Head & Neck MM, Special case

Supraglottic Spindle Cell Sarcoma

Presenter R1吳仲升
Supervisor VS 羅武嘉

Patient’s profile

- Name:　　陳o菁
- Chart num: 1694280
- Age: 33y/o
- Gender: female
- BH:145cm BW:62kg　　BMI: 29.5 kg/m2　(Overweight)
- Social status: married
- A(-)B(-)C(-)

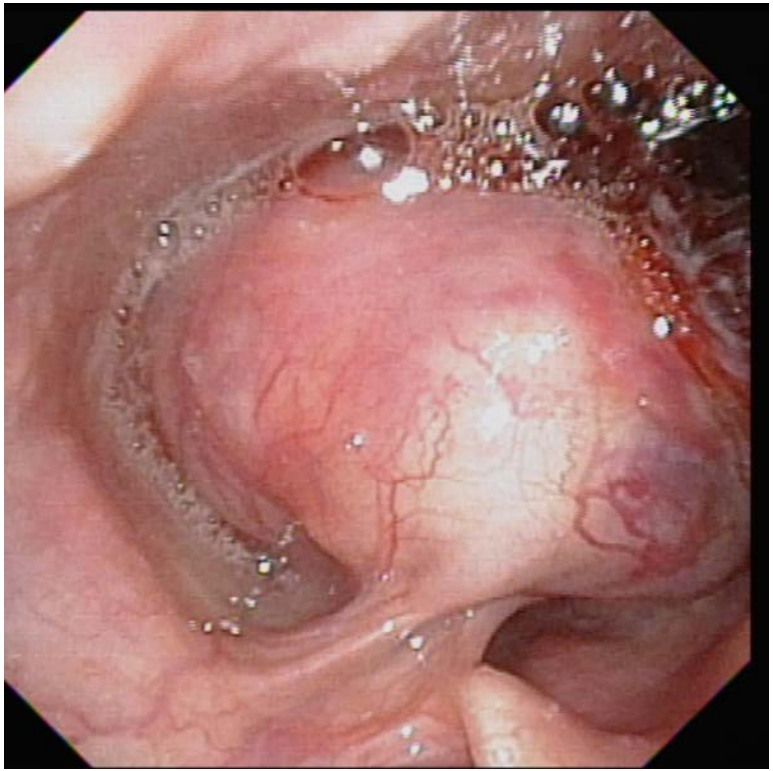
Personal history

- Important medical history
- Systemic disease: HTN(-), DM(-), CAD(-),asthma(-),HBV(-),HCV(-), other: nil
- Drug allergy: NKA
- Long-term medications: nil
- Operation history: nil

Present Illness

- 2025-02-07 OPD
  - Cough with blood for 1 day
  - hoarseness with dyspnea for 6 months

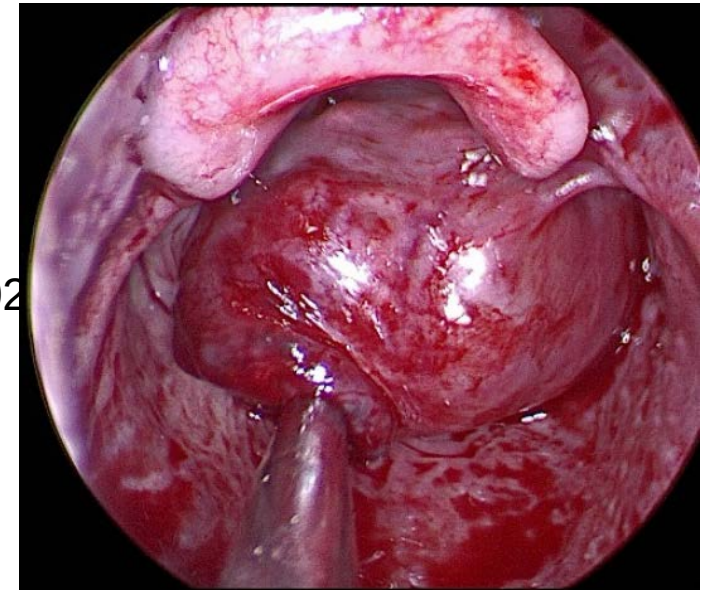
- Fiber: NP lymphoid tissue, huge mass over arytenoids with blood clot, bil VF unseen, airway compromised

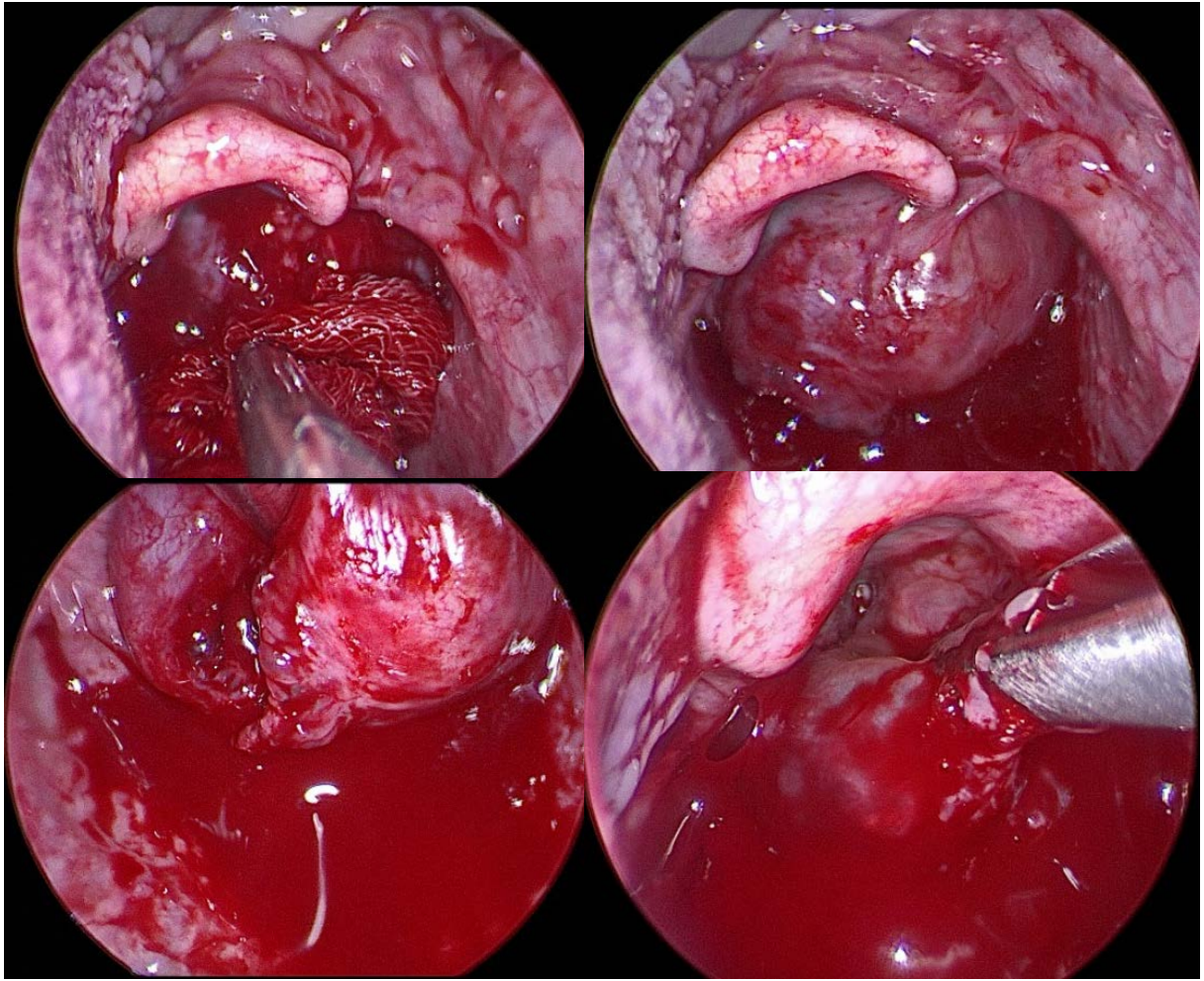


Present Illness

- 2025.02.07- 02.13 Admitted to ENT ward

- s/p emergent tracheostomy + LMS biopsy on 202
  02-07

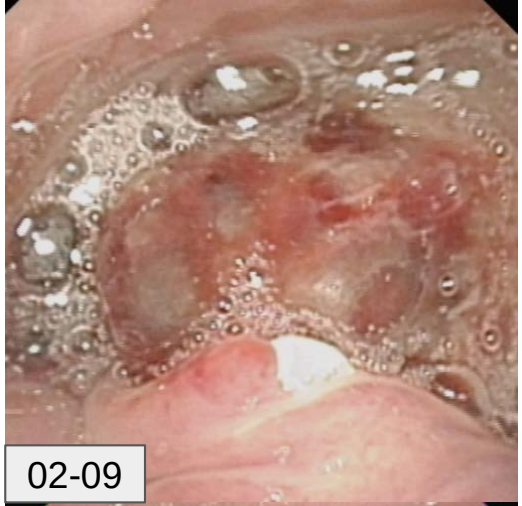


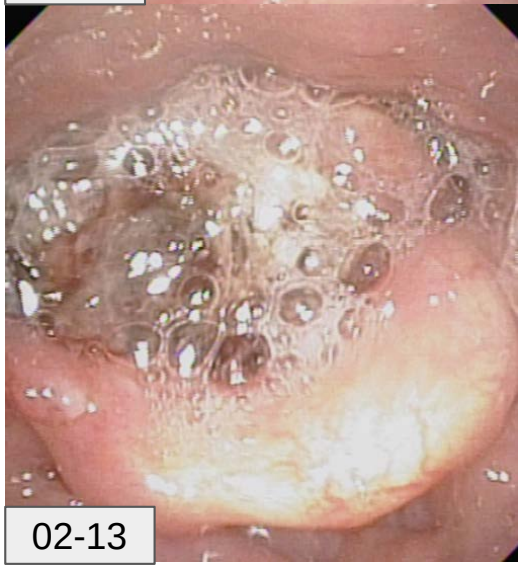


Present Illness

- 2025.02.07- 02.13 Admitted to ENT ward

- Patho:
- Larynx, supraglottis, right, LMS biopsy, spindle cell sarcoma





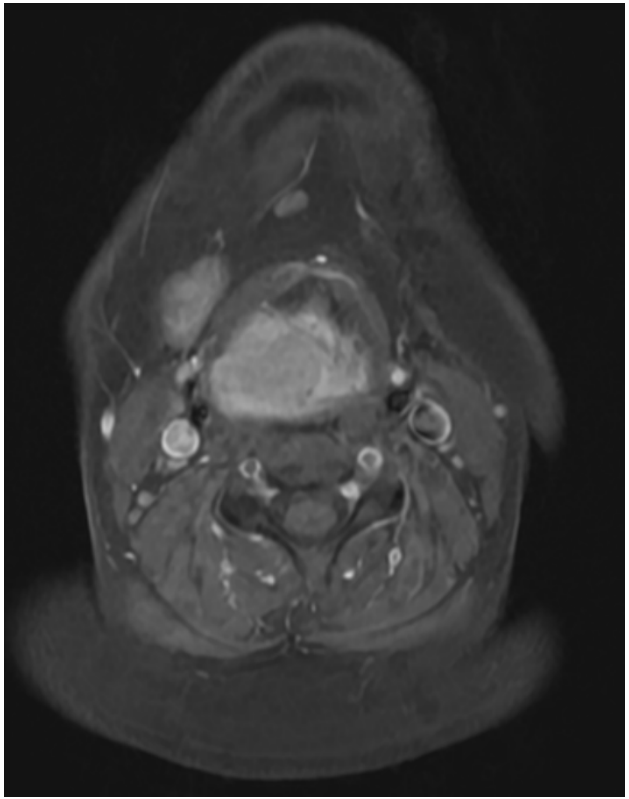
Pathology 2025-02-27

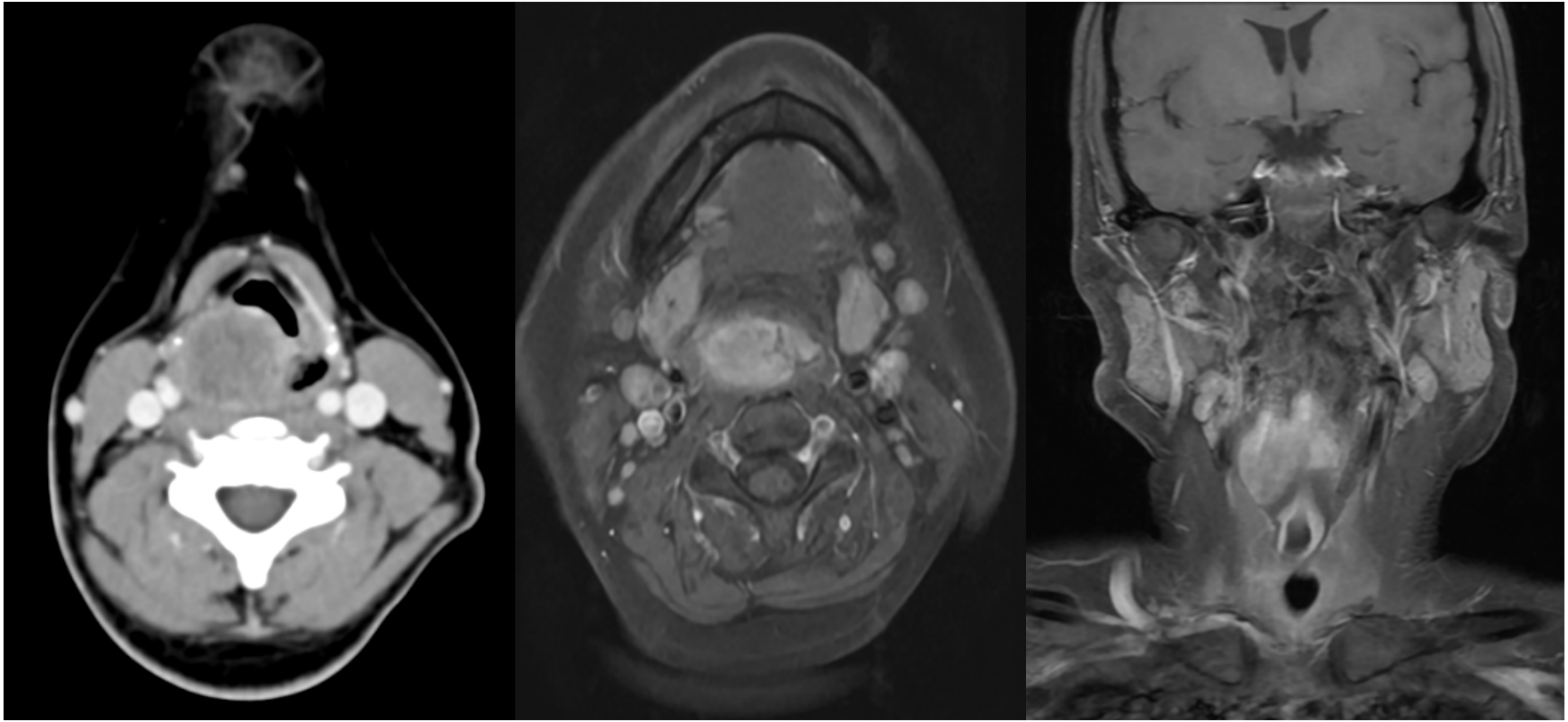
Microscopically, it shows laryngeal mucosa with unremarkable squamous epithelium and subepithelial proliferation of oval to short spindle tumor cells arranged in sheets or fascicles. The tumor cells exhibit fine chromatin, inconspicuous nucleoli, and frequent mitosis (10-15 mitosis/10 HPF). By immunohistochemistry, the tumor cells are CK (-), EMA (-), TLE1 (partial +), S100 (-), SOX10 (-), H3K27me3 (preserved), synaptophysin (+), chromogranin A (-), CD99 (partial +), WT-1 (cytoplasmic staining), ERG (-), Cyclin D1 (partial +), SATB2 (partial +), desmin (-), actin (-), myogenin (-), myoD1 (-), INI1 (preserved), CD34 (-), STAT6 (-), and MDM2 (-).

The **immunoprofile is inconclusive**. The tumor differentiation and classification cannot be determined. The diagnosis has been concurred in the intradepartment consensus meeting. The differential diagnoses include **synovial sarcoma, sarcoma with BCOR genetic alteration, CIC-rearranged sarcoma**, and other sarcomas.

Present Illness

- 2025-02-19 ~ 2025-02-24 Staging
- MRI:
  - prominent lymph nodes in **bilateral level IB and II**. An enlarged lymph node with mild central necrosis noted in right level II lymph nodes, suspect lymphadenopathy.
  - AJCC 8th edition Staging status:cT3N1M0
- Neck echo: left neck level IV small LN
- Fluoroscopic swallowing study:
  - Penetration-Aspiration Scale: **8**. Material enters the airway, passes below the vocal folds, and no effort is made to eject.
- CXR: no lung lesion noted





Present Illness

- PET CT
  - suspicious oropharyngeal or hypopharyngeal malignancy around **supraglottic area**, extending to the level of **tonsil**, and near **cricoid cartilage**. (consider TX)
  - consider metastatic lymph nodes in **right neck level IB, right level II**, and suspicious **left neck level III**. (consider NX)
  - suspicious reactive lymph nodes in **right interlobar region**.
  - suspicious chronic change or inflammation at infiltration in **RLL and LLL**.

Diagnosis

- Supraglottic spindle cell sarcoma, cT3N1M0

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Contents ⌄ Calcula

NCCN National Comprehensive Cancer Network®

NCCN Guidelines Version 4.2024
Soft Tissue Sarcoma

Received: 19 September 2018 | Revised: 15 January 2019 | Accepted: 29 January 2019

DOI: 10.1002/hed.25701

WILEY

ORIGINAL ARTICLE

Staging soft tissue sarcoma of the head and neck: Evaluation of the AJCC 8th edition revised T classifications

Justin M M. Cates MD, PhD

Cancer
An International Interdisciplinary Journal of the American Cancer Society

Original Article | Free Access

Analysis of prognostic factors in 146 patie
skull base sarcoma: An international colla

Ziv Gil MD, PhD, Snehal G. Patel MD, Bhuvanesh Singh MD, Giulio Cant
Luiz P. Kowalski MD, Dennis H. Kraus MD, Carl Snyderman MD, Jatin P. Shah MD ... See all authors

Home > Head and Neck Pathology > Article

Primary Sarcomas of the Larynx: A Single Institutional Experience with Ten Cases

Original Paper | Published: 07 December 2019

Volume 14, pages 707–714, (2020) Cite this article

Introduction to Sarcoma

- Sarcoma is a rare type of cancer that arises from connective tissues (e.g., muscles, fat, blood vessels, nerves, bones, and cartilage).

- It is classified into **Soft Tissue Sarcomas (STS)** and **Bone Sarcomas**.

- Represents approximately **1% of all adult cancers** but is more common in children.

Classification of Sarcoma

Soft Tissue Sarcomas (STS)

- **Undifferentiated/Unclassified Sarcoma**
- **Fibrosarcoma**
- **Liposarcoma**
- **Leiomyosarcoma**
- **Synovial Sarcoma**
- **Malignant Peripheral Nerve Sheath Tumor (MPNST)**
- **Rhabdomyosarcoma** (more common in children)
- **Angiosarcoma**
- **Dermatofibrosarcoma Protuberans (DFSP)**

Bone Sarcomas

- **Osteosarcoma**
- **Chondrosarcoma**
- **Ewing Sarcoma**
- **Chordoma**

Spindle Cells

- Spindle cells are elongated, fusiform-shaped cells commonly found in various tissues, particularly those derived from **mesenchymal origins**.
  - **Connective Tissue**
    - Fibroblasts
    - Myofibroblasts
  - **Muscle Tissue**
    - Smooth Muscle Cells
    - Skeletal Muscle Satellite Cells
  - **Nervous Tissue**
    - Schwann Cells
    - Glial Cells
  - **Vascular Tissue**
    - Endothelial Cells & Pericytes
  - **Bone and Cartilage**
    - Osteoblasts/Osteocytes
      Chondroblasts/Chondrocytes
  - **Synovial and Mesothelial Tissue**
    - Synovial Fibroblasts
    - Mesothelial Cells

Spindle Cell Sarcoma and Its Presence in Sarcoma Types

- **Spindle Cell Sarcoma** is a descriptive term for sarcomas characterized by spindle-shaped cells.
- Can be found in multiple sarcoma subtypes, including:
  - **Undifferentiated/Unclassified Sarcoma**
  - **Fibrosarcoma**
  - **Leiomyosarcoma**
  - **Synovial Sarcoma (Monophasic Type)**
  - **Malignant Peripheral Nerve Sheath Tumor (MPNST)**
  - **Bone Spindle Cell Sarcoma** (variant of osteosarcoma)

Pathology 2025-02-27

Microscopically, it shows laryngeal mucosa with unremarkable squamous epithelium and subepithelial proliferation of oval to short spindle tumor cells arranged in sheets or fascicles. The tumor cells exhibit fine chromatin, inconspicuous nucleoli, and frequent mitosis (10-15 mitosis/10 HPF). By immunohistochemistry, the tumor cells are CK (-), EMA (-), TLE1 (partial +), S100 (-), SOX10 (-), H3K27me3 (preserved), synaptophysin (+), chromogranin A (-), CD99 (partial +), WT-1 (cytoplasmic staining), ERG (-), Cyclin D1 (partial +), SATB2 (partial +), desmin (-), actin (-), myogenin (-), myoD1 (-), INI1 (preserved), CD34 (-), STAT6 (-), and MDM2 (-).

The **immunoprofile is inconclusive**. The tumor differentiation and classification cannot be determined. The diagnosis has been concurred in the intradepartment consensus meeting. The differential diagnoses include **synovial sarcoma, sarcoma with BCOR genetic alteration, CIC-rearranged sarcoma**, and other sarcomas.

Head and neck sarcomas **Epidemiology & Histologic Distribution**

- Head and neck sarcomas constitute **~2%** of all **head and neck** malignancies.
- **~1000 cases** diagnosed annually in the U.S.
- Key histologic subtypes:
  - **Children:** Rhabdomyosarcoma (RMS)
  - **Adults:** Osteosarcoma, chondrosarcoma, angiosarcoma, liposarcoma, leiomyosarcoma
  - **Rare types:** Solitary fibrous tumor, Ewing sarcoma, alveolar soft part sarcoma

Risk Factors

- **Radiation exposure** – Associated with secondary sarcomas.
- **Genetic predisposition** – Li-Fraumeni syndrome, NF1 (MPNST).
- **Environmental exposures** – Chemical carcinogens.

Clinical Presentation & Diagnosis

- **Common symptoms:**
  - Palpable mass, skin changes, subsite-specific symptoms **(hoarseness, dysphagia, epistaxis).**
- **Diagnostic workup:**
  - MRI (preferred for soft tissue assessment).
  - CT (useful for bone involvement, surgical planning).
  - **Biopsy** planning crucial for definitive surgery.
  - Metastatic evaluation with **chest CT, PET** scan if indicated.

Staging Systems

- **Head and Neck Soft Tissue Sarcomas: TNM staging (AJCC 8th Edition)**.
- **Key Prognostic Factors:**
  - Tumor size, grade, surgical margins, nodal involvement.

Soft tissue sarcoma of the head and neck TNM staging AJCC UICC 8th edition*

Primary tumor (T)	
T category	**T criteria**
TX	Primary tumor cannot be assessed
T1	Tumor ≤2 cm
T2	Tumor >2 to ≤4 cm
T3	Tumor >4 cm
T4	Tumor with invasion of adjoining structures
T4a	Tumor with orbital invasion, skull base/dural invasion, invasion of central compartment viscera, involvement of facial skeleton, or invasion of pterygoid muscles
T4b	Tumor with brain parenchymal invasion, carotid artery encasement, prevertebral muscle invasion, or central nervous system involvement via perineural spread
Regional lymph nodes (N)	
N category	**N criteria**
N0	No regional lymph node metastases or unknown lymph node status
N1	Regional lymph node metastasis
Distant metastasis (M)	
M category	**M criteria**
M0	No distant metastasis
M1	Distant metastasis

TNM: tumor, node, metastasis; AJCC: American Joint Committee on Cancer; UICC: Union for International Cancer Control.

* This is a new classification that needs data collection before defining a prognostic stage grouping.

G	**Definition of Grade** **FNCLCC Histologic Grade - see Histologic Grade (G)**
GX	Grade cannot be assessed
G1	Total differentiation, mitotic count and necrosis score of 2 or 3
G2	Total differentiation, mitotic count and necrosis score of 4 or 5
G3	Total differentiation, mitotic count and necrosis score of 6, 7, or 8

Anatomic Stage/Prognostic Groups

This is a new classification that needs data collection before defining a stage grouping for head and neck sarcomas.

Natural History & Prognostic Factors

- High local recurrence rates compared to extremity sarcomas.
- **Key determinants of prognosis:**
  - **Tumor grade** and **size**.
  - Negative surgical **margins**.
  - **Histologic subtype** and **lymph node** status.
  - Positive/close surgical margins significantly worsen survival.

Received: 19 September 2018 | Revised: 15 January 2019 | Accepted: 29 January 2019

DOI: 10.1002/hed.25701

WILEY

ORIGINAL ARTICLE

Staging soft tissue sarcoma of the head and neck: Evaluation of the AJCC 8th edition revised T classifications

Justin M.M. Cates MD, PhD

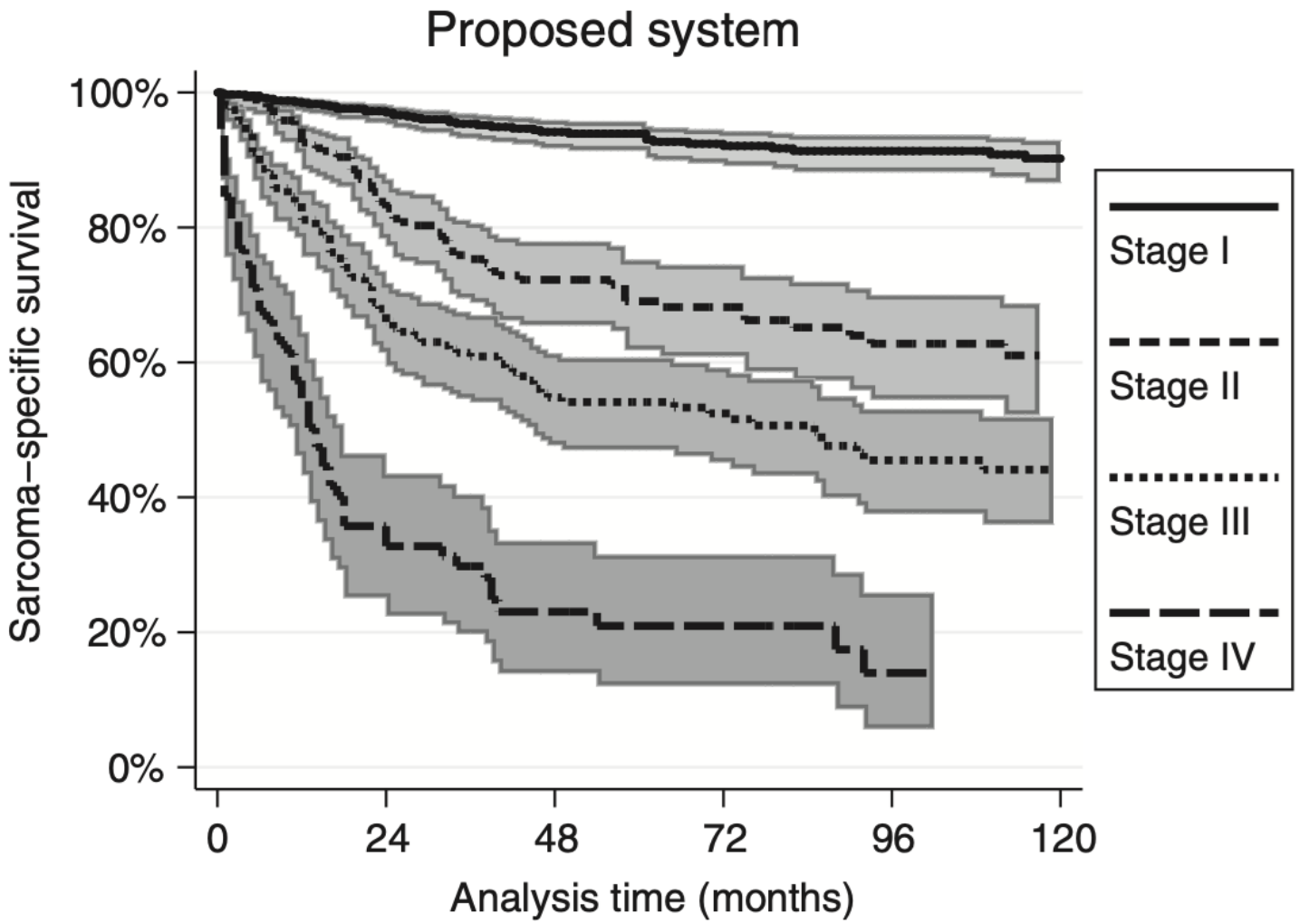
Head & Neck
IF: 2.4
Rank: Q2
USA

Summary

- Evaluate the **predictive ability** of revised **AJCC 8th Edition T classifications.**
- Develop and validate a **new staging algorithm** for head and neck sarcomas.
- **Data Source:** SEER Database (**N = 2756 patients**)
- **Analysis:** Nonlinear multivariable regression
- **Validation:** Separate dataset for comparison of staging systems
- **Outcome Measure:** Sarcoma-specific survival prediction
- **Tumor size categorization** is an informative predictor of sarcoma-specific mortality.
- The **proposed staging system** enhances risk stratification.

TABLE 4 Proposed staging system for soft tissue sarcoma of the head and neck based on revised AJCC 8th edition T classifications

Stage group	GTNM classification	No. of cases (%)	HR (95% CI)	*P*
Stage I	G1 Tx N0 M0	619 (46%)	1.00	NA
	G2 T1 N0 M0			
	G2 T2 N0 M0			
	G3 T1 N0 M0			
Stage II	G2 T3 N0 M0	301 (22%)	3.12 (2.27-4.28)	<0.0005
	G2 T4 N0 M0			
	G3 T2 N0 M0			
Stage III	G3 T3 N0 M0	304 (22%)	6.36 (4.78-8.48)	<0.0005
	G3 T4 N0 M0			
	Gx Tx N1 M0			
Stage IV	Gx Tx Nx M1	129 (10%)	18.7 (13.6-25.9)	<0.0005


Proposed system
Sarcoma−specific survival
100%
80%
60%
40%
20%
0%
0
24
48
72
96
120
Analysis time (months)
Stage I
Stage II
Stage III
Stage IV

Surgical Margins & Prognosis

- **Clear margins difficult due to anatomical constraints**

Cancer
An International Interdisciplinary Journal of the American Cancer Society

Original Article | Free Access

Analysis of prognostic factors in 146 patients with anterior skull base sarcoma: An international collaborative study†

Ziv Gil MD, PhD, Snehal G. Patel MD, Bhuvanesh Singh MD, Giulio Cantu MD, Dan M. Fliss MD, Dennis H. Kraus MD, Carl Snyderman MD, Jatin P. Shah MD ... See all authors

Cancer(2007)
IF: 6.1
Rank: Q1
USA

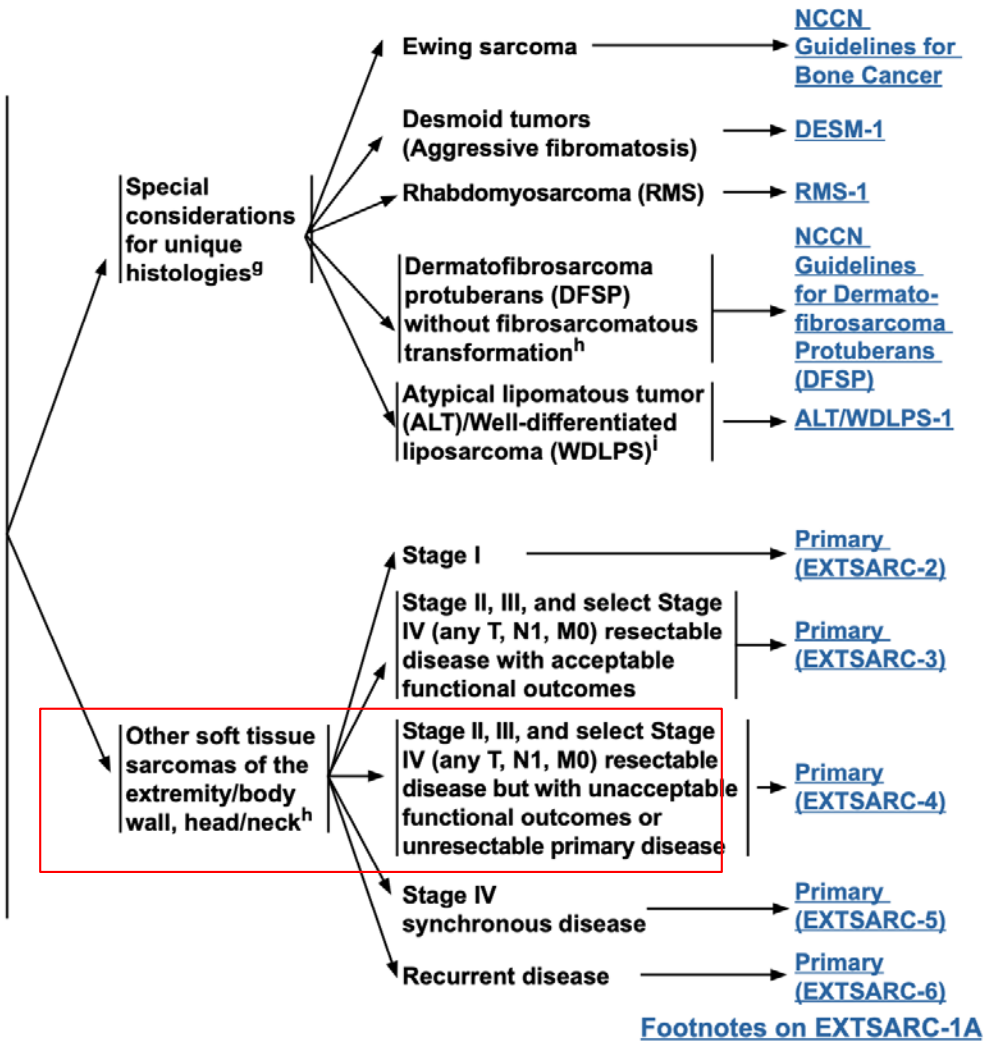
Surgical Margins & Prognosis

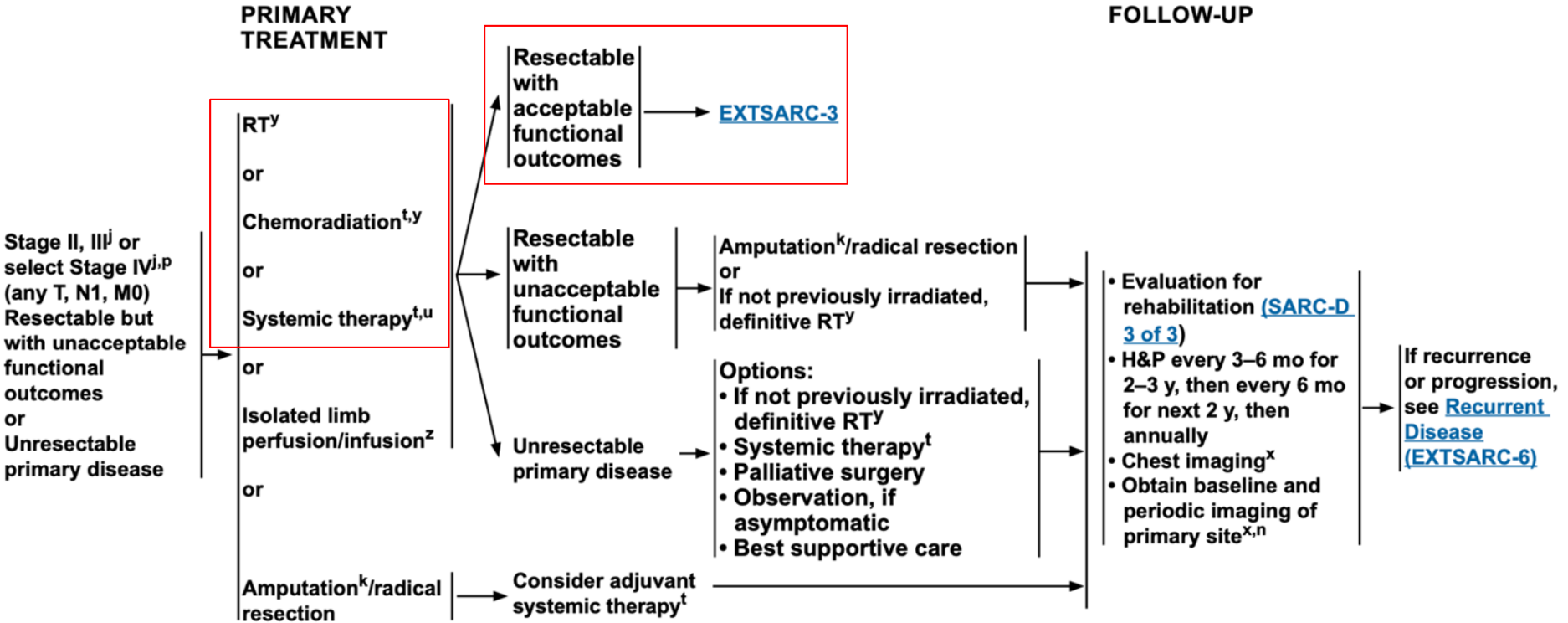
- **International collaborative study group**
- **N= 146, skull base sarcoma, aged > =21 y/o(77%)**
- **Adjuvant RT: 35% of patients, chemotherapy: 10% of patients.**
- **5-year survival rates by margin status:**
  - **Negative margins:** 77%
  - **Close margins (<1 mm):** 43%
  - **Positive margins:** 36%
- Positive margins = **only independent predictor of poor survival**

General Treatment Principles

- **Surgery** – Aim for complete resection with negative margins.
- **Radiation Therapy (RT):**
  - Adjuvant RT recommended for **high-grade** tumors or **positive margins**.
- **Chemotherapy:**
  - Used for specific subtypes (e.g., osteosarcoma, RMS).
  - Role in adult sarcomas remains **controversial**.

NCCN guideline




PRIMARY TREATMENT
FOLLOW-UP
Stage II, III[j] or select Stage IV[j,p] (any T, N1, M0) Resectable but with unacceptable functional outcomes or Unresectable primary disease
RT[y]
or
Chemoradiation[t,y]
or
Systemic therapy[t,u]
or
Isolated limb perfusion/infusion[z]
or
Amputation[k]/radical resection
Resectable with acceptable functional outcomes
EXTSARC-3
Resectable with unacceptable functional outcomes
Amputation[k]/radical resection or If not previously irradiated, definitive RT[y]
Unresectable primary disease
Options:
• If not previously irradiated, definitive RT[y]
• Systemic therapy[t]
• Palliative surgery
• Observation, if asymptomatic
• Best supportive care
Consider adjuvant systemic therapy[t]
• Evaluation for rehabilitation (SARC-D 3 of 3)
• H&P every 3–6 mo for 2–3 y, then every 6 mo for next 2 y, then annually
• Chest imaging[x]
• Obtain baseline and periodic imaging of primary site[x,n]
If recurrence or progression, see Recurrent Disease (EXTSARC-6)

Radiation Therapy Guidelines for Soft Tissue Sarcoma of Extremity/Body Wall/Head and Neck[1,2]

Neoadjuvant RT:

- The panel has expressed a general preference for preoperative over postoperative radiotherapy.[3]
- Potential benefits of preoperative radiation include lower total radiation dose, shorter treatment time, smaller field sizes, potential for reduced late toxicities (ie, fibrosis, edema, joint stiffness), and potential for tumor downstaging. Potential disadvantages include increased frequency of early wound healing complications, particularly for lower extremity tumors.[4,5]
- A preoperative dose of 50 to 50.4 Gy external beam RT (EBRT) (1.8–2 Gy per fraction) is recommended.[6-9]
- Radiation should be delivered using the most appropriate technique that will cover the target volume while maintaining dose constraints to normal tissues (which may include bone, lymphatics, and soft tissue). These techniques may include electron beam therapy, 3D conformal radiation therapy, IMRT, or (in challenging anatomic locations) proton beam therapy delivered with image guidance.[10,11]
- If an R1 or R2 resection is anticipated, placement of clips in areas at high risk for recurrence is encouraged.
- For positive margins after preoperative radiation and surgery, consider observation or RT boost in select situations.
  - There are data to suggest that some patients with positive margins following neoadjuvant RT such as those with low-grade WDLPS and a focally, "planned" positive margin on an anatomically fixed critical structure may have acceptable local control without a boost.[12]
  - There are also data to suggest that delivery of a boost for positive margins does not improve local control. Since delivery of an adjuvant RT boost does not clearly add benefit, the decision should be individualized and the potential toxicities should be carefully considered.[13,14]
  - If adjuvant boost radiation for a positive margin is felt to be appropriate, an additional 14–20 Gy can be considered with fractionated EBRT or brachytherapy.[15]
- A dose reduction to 36 Gy for myxoid liposarcoma can be considered.[16]
- Conventionally fractionated RT remains the standard of care. A moderately hypofractionated preoperative regimen has demonstrated acceptable early local control, but long-term local control and toxicity data are not yet available. A hypofractionated regimen may be appropriate for select patients who might otherwise be unable to receive a standard course of preoperative radiotherapy.[17]

SYSTEMIC THERAPY AGENTS AND REGIMENS WITH ACTIVITY IN SOFT TISSUE SARCOMA SUBTYPES[a,b,c,d] AND AGGRESSIVE SOFT TISSUE NEOPLASMS

Regimens Appropriate for General Soft Tissue Sarcoma[e,f]; see other sections for histology-specific recommendations[g]

	Preferred Regimens	Other Recommended Regimens	Useful in Certain Circumstances
Neoadjuvant/ Adjuvant Therapy	• AIM (doxorubicin, ifosfamide, mesna)[1-4] • Ifosfamide, epirubicin, mesna[5]	• AD[1,2,10,11] for LMS, or if ifosfamide is not considered appropriate • Doxorubicin[1,2,6,7]	• Ifosfamide[5,7,21-25] • Trabectedin (for myxoid liposarcoma)[30] • Gemcitabine and docetaxel[21,22] (category 2B)
First-Line Therapy Advanced/ Metastatic	• Anthracycline-based regimens: ▸ Doxorubicin[1,2,6,7] ▸ Epirubicin[8] ▸ Liposomal doxorubicin[9] ▸ AD (doxorubicin, dacarbazine)[1,2,10,11,12] ▸ AIM[1-4,6] ▸ Ifosfamide, epirubicin, mesna[5] • *NTRK* gene fusion-positive sarcomas only (regardless of soft tissue sarcoma subtype) ▸ Larotrectinib[h,13] ▸ Entrectinib[i,14] ▸ Repotrectinib[15]	• Gemcitabine • Gemcitabine and docetaxel[21,22] (category 2B)	• Pazopanib[k,16] (patients ineligible for IV systemic therapy or patients who are not candidates for anthracycline-based regimens) • MAID (mesna, doxorubicin, ifosfamide, dacarbazine)[1,2,31,32] • Trabectedin and doxorubicin (for LMS)[33,34] • Selpercatinib (for *RET* gene fusion-positive tumors)[35] (regardless of soft tissue sarcoma subtype) • Gemcitabine and dacarbazine[23] (category 2B)
Subsequent Lines of Therapy for Advanced/ Metastatic Disease	• Pazopanib[j,k,16] • Eribulin[j,17] (category 1) recommendation for liposarcoma, category 2A for other subtypes • Trabectedin[j,18-20] (category 1 recommendation for liposarcoma and LMS, category 2A for other subtypes) • Gemcitabine and docetaxel[21,22] • *NTRK* gene fusion-positive sarcomas only (regardless of soft tissue sarcoma subtype) ▸ Repotrectinib[15] (if not previously given)	• Dacarbazine[23] • Ifosfamide[5,7,22,24,25,26] • Temozolomide[j,27] • Vinorelbine[j,28] • Regorafenib[k,29] • Gemcitabine • Gemcitabine and dacarbazine[23]	• Gemcitabine and vinorelbine[24] (category 2B) • Gemcitabine and pazopanib[36] (category 2B) • Pembrolizumab[37,38] or nivolumab ± ipilimumab[39-42] ▸ For myxofibrosarcoma, UPS,[f] dedifferentiated liposarcoma, cutaneous angiosarcoma, and undifferentiated sarcomas OR ▸ For TMB-H (≥10 mutations/megabase [mut/Mb])[l] regardless of soft tissue sarcoma subtype • Pembrolizumab[43] ▸ For MSI-H or dMMR tumors[m] (regardless of soft tissue sarcoma subtype) • Cabozantinib[44] (category 2B) • Afamitresgene autoleucel[117] ▸ HLA-A*02:01P, HLA-A*02:02P, HLA-A*02:03P or HLA-A*02:06P positive and whose tumor expresses the MAGE-A4 antigen (synovial sarcomas only)

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Primary Sarcomas of the Larynx: A Single Institutional Experience with Ten Cases

Original Paper | Published: 07 December 2019

Volume 14, pages 707–714, (2020) Cite this article

Head & Neck pathology
IF: 3.2
Rank: Q1
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Summary

- **Total cases:** 10 (9 males, 1 female; mean age: 59 years).
- **Histologic subtypes:**
  - **Chondrosarcoma (*4), Osteosarcoma (*1), Embryonal rhabdomyosarcoma (*1)**
  - **Undifferentiated spindle cell sarcoma (*1), Well-differentiated liposarcoma (*1)**
  - **Kaposi sarcoma (*1 case), Synovial sarcoma (*1)**
- All patients **presented with vocal and/or respiratory symptoms**.
- **Surgical excision** was the primary treatment in all but **Kaposi sarcoma**.
- **Local recurrence** was observed in **2 chondrosarcoma cases**.
- **No distant metastases or disease-related deaths** were reported.

Patient #	Diagnosis	Age (years)/sex	Size (cm)	Site of tumor Epicenter	Symptoms	Treatment (margins)	Recurrence/ metastasis (months)	Status & total follow-up (years)
1	Chondrosarcoma, grade I	71/M	2.5	L Arytenoid	Hoarseness, dysphagia	Total laryngectomy (−)	N/N	DOC (3)
2	Chondrosarcoma, grade I	65/F	4.6	R Arytenoid	Hoarseness, dysphagia	Fragmented local excision (N/A); Laryngectomy (−)	Y(6)N/N	NED (12)
3	Chondrosarcoma, grade II	54/M	3.5	Cricoid	Hoarseness, discomfort	Partial laryngectomy (+); Total laryngectomy (−)	Y(24)Y(15)/N	AWD (7)
4	Chondrosarcoma, grade I	58/M	2.6	Subglottis	Stridor	Partial laryngectomy (−)	N/N	NED (2)
5	Osteosarcoma, high grade	75/M	2.1	Anterior commisure	Hoarseness	Laryngectomy (−)	N/N	LOST
6	Embryonal rhabdomyosarcoma	61/M	<1	R Arytenoid	Dyspnea, stridor	Fragmented local excisions x2 (N/A), CT, RADx	N/N	NED (1)
7	Well-differentiated liposarcoma	36/M	2.4	Epiglottis/vallecula	Sleep apnea, dysphagia	Partial laryngectomy (−), RADx	N/N	NED (1)
8	Kaposi Sarcoma	34/M	N/A	Epiglottis, aryepiglottic folds	Vocal changes	CT	–	NED (9)
9	Synovial sarcoma, FNCLCC grade 3	69/M	6.8	Vallecula	Cough	Partial laryngectomy (−), RADx	N/N	LOST
10	Undifferentiated spindle cell sarcoma, FNCLCC grade 3	68/M	2.5	Subglottis posterior	Dyspnea, stridor, hemoptysis	Laryngectomy (−); RADx	N/N	LOST (0.25)

Home › Head and Neck Pathology › Article

Primary Sarcomas of the Larynx: A Clinicopathologic Study of 27 Cases

Original Paper | Published: 08 March 2021

Volume 15, pages 905–916, (2021) Cite this article

Head & Neck pathology
IF: 3.2
Rank: Q1
USA
University of Miami Miller School of Medicine

- **Total cases:** 27 (25 males, 2 females; mean age: 60 years, range: 33–85).
- **Histologic subtypes:**
  - **Conventional chondrosarcoma (16), Well-differentiated liposarcoma (2)**
  - **Clear cell chondrosarcoma (1), Leiomyosarcoma (2)**
  - **High-grade myxofibrosarcoma (2), High-grade myofibroblastic sarcoma (1)**
  - **Low-grade myofibroblastic sarcoma (1), Malignant granular cell tumor (1)**
  - **Kaposi sarcoma (1)**
- **Local recurrence (3), Metastases (2).**
- **Other followed patients remained disease-free.**
- **Surgical removal remains the mainstay of treatment.**
- **Extent of surgery depends on tumor type and grade.**
- **Adjuvant therapy (neoadjuvant or post-surgical) is reserved for high-grade sarcomas.**

Table 1 Clinicopathologic features of laryngeal sarcomas

Case	Diagnosis	Age (years)/ Gender	Symptoms	Size (cm)	Grade	Site	Recurrence (months)	Treatment/Margin status	Status & Total follow up (months)
1	Myxofibrosarcoma	62/M	Stridor and shortness of breath	4.0	3	Vocal cords, bilateral	No	Total laryngectomy and bilateral neck dissection/Negative margins	LOST; 1 month
2	Leiomyosarcoma	65/M	Difficulty swallowing	5.3	2	Cricoid cartilage	Yes; (13 months after resection)	Total laryngectomy, partial pharyngectomy, and partial thyroidectomy/Negative margins	DOD; 48 months
3	Conventional chondrosarcoma	33/M	Voice changes	2.5	1	Cricoid cartilage	No	Segmental wide resection of the cricoid/ Negative margins	NED; 40 months
4	Myofibroblastic sarcoma	79/M	Cough and voice changes, hoarseness, fatigue, and fainting episodes	1.2	3	Left vocal cord	No	Left partial laryngectomy/Negative margins	LOST; 24 months with NED
5	Conventional chondrosarcoma, hyaline and myxoid type	85/M	Voice changes	5.5	2	Left thyroid and cricoid cartilage	No	Total laryngectomy/ radical resection of soft tissue tumor/left hemithyroidectomy/ Negative margins	NED; 24 months
6	Well-differentiated liposarcoma, lipoma-like	54/M	Progressive shortness of breath	5.1	1	Left epiglottis	No	Left partial laryngectomy with limited pharyngectomy/Negative margins	LOST; 12 months with NED
7	Conventional chondrosarcoma, arising from chondroma	52/M	Hoarseness	2.5	1	Left cricoid cartilage	No	Left hemilaryngectomy/ Negative margins	LOST; 1 month
8	Conventional chondrosarcoma	72/M	Left vocal cord paralysis	4.1	1	Left posterior thyroid cartilage	NA	NA	NA
9	Conventional chondrosarcoma, hyaline type	50/M	Dysphonia	NA	1	Cricoid cartilage	NA	NA	NA
10	Conventional chondrosarcoma, hyaline type	48/M	Hoarseness	2.0	2	Subglottis	NA	NA	NA
11	Conventional chondrosarcoma, hyaline type, arising from a chondroma	69/M	NA	NA	1	Larynx	NA	NA	NA
12	Conventional chondrosarcoma, hyaline type	50/M	Voice changes	2.8	2	Left cricoid cartilage	No	Left hemicricoid resection/Negative margins	NED; 18 months

Table 1 (continued)

Case	Diagnosis	Age (years)/ Gender	Symptoms	Size (cm)	Grade	Site	Recurrence (months)	Treatment/Margin status	Status & Total follow up (months)
13	Kaposi sarcoma	44/M	Voice changes and hemoptysis	1.5	–	Left false vocal cord	No	Six cycles of chemotherapy and highly active antiretroviral therapy	NED; 24 months
14	Conventional chondrosarcoma	75/M	Dysphagia	6.0	1	Cricoid cartilage	No	Total laryngectomy/ Negative margins	NED; 79 months
15	Clear cell chondrosarcoma, hyaline type	72/M	Voice changes	3.2	2	Thyroid cartilage	Yes; 204 months	Partial laryngectomy × 2/ Positive margins	NED; 90 months
16	Well-differentiated liposarcoma, mixed lipoma, and sclerosing type	71/M	Progressive shortness of breath	3	1	Left supraglottis	Yes;4 times (1995, 2002, 2004, and 2013)	Supraglottic laryngectomy/Positive margins	LOST; 1 month
17	Low-grade myofibroblastic sarcoma	69/F	Dysphonia and globus sensation	1.6	1	Left false and true vocal cords	No	Supracricoid partial laryngectomy/Negative margins	NED; 12 months
18	Conventional chondrosarcoma, hyaline type	45/M	Hoarseness	NA	1	Left larynx	NA	NA	NA
19	Conventional chondrosarcoma, hyaline type	65/M	Hoarseness	NA	1	Cricoid cartilage, posterior aspect	NA	NA	NA
20	Conventional chondrosarcoma, hyaline type	42/M	Hoarseness, dysphonia	1.3	1	Left arytenoid cartilage	NA	NA	NA
21	Conventional chondrosarcoma, hyaline type	49/M	Progressive dysphagia, hoarseness	NA	2	Left posterior cricoid	NA	Partial excision/subtotal tumor removal/ Margin status NA	NA
22	Conventional chondrosarcoma, mixed hyaline and myxoid type, arising from chondroma	68/M	Hoarseness	4.0	1	Right cricoid cartilage	NA	Left partial cricoidectomy/Positive peripheral margins	NED; 1 month
23	Conventional chondrosarcoma, myxoid and hyaline type, arising from chondroma	72/M	NA	3.5	2	Cricoid cartilage	No	Total laryngectomy/ Negative margins	DOC; 116 months
24	Malignant granular cell tumor	38/F	Dysphagia and odynophagia	6.8	2	Left lateral pharynx, pyriform sinus and left aryepiglottic fold	NA	Left partial pharyngectomy and partial vertical laryngectomy/ Positive margin	NED; 14 months
25	Conventional chondrosarcoma, hyaline type	48/M	Shortness of breath	2.9	1	Left thyroid cartilage	NA	NA	NA

Table 1 (continued)

Case	Diagnosis	Age (years)/ Gender	Symptoms	Size (cm)	Grade	Site	Recurrence (months)	Treatment/Margin status	Status & Total follow up (months)
26	Leiomyosarcoma	81/M	Shortness of breath	1.5	2	Left arytenoid	NA	NA	NA
27	Myxofibrosarcoma	78/M	Shortness of breath	4.0	3	Right vocal fold	NA	NA	NA

LOST Lost to follow up; *DOD* Died of disease; *NED* No evidence of disease; *AWD* Alive with disease; *DOC* Died of other causes; *NA* Not available

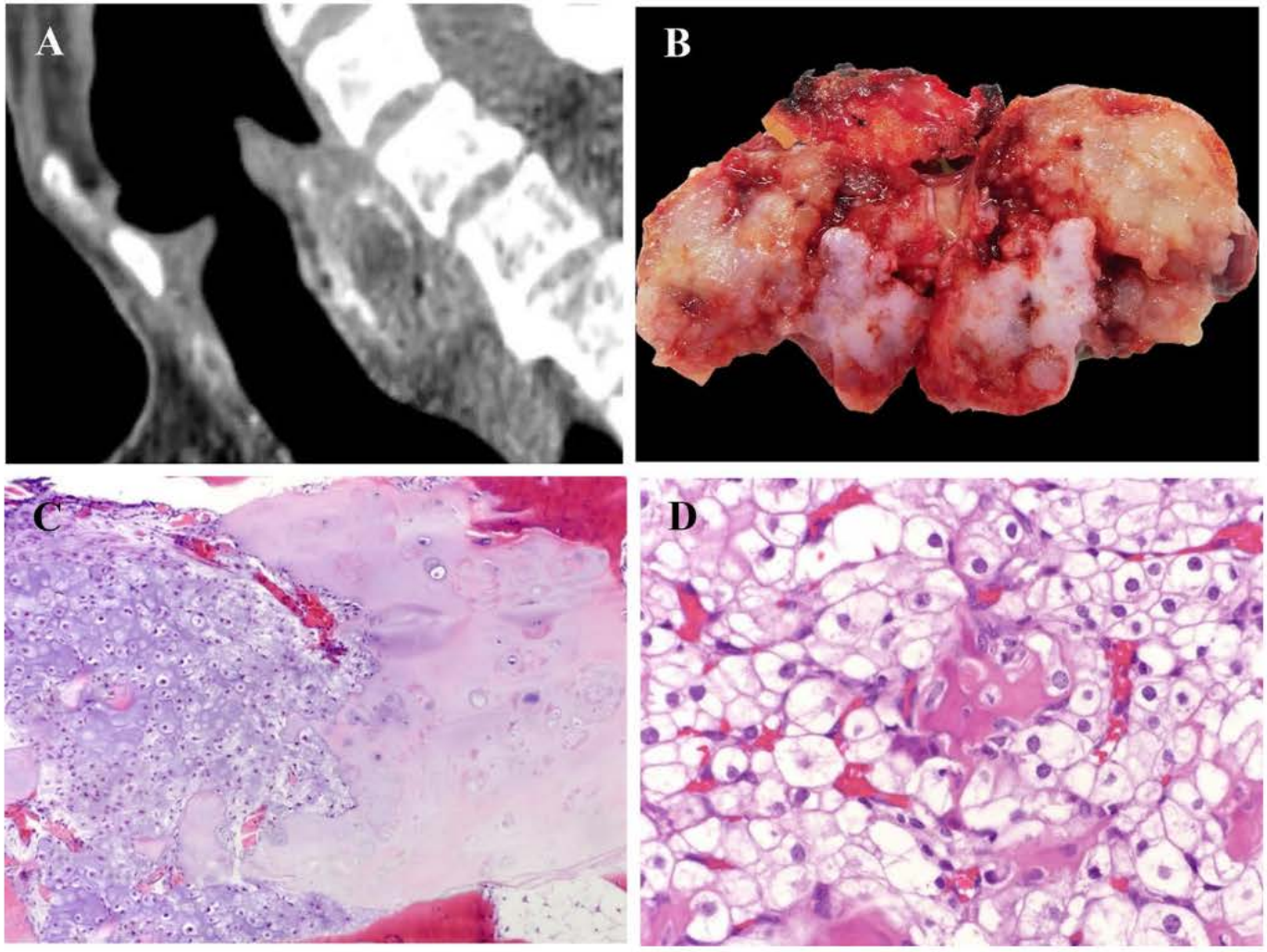

Fig. 1 **a** Reformatted sagittal computed tomography image of the larynx demonstrates a focally calcified mass located posteriorly, causing stenosis of the lumen. **b** Excised chondrosarcoma is solid, lobular, and has an area that is blue-white and regions that are glistening pale tan-yellow. **c** Low-grade chondrosarcoma arising from chondroma. The chondrosarcoma shows increased cellularity and mild nuclear atypia juxtaposed to the chondroma that is less cellular and lacks atypia. (HES × 10). **d** Clear cell chondrosarcoma. Sheets of large polygonal tumor cells with abundant clear to pale eosinophilic cytoplasm closely admixed with trabeculae of metaplastic woven bone focally lined by osteoblasts (HES × 40)

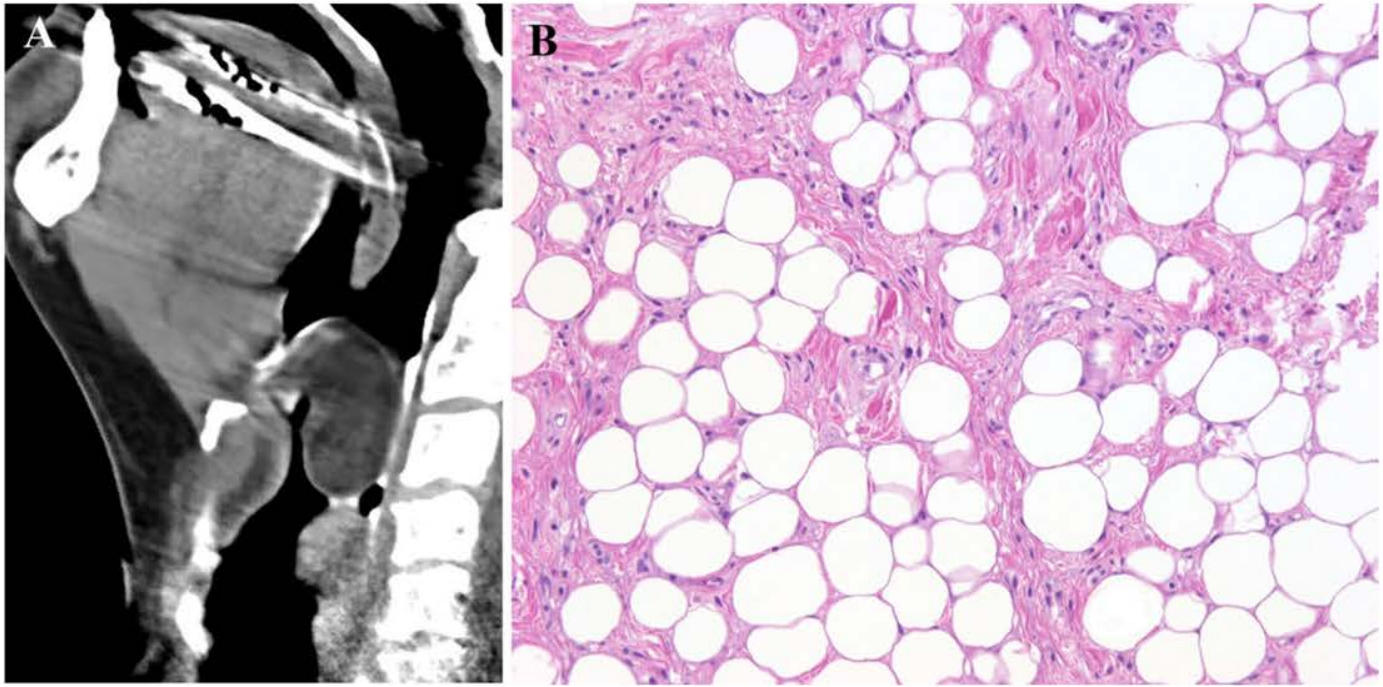

Fig. 2 **a** Reformatted sagittal computed tomography. Fat density mass seen extends from the inferior oropharynx to the larynx just above the level of the false vocal cords. **b** Well differentiated liposarcoma is composed of lobules of white adipocytes that vary in size, and scattered cells had enlarged hyperchromatic nuclei. The septae are thick, collagenous, and also contain spindle cells, some of which had enlarged hyperchromatic nuclei (HES × 20)

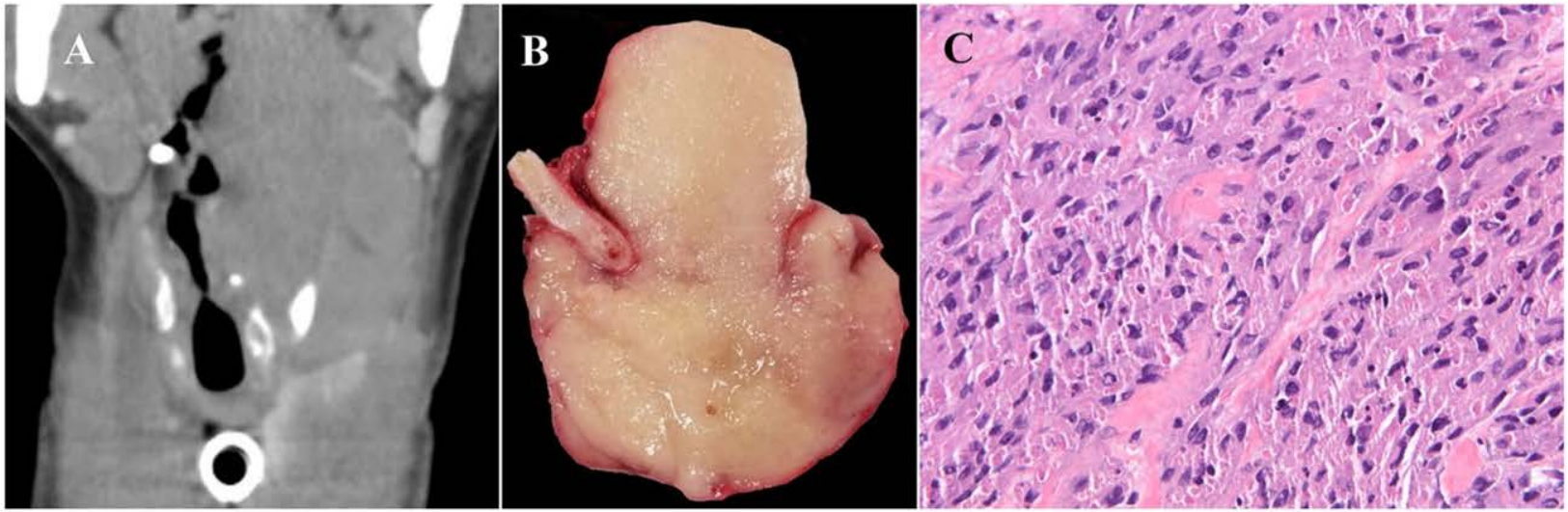

Fig. 6 **a** Reformatted coronal computed tomography demonstrates a large mass involving the left pharyngeal wall, the base of tongue, left true vocal cord, and subglottic region. **b** Excised malignant granular cell tumor shows glistening tan-yellow cut surface. **c** Sheets of spindle and polygonal cells with granular cytoplasm (HES × 40)

Back to our patient

- AJCC 8th: cT3N1M0, proposed stage III, 5 year survival ~ 50-60%

- NCCN: adjuvent CT or RT > surgery, with safe margin > 1mm
  - CT: AIM (doxorubicin, ifosfamide, mesna)
  - RT: 50 to 50.4 Gy external beam RT (EBRT)

- Surgical removal remains the mainstay of treatment.

Head & Neck MM, Special case

Supraglottic Spindle Cell Sarcoma

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