

## Pediatric Solitary Fibrous Tumor of the Sublingual Gland with Malignant Potential

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### – ABSTRACT –

Solitary fibrous tumor(SFT) is an unusual primary mesenchymal spindle cell tumors, most commonly arising in the pleura. Although the majority of these tumors exhibit benign behavior and have a good prognosis, but minority of patients with these tumors are at risk for local recurrence and distant metastaseis. Recently, we experienced a rare case of 12-years-old pediatric SFT patient arising in the sublingual gland, which shows histopathologically malignant potential. So we report this case with a review of literatures. (J Clinical Otolaryngol 2018;29:114-118)

**KEY WORDS** : Solitary fibrous tumors · Mouth floor · Sublingual gland.

### Introduction

Solitary fibrous tumors were first identified in 1931 and were originally thought to be primary pleural neoplasm. The most common primary sites of SFT are pleural, meningeal, and extrathoracic soft tissues, although it can arise in a variety of anatomical locations.<sup>1)</sup> In the head and neck region, SFT are reported in the meninges, orbit, nose, paranasal sinuses, nasopharynx, parapharyngeal space, salivary glands, epiglottis, larynx, thyroid, skin, and deep soft tissues of the neck.<sup>2)</sup> SFT of the sublingual gland are extremely rare, and only 4 previous cases reported in English literature by the author's best knowledge.<sup>3-6)</sup> Recently

we treated first pediatric SFT in sublingual gland with malignant potential, so we describe the case with review of literatures.

### Case Report

A 12-year-old female patient was referred to the authors' department with a two month history of a painless submucosal tumor of the mouth floor. Clinical examination revealed a tumor in the left side of the mouth floor, covered by healthy mucosa (Fig. 1). Computed tomography (CT) and ultrasonography showed a solid smooth mass in sublingual gland of the floor of mouth. Magnetic resonance imaging (MRI) revealed a 30 × 35 mm well circumscribed mass in the floor of mouth (Fig. 2). Fine needle aspiration was performed and the cytomorphicologic examination showed collagen scattered in the bloody background which had no malignant features.

We could resect the encapsulated tumor en loc under general anesthesia (Fig. 3A). Glossly, the surgical specimen was a circumscribed, spherical mass of

논문접수일 : 2018년 1월 4일  
논문수정일 : 2018년 4월 26일  
심사완료일 : 2018년 5월 29일  
교신저자 : 박정제, 52727 경남 진주시 강남로 79  
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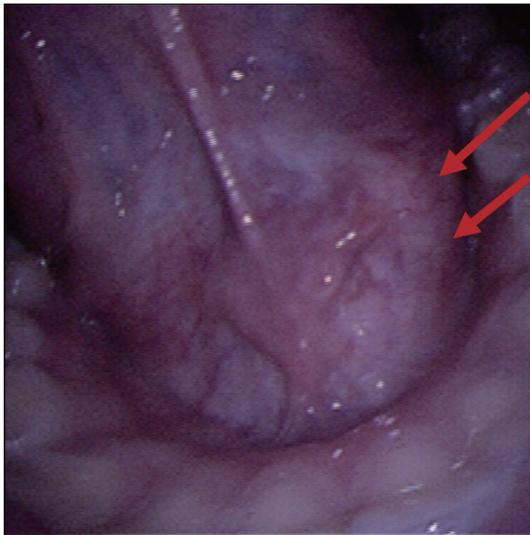
firm consistency measuring 35 mm in its greatest dimension (Fig. 3B). The postoperative clinical course was uneventful.

Histopathologic analysis revealed a hypocellular fibrous tumor with patternless architecture and abundant scattered vessels among the cells (Fig. 4A). Mitotic activity exceed up to 7 mitotic figures per 10 high power fields. They were mostly of small size, and of-

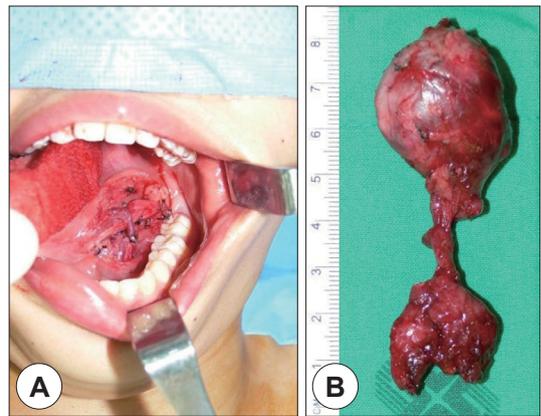
ten displayed branching lumens (Fig. 4A).

Immunohistochemical stains were performed. The tumor cells were diffusely positive for CD34 antigen (Fig. 4B). There was also focal positivity of tumor cells for smooth muscle actin, and staining for smooth muscle actin S-100 protein was negative (Fig. 4C, D).

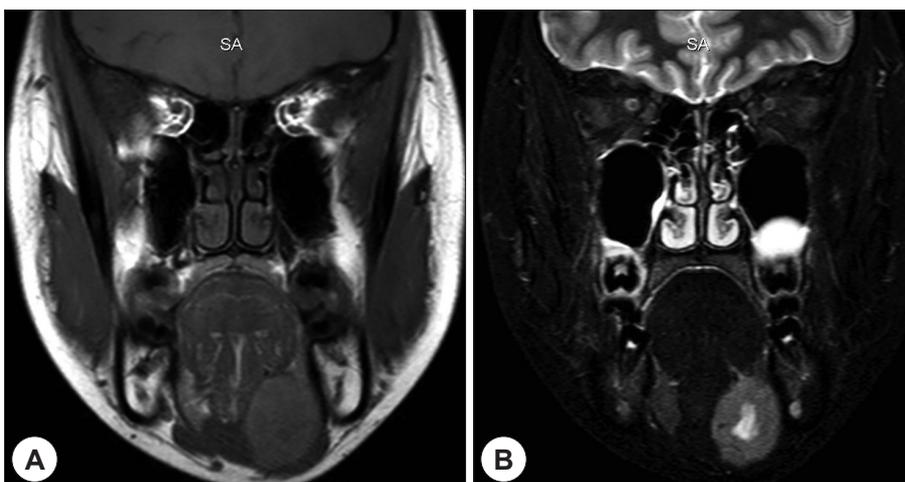
These finding led to the diagnosis of extrapleural SFT with malignant potential. And also, there were tumor cells on the surface with undetermined resection margin. Thus, we recommended adjuvant concurrent radiotherapy. However, the patient's mother



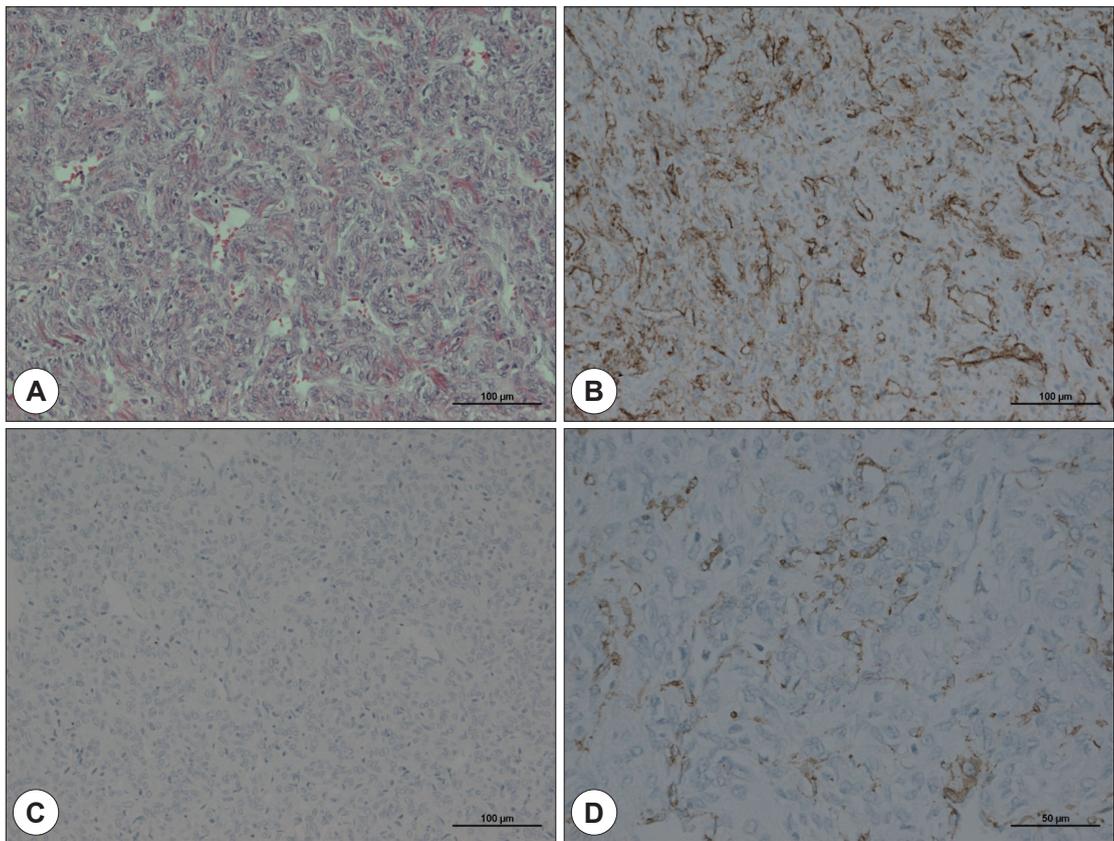
**Fig. 1.** The submucosal tumor in the left side of mouth floor covered with healthy mucosa.



**Fig. 3.** A : View of operation field after mass excision. B : En bloc resection specimen of the tumor with sublingual gland.



**Fig. 2.** MRI revealed a 30×35 mm sized mass at the left side of the mouth floor which has non-enhancing pattern and cystic fluid component. A : T1 weighted enhancing image. (B) T2 weighted image.



**Fig. 4.** A : Microscopic appearance of high cellular pleomorphism, high mitotic activity, and abundant vascularity. H-E stain ( $\times 200$ ). B : Tumor cells are diffusely and strongly stained with CD34 ( $\times 200$ ). C : Negative for S-100 ( $\times 200$ ). D : Weakly positive for SMA (Smooth muscle actin) ( $\times 400$ ).

refused further treatments. We are closely observing the patient for 3 years without evidence of recurrence.

## Discussion

Solitary fibrous tumors are primary mesenchymal spindle cell neoplasm.<sup>7)</sup> This tumor was first reported in 1931 arising in pleura, it has since acquired several synonyms including hemangiopericytoma, localized fibrous mesothelioma, localized pleural mesothelioma, fibrous mesothelioma, solitary fibrous mesothelioma, pleural fibroma, subserosal fibroma, and submesothelial fibroma.<sup>8)</sup> Subsequently, this has been documented in extrapleural, almost every anatomic site including the deep soft tissue, skin, gastro-

intestinal tract, urogenital system, head and neck, female genital tract, intracranial and spinal cord meninges, adrenal gland, pelvis, retroperitoneum, pancreas, liver, kidney, lung, and bones.<sup>9)</sup> A better knowledge of immunohistochemical and molecular features has grouped these entities into the same tumor histotype, named as SFT. Recently, the 2016 WHO Classification of Tumors of the Central Nervous System still distinguishes SFT of the meninges and meningeal hemangiopericytoma, belonging to WHO grade II and III tumors, respectively.<sup>9)</sup>

SFTs of the head and neck are very unusual. Those that develop in the oral cavity arise most commonly in the buccal region including the mucosa, then in the tongue and the lips.<sup>10)</sup> SFT of the sublingual gland are

extremely rare, and only 4 previous cases reported in English literature.<sup>3-6)</sup>

The etiology of extrapleural SFT is currently not defined, and no association has been demonstrated with smoking habit or asbestos exposure.<sup>9)</sup> Some author suggested that trauma may impact the site predilection for SFT within the oral cavity.<sup>11)</sup>

The majority of these tumors exhibit benign behavior and have a good prognosis, the malignancy incidence estimated to be lower than 0.1/100,000/year.<sup>9)</sup> Malignant SFT is usually grossly indistinguishable from the benign forms, but it may also show a more irregular cut surface, with evidence of necrotic areas or infiltration of the nearest tissues.<sup>12)</sup> It could be distinguished from benign SFTs only by an invasive growth pattern, high cellular pleomorphism, and abundant mitotic activity (mitotic index > 4/10 HPF).<sup>8,9,12)</sup> In this case, microscopic examination shows high cellular pleomorphism, and high mitotic activity up to 7 mitotic figures per 10 high power fields. These findings lead us to decide the tumor has a malignant potential, and decide to regarding post-operative adjuvant radiation therapy.

Extrapleural SFT occur as slowly growing masses, being often an asymptomatic incidental finding, but sometimes symptoms are due to the pressure effects on adjacent structures.<sup>9)</sup> In addition, clinical features can be rarely associated with multiple paraneoplastic syndromes, including refractory hypoglycemia, hypertrophic pulmonary osteoarthropathy, and elevated beta human chorionic gonadotropin.<sup>13)</sup> Out patient presents just complains of oral discomfort due to mass lesion, and any other studies shows no evidence of paraneoplastic syndrome.

Diagnosis of SFT is challenging and requires an integrated approach that includes clinical, histological, immunohistochemical, and molecular findings.<sup>9)</sup> Computed tomography and magnetic resonance imaging were obtained for further characterization of the mass, but give limited information for both diagnostic purpose and differentiating malignancy.<sup>8)</sup> The diagnostic performance of 18F-FDG-PET/CT also

seems to be suboptimal in these tumors, especially in their malignant variants.<sup>14)</sup> The use of fine needle aspiration cytology provides good diagnostic opportunities in extrapleural SFTs with ultrasound-guided approachable locations and provides the clue for management.<sup>9)</sup>

The final diagnosis is confirmed by histopathology with immunohistochemistry staining. Histologically, conventional SFT shows a variable cellularity constituted by bland ovoid or spindle cells arranged in a "patternless" distribution in a variable collagen stroma.<sup>15)</sup> The stroma may present a variable degree of myxoid changes.<sup>9)</sup> Large branching or "staghorn"-shaped thin-walled vessels are almost invariably present, and medium-sized vessels with variable perivascular hyalinization are also a common feature.<sup>16)</sup> The background contains irregular fragments of collagen and a few inflammatory elements.<sup>9)</sup> Our specimen also showed a hypocellularity, mixed ovoid or spindle cell pleomorphism, fibrous tumor with patternless architecture and many branching vessels (Fig. 4A). Immunohistochemically, SFT shows reactivity for CD34 (80–90%), Bcl-2 (30%), factor XIIIa and CD99 (70%), nuclear  $\beta$ -catenin (40%), and EMA (20–30%). CD34 is the best sensible marker to diagnose SFT.<sup>2)</sup> Recently, immunohistochemical markers such as STAT-6 and  $\beta$ -catenin have been identified.<sup>2,9)</sup> We performed CD34, S-100, SMA staining for more precise diagnosis and the results were strongly stained for CD 34, weakly for SMA, and negative for S-100 (Fig. 4B, C, D).

Complete surgical excision with clear margin is the primary treatment.<sup>17)</sup> Although in patients with malignant components or positive surgical margins, adjuvant radiation may be beneficial.<sup>9)</sup> Adjuvant chemotherapy also remains controversial.<sup>9)</sup> Several recent studies reported the efficacy of molecular targeted drugs for SFT.<sup>9,18)</sup> But it's effect also has debates, and more larger sample sized research should be required.

Typically the prognosis is known as good. The occurrence of metastasis is associated with poorer prognosis because approximately 75% of patients with

metastasis die with a median survival duration ranging from 22 to 46 months.<sup>9)</sup> Although there is no total agreement, some data seem to suggest that cases of extrapleural SFT with a more aggressive clinical behavior are usually characterized by infiltrative margins, pleomorphism, hypercellularity, high mitotic index, and necrosis.<sup>9)</sup> In our case, we were expected to have a more aggressive clinical course because of the findings of histologic examination, which was why we intended adjuvant therapy. Because of the lack of additional treatment due to her parent's objection, close follow-up is performed for 3 years after surgery. Up to now, no evidence of recurrence or metastasis has been observed.

## Conclusion

In summary, we experience a case of pediatric SFT arising from sublingual gland with malignant potential, which has performed surgical treatment only. SFT is usually benign; however, it can be lead to aggressive course. In such a cases, risk of local recurrence or distant metastasis is non-negligible, and more careful watching and close follow-up should be needed.

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