

原文題目(出處)：	Synovial Sarcoma of the Tongue: Report of a Case and Review of the Literature. J Oral Maxillofac Surg 2008;66:154-160
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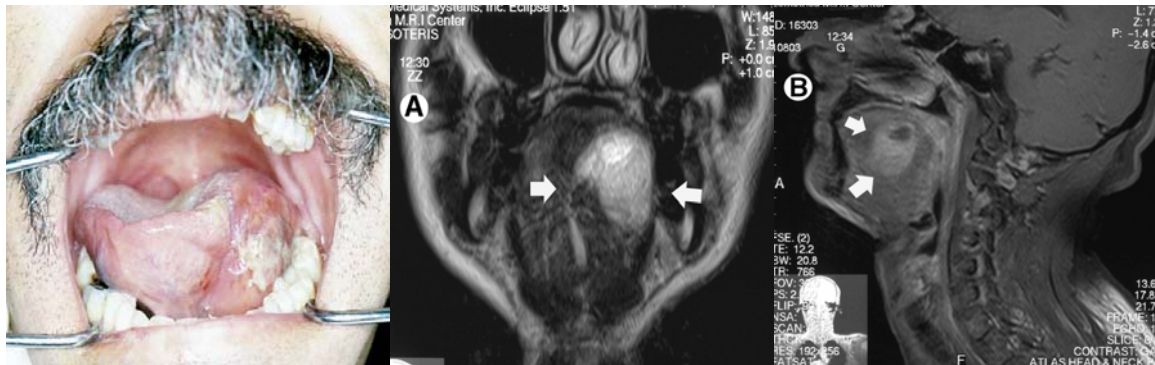
內文：

Abstract

1. Soft tissue tumors 包含不同種mesenchymal lesions，可由cytogenetic、molecular techniques的結果來分類。
2. Synovial tissue由development of cavities in pre-existing mesenchyme所組成。
3. Progenitor mesenchyme分化成2種組成：epithelioid cell 與spindle cell，這兩類cell被認為是separate entities，且被認為是interchangeable。
4. Synovial sarcoma (SS)約佔8%的soft tissue tumor且好發年齡在adulthood。
5. 超過90%的SS好發在四肢部位；起源於頭頸部位則佔3~10%，且最常發生部位是parapharyngeal，在口內則甚稀少。
6. 過去40年內，SS發生在tongue只有10 cases，非常稀少，因此值得報導。

Report of a Case

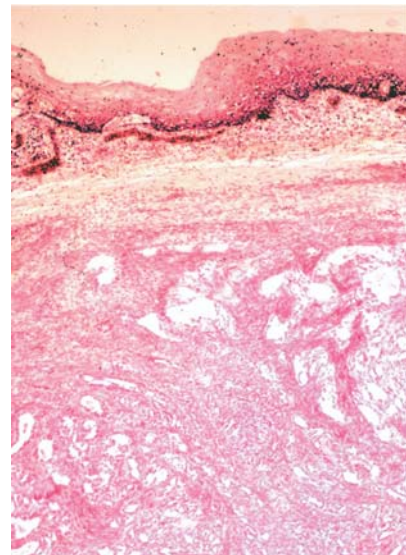
	General	Case
Age	Adulthood	49 y/o
predilection	Slight male predilection	Male
Site	Extremities (paravertebral、parapharyngeal area)	Tongue的左後1/3
size	Variable	3×4.5×4 cm
growth	Gradually enlarging mass	Rapidly
Duration		4 months
dysfunction	Dysphagia、dyspnea、hoarseness	Speech impairment food intake hardly
Progress	Poor	poor
Survival rate	36~64%(5 years) 20~38%(10 years)	
其他	Pain Tenderness Ref: Oral & Maxillofacial PATHOLOGY, 2 nd , p.488 Neville,Damm,Allen,Bouquot	Painless exophytic mass Ulcer with Bleeding Firm、bluish appearance No palpable lymph node of neck A(+); B(-); C(-)



<p>Clinical preoperative intraoral photograph of the tongue lesion at presentation</p>	<p>Coronal MRI (T2 mode). A well delineated, ovoid, circumscribed lesion inside the tongue can be depicted (<i>arrows</i>)</p>	<p>Lateral MRI (T1 mode). The lesion can be seen inside the musculature of the tongue occupying the middle and posterior third but not extending to the base of the tongue (<i>arrows</i>)</p>
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Lesion超過midline of the tongue，但無 cervical lymphadenopahty被發現。

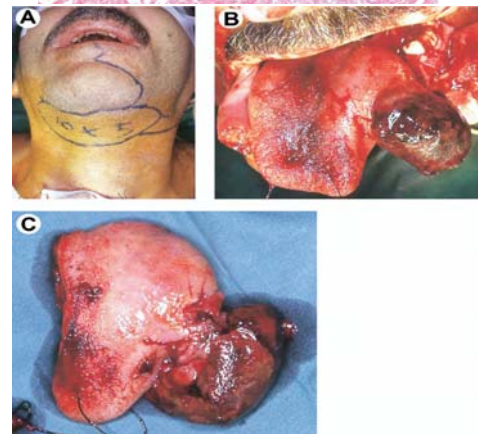
1. 在local anesthesia下做biopsy，得到組織學報告為synovial sarcoma。
2. Tumor包含2種不同形態細胞，形成typical biphasic pattern of epithelial cells with a surrounding spindle component。
3. No metastasis was found(CT scan of lung, bone scan, abdominal ultrasonography)。
4. Plan: hemiglossectomy with submental island skin flap, and no post-op CT、RT。
5. Surgery:
 - (1) Temporary tracheostomy was performed
 - (2) 10×5cm fasciocutaneous submental island flap was raised
 - (3) Median lipo split incision
 - (4) Left paramedian mandibular osteotomy
 - (5) Hemiglossectomy
 - (6) Submental island skin flap was transported



A, Incision lines of the midline split and the island skin flap.

B, Distortion of the tumor from the intralesional hemorrhage. A paramedian mandibular split has been performed.

C, The excised specimen of hemiglossectomy is shown



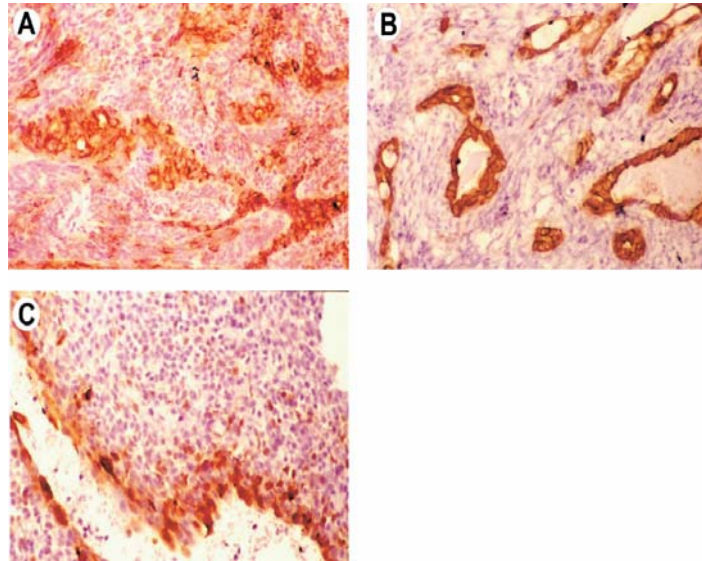
6. surgical specimen:
 - (1) Classic biphasic SS
 - (2) Dual differentiation into epithelial nest of cells and spindle cell collections surrounding the epithelial components

- (3) High and low molecular weight cytokeratin antigen and epithelial membrane antigen

A, The immunohistochemical stain with the epithelial membrane antigen shows strong positivity. (EMA stain; magnification ×250)

B, The immunohistochemical stain with high and low molecular weight cytokeratin. (AE1/AE3 stain; ×150)

C, Higher magnification of the same histologic slide (B). (×400)



7. 從右圖可知除了要定期口內刮皮膚外，整個外觀及功能非常完美。
8. post-op 18 months, patient覺得呼吸愈來愈不順，因而進行chest x-rays及CT scan，懷疑有lung metastases，但未確定，因為patient拒絕bronchoscopy檢查
9. 但病人用藥治療後逐漸好轉，且在術後2年內沒有症狀發生。



Discussion

- SS是由兩種形態相異，但組織來源相關的cell組成的biphasic pattern.
 - SS可分成下列四型，但最近亦有一種myxoid variant type 被描述
 - Biphasic type
 - Monophasic fibrous type
 - Monophasic epithelial type
 - Poorly differentiated (round cell)type

* Monophasic type，大部分由spindle組成，罕見來自epithelial cell
 - 舌部的epithelial-like cells排成glandular structures，而spindle cell存在其外部。
 - Epithelial-like cells對cytokeratins、epithelial-membrane antigen為positive。Spindle cells對vimentin、fibronectin為positive。
- Oral SS from 1962~2006年，共31 cases，有11(33%)位於tongue;在這11 cases中，有7個在base of the tongue.
5. SS在這11 cases中，有明顯偏向male，這與soft tissue sarcoma的1:1特點不同。

Table 1. CLINICOPATHOLOGIC CHARACTERISTICS OF SYNOVIAL SARCOMAS OF THE TONGUE

Study	Year	Age/ Gender	Location	Size (cm)	Treatment	Follow-Up (mos)	Distant Metastases	Outcome
Mir-Abedy ¹⁶	1962	23/F	Base of tongue	5	Surgery	36	NR	AND
Novonty and Fort ¹⁰	1971	20/M	Base of tongue	3.5 × 2	Surgery/Radiotherapy	72	NR	AND
Mousavi and Ghodsi ⁹	1974	40/M	Base of tongue	6 × 5 × 5	Surgery/Radiotherapy	4	NR	AND
Guzman et al ¹⁷	1975	21/M	Base of tongue	7	Surgery	12	Lungs	AWD
Engelhardt and Leafstedt ¹¹	1983	25/M	Base of tongue	NR	Surgery	NR	NR	NR
Smookler et al ¹⁵	1982	16/M	Mobile tongue	1.2	Surgery	96	NR	AND
Holtz and Magielski ⁸	1985	16/M	Base of tongue	6 × 5 × 4	Surgery/Radiotherapy/ Chemotherapy	37	Multiple sites	DOD
Bridge et al ¹²	1988	31/M	Base of tongue	3 × 3	Surgery	NR	NR	NR
Carrillo et al ⁷	1992	13/M	Middle third	1.1 × 5 × 5	Surgery	12	NR	AND
Fortuno-Mar et al ¹⁴	2000	26/M	Unspecified	NR	Surgery	NR	Lungs	AWD
Present case	2006	49/M	Middle third	5.8 × 4.5 × 2.8	Surgery	24	Lungs	AWD

NOTE: Data from Meer et al.¹³
Abbreviations: AND, alive no disease; AWD, alive with disease; DOD, died of disease; NR, not reported.

6. 31 cases的好發年紀

Age	range	Median age
ss in oral	10~50 y/o	
ss in all site	5~87 y/o	32 y/o

7. DD:

- (1) High grade SS: Ewing sarcoma, primitive neuroectodermal tumor 、 malignant peripheral nerve sheath tumor
 - (2) Predominant epithelioid SS: sarcomas 、 myoepitheliomas
 - (3) Biphasic type SS: no difficulty
 - (4) Monophasic epithelioid variants & poor differential SS: spindle cell sarcomas 、 round cell sarcomas 、 myoepitheliomas 、 epithelioid fibrosarcomas
8. Soft tissue sarcoma divide into 2 major genetic groups:
- (1) Sarcomas with specific genetic alterations and specific oncogenic mutations
 - (2) Sarcomas with nonspecific genetic alterations complex unbalanced karyotypes
9. SS 被認為(X;18)translocation所造成，這源於fusion of SYT on chromosome 18 & SSX1 、 SSX2 、 SSX4 on chromosome X 。
10. 95%的SS可由上述translocation gene鑑定，所以是相當可靠的診斷方法。
11. Survival of SS is poor且5-year survival rate為40~50% 。
12. 根據Carrillo et al ， SS in tongue是less aggressive ， 歸因於early detection, small size , young age of patients 。
13. Patient常有blood borne metastases ， 常會轉移到lung ， 這常常是致死的主因。 sarcoma罕見lymphatic spreading ， 因而SS沒有做elective neck dissection ， 除非有palpable nodes出現 。
14. Local recurrences常發生在術後2年或更晚 。
15. 雖然SS被認為high-grade sarcoma ， 但lesion的行為常因病人的因素而變化 。
16. Negative prognostic factors:
- (1) Large tumor size (>5cm)
 - (2) Tx in noncancer-specific hospital
 - (3) Patient age> 25y/o
 - (4) Poorly differentiated histologic appearance 。
17. Submental island flap在男性會有hair growth的問題 。
18. 若有進行radial neck dissection ， 則需進行radial forearm free flap ， 但這手術是從前臂取下一塊flap ， 因此需另外進行取flap的手術 。
19. Malignant mesenchymal tumors, especially sarcomas different SCC
- (1) SCC: spread into adjacent lymph node 30~40%
spread into adjacent lymph node 20~30%(with Neck Dissection)
 - (2) Sarcoma: metastases in distant sites
20. Post-op RT is still debatable issue. But RT seems to improve locoregional control of the disease.
21. SS is sensitive to chemotherapy, reduce the rates of local recurrences and distant metastases.
22. Chimeric protein produced by fusion of SYT-SSX genes成為未來具有 antigen-directed, individualized, antineoplastic的免疫治療目標 。

題號	題目
1	synovial sarcoma的classic histologic type為哪一型？ (A) Biphasic type (B) Monophasic fibrous type (C) Monophasic epithelial type (D) Poorly differentiated (round cell)type
答案 (A)	出處：Oral & Maxillofacial PATHOLOGY, 2 nd , p488 Neville,Damm,Allen,Bouquot
題號	題目
2	Synovial sarcoma的好發位置為何？ (A) Extremities (B) Parasvertebral area (C) Parapharyngeal area (D) Base of the tongue
答案 (A)	出處：Oral & Maxillofacial PATHOLOGY, 2 nd , p488 Neville,Damm,Allen,Bouquot