoshida *et al.*[10] reported that all SFT cases (49 cases) showed STAT6 staining positivity, and 4 of 159 non-SFT cases (2.5%, two low-grade fibromyxoid sarcomas, one myxoid/round-cell liposarcoma and one ovarian fibroma) showed weak nuclear expression. Currently, as most SFTs show strong and diffuse nuclear expression, STAT6 is a highly sensitive and almost perfectly specific immunohistochemical marker for SFT and can be helpful to distinguish this tumour type from histological mimics[9,10].

In SFTs of the oral region, Kao *et al.*[3] described the variability in *NAB2-STA6* fusion variants in oral SFTs, and their immunohistochemical study showed a positive staining rate of STAT6 of oral SFTs of 97.2% (35 of 36 cases). In the present case, immunohistochemically, the tumour cells were immunoreactive for CD34 and lacked CD68, S-100 protein, EMA and α-SMA immunoreactivity. Furthermore, STAT6 immunoreactivity suggested the diagnosis of SFT. Immunostaining with STAT6 is the most promising tool for the diagnosis of SFTs, but since it is difficult to differentiate SFTs with only STAT6 staining, combined staining with STAT6, CD34, and other appropriate antibodies is used practically for accurate diagnosis[2–5,13].

Most SFTs are benign lesions and cured by surgical resection, whereas in about 10% of cases, they are aggressive and show local or distant recurrences

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even many years after resection[2]. With respect to malignant transformation, clinically aggressive classical SFTs cannot always be distinguished morphologically from those that will behave indolently. There have been reports of malignant oral SFTs[14,15]. It is crucial that patients with SFTs, including those in the oral cavity, receive long-term follow-up.

Conflict of interest

The authors have no financial interests to disclose. This study had no funding sources.

Ethical approval

None required.

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