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Radiotherapy

Primary Myxofibrosarcoma of the Esophagus: A Case Report and Literature Review

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Abstract | **Case Report**

Background: Squamous cell, and adenocarcinoma are the most common histological types of esophageal tumors. Primary sarcomas of the esophagus are extremely rare, and even more rare myxofibrosarcoma. ***Case Presentation:*** We describe the case of a 42 years old male with a history of Plummer-Vinson syndrome, presenting with progressive dysphagia and weight loss. Clinical and radiological investigations were inconclusive. Histopathological analysis of biopsy specimens revealed a poorly differentiated spindle cell tumor with a myxoid stroma. Immunohistochemical profiling confirmed the diagnosis of grade II myxofibrosarcoma according to the FNCLCC grading system. Given the lesion's inaccessibility for surgical resection, the patient was treated with exclusive radiotherapy using Volumetric-modulated arc therapy (VMAT) at a total dose of 66 Gy in 33 sessions. The patient tolerated the treatment well, with a complete resolution of dysphagia by the end of treatment. ***Conclusion:*** This is the first reported case of a primary esophageal myxofibrosarcoma treated with exclusive radiotherapy. This case supports the role of radiotherapy as a curative option in non-operable cases.

Keywords: Esophageal cancer, Myxofibrosarcoma, Radiotherapy.

I. INTRODUCTION

Esophageal cancer is the seventh cause of cancer mortality worldwide. Two major pathological subtypes exist: esophageal squamous cell carcinoma and esophageal adenocarcinoma ([1]).

Primary sarcomas of the esophagus are exceptionally rare, accounting for less than 1% of all esophageal malignancies ([2]). Among them, leiomyosarcoma ([3]) is the most common primary esophageal sarcoma, while rhabdomyosarcoma ([4]) is rare. Other reported histopathological diagnoses include chondrosarcoma ([5]), and liposarcoma.

Myxofibrosarcoma (MFS) is a rare soft tissue sarcoma characterized by a malignant fibroblastic lesion with a variable myxoid stroma, and pleomorphism. It usually presents as superficial lesions, painless, and slow-growing mass.

MFS accounts for approximately 5% of all adult soft tissue sarcomas, with an annual incidence of 0.3–0.4 cases per 100,000 individuals. It usually presents as superficial lesions, painless, and slow-growing mass and the majority of cases occur in patients aged 50–80 years, with a slight male predominance.

Most cases of MFS occur in the extremities, but also can arise in less common anatomical locations, including the trunk, retroperitoneum, heart ([6]), and rarely, the gastrointestinal (GI) tract. Surgical resection and local adjuvant radiotherapy are the first choice to improve the prognosis ([7]).

This article aims to report and discuss a rare case of primary myxofibrosarcoma (MFS) of the esophagus, highlighting its clinical presentation, diagnostic challenges, and therapeutic management.

II. MEDICAL CASE

A 42-year-old patient with a history of Plummer-Vinson syndrome for the past four years, treated with repeated endoscopic dilations, presents with intermittent dysphagia to solid foods for two years. Initially attributed to his underlying condition, the progressive worsening of dysphagia associated with weight loss warrants further investigation

Clinical and Paraclinical Assessment:

- Clinical examination: No significant findings.
- Cervicofacial CT scan: No detectable abnormalities.
- Nasofibroscopy findings:
- Normal-appearing nasal mucosa and clear nasopharynx.
- Healthy and mobile base of the tongue and epiglottis.
- Difficult visualization of the remaining structures due to significant salivary stasis.

Additional Investigations

Direct laryngoscopy and hypopharyngoscopy under sedation revealed:

- Macroscopically normal vocal cords and arytenoids (mobility not assessed).
- Free piriform sinuses.
- Presence of leukoplastic lesions in the cricoarytenoid region.
- Multiple biopsies performed.

Pathological Examination

- Identification of a poorly differentiated spindle-cell tumor with occasional pleomorphic cells, showing moderate to marked cytonuclear atypia.
- Presence of an abundant myxoid stroma traversed by branched capillaries with thickened walls.

Immunohistochemistry:

- Negative markers: EMA, AE1/AE3, CD34, AML, Desmin, S100, Myogenin, MUC4, CD30, ERG, and CD45.
- Ki-67 proliferation index estimated at 20%.

III. CONCLUSION

Morphological and immunohistochemical features suggestive of a grade II myxofibrosarcoma (3+1+0) according to the FNCLCC classification. (Figure 1)

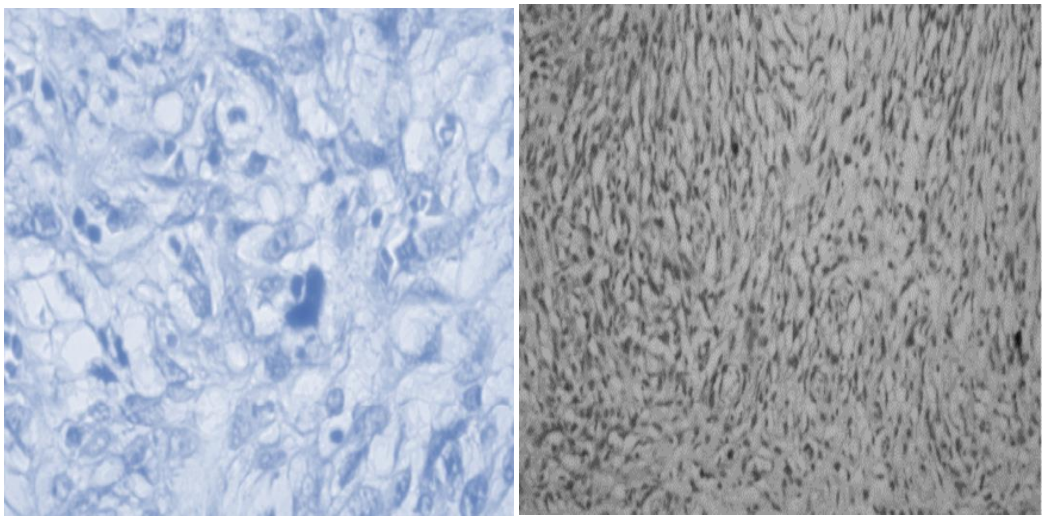


Figure 1: Histologic images of the myxofibrosarcoma

Complementary Imaging

- **Cervicothoracic CT scan:**
  - Preservation of the pharyngolaryngeal tract.
  - Complementary MRI performed (T2 Fat-Sat and diffusion sequences) reveals a small signal anomaly located retro-cricoid on the left side, involving the inferior constrictor muscle of the pharynx and the posterior cricoarytenoid muscle, showing an intermediate signal on T2 sequences and mild diffusion hyperintensity, measuring 11 × 9.5 mm.

TREATMENT

The patient was initially considered for surgical resection. However, the lesion was inaccessible for curative surgical treatment. After reviewing the literature and discussing the case in a multidisciplinary board, a decision was made to proceed with exclusive radiotherapy.

The treatment plan was established using Volumetric-modulated arc therapy (VMAT). The simulation was performed using the CT simulator. The patient was immobilized in a supine position with a five-point thermoplastic head and neck mask.

Planning CT images were acquired with a slice thickness of 3 mm. The CT DICOM images were transferred to the treatment planning system for the delineation of target volumes, including the gross tumor volume (GTV), the clinical target volume (CTV), and the planning target volume (PTV).

The gross target volume (GTV) includes the primary tumor, The CTV includes the GTV with a 5 mm margin, the PTV defined as the CTV with a 5 mm.

A prescription dose of 66 Gy in 33 fractions was given to PTV en 2 Gy per fraction en 33 sessions,5 sessions per week.

Organs at risk (OARs) were contoured according to the Radiation Therapy Oncology Group (RTOG) normal tissue contouring: (Figure 2)

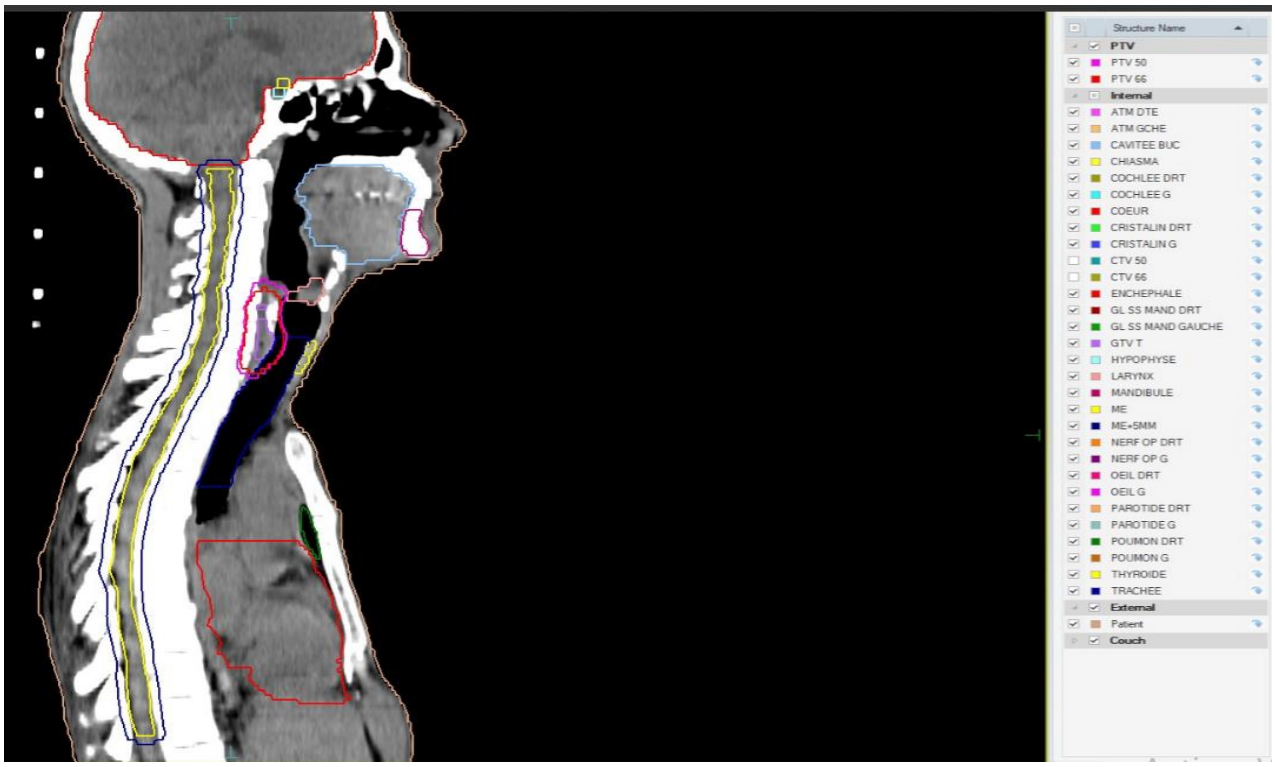


Figure 2: Organs at risk

Volumetric modulated arc therapy (VMAT) with a two-arc technique plan was generated using 6 MV photon beams. PTV coverage and dose to OARs were acceptable. (Figure 3)

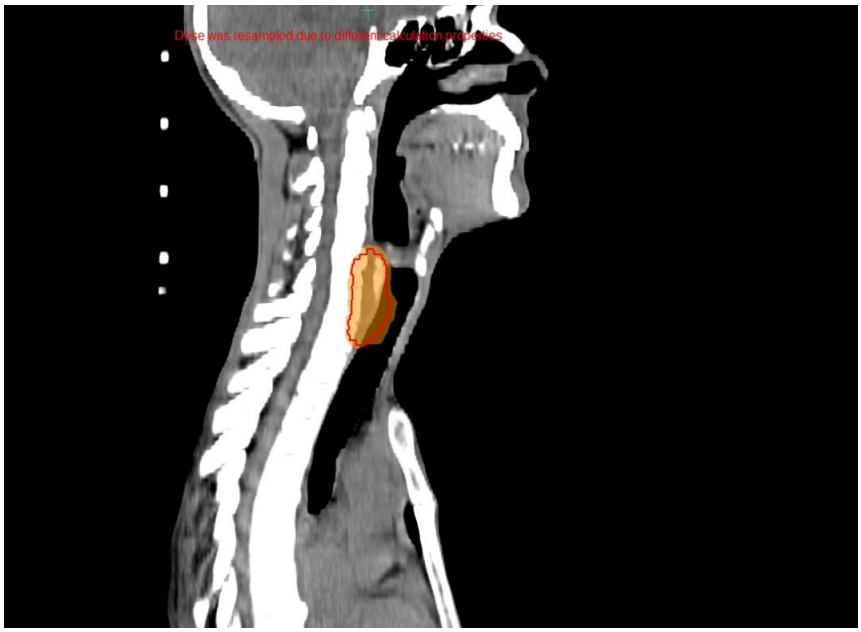
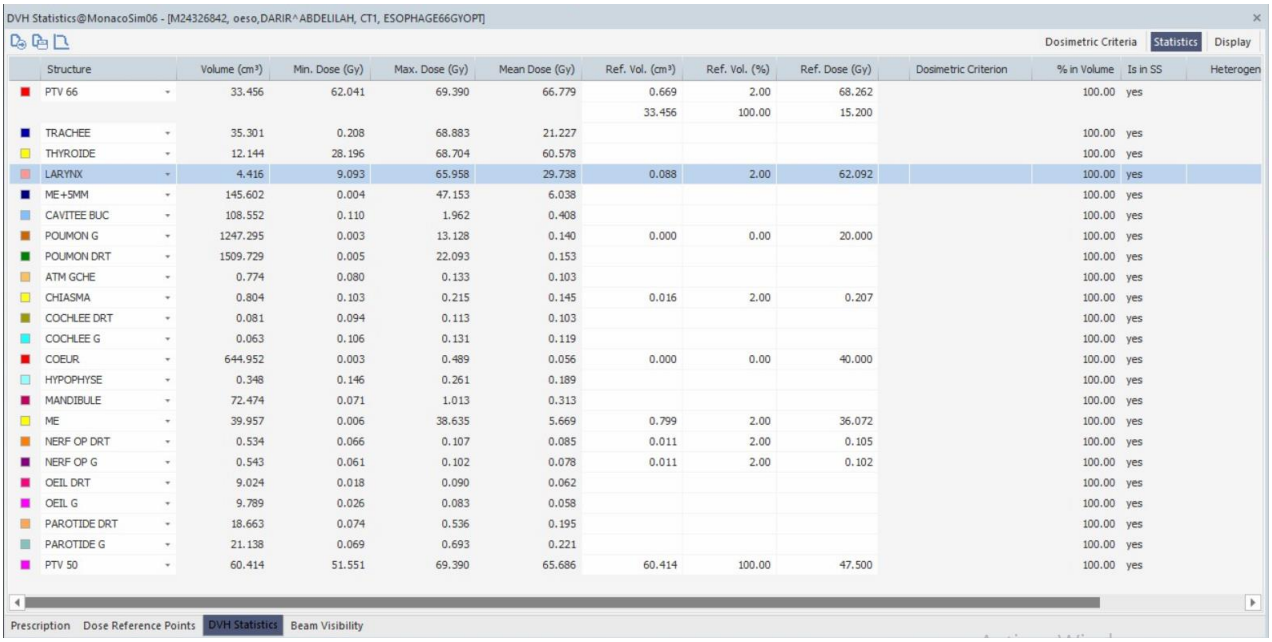


Figure 3: Radiotherapy isodose curve (sagittal view): the planning CT scan with 66 Gy (in orange) isodose lines

Dose constraints to OARs were defined according to the RECORD (Figure 4), Patient setup was verified daily by cone beam CT imaging before treatment.


DVH Statistics@MonacoSim06 - [M24326842, oeso,DARIR^ABDELILAH, CT1, ESOPHAGE66GYOPT]

Dosimetric Criteria | Statistics | Display

Structure	Volume (cm³)	Min. Dose (Gy)	Max. Dose (Gy)	Mean Dose (Gy)	Ref. Vol. (cm³)	Ref. Vol. (%)	Ref. Dose (Gy)	Dosimetric Criterion	% in Volume	Is in SS	Heterogen
PTV 66	33.456	62.041	69.390	66.779	0.669	2.00	68.262		100.00	yes	
					33.456	100.00	15.200				
TRACHEE	35.301	0.208	68.883	21.227					100.00	yes	
THYROIDE	12.144	28.196	68.704	60.578					100.00	yes	
LARYNX	4.416	9.093	65.958	29.738	0.088	2.00	62.092		100.00	yes	
ME+5MM	145.602	0.004	47.153	6.038					100.00	yes	
CAVITEE BUC	108.552	0.110	1.962	0.408					100.00	yes	
POUMON G	1247.295	0.003	13.128	0.140	0.000	0.00	20.000		100.00	yes	
POUMON DRT	1509.729	0.005	22.093	0.153					100.00	yes	
ATM GCHE	0.774	0.080	0.133	0.103					100.00	yes	
CHIASMA	0.804	0.103	0.215	0.145	0.016	2.00	0.207		100.00	yes	
COCHLEE DRT	0.081	0.094	0.113	0.103					100.00	yes	
COCHLEE G	0.063	0.106	0.131	0.119					100.00	yes	
COEUR	644.952	0.003	0.489	0.056	0.000	0.00	40.000		100.00	yes	
HYPOPHYSE	0.348	0.146	0.261	0.189					100.00	yes	
MANDIBULE	72.474	0.071	1.013	0.313					100.00	yes	
ME	39.957	0.006	38.635	5.669	0.799	2.00	36.072		100.00	yes	
NERF OP DRT	0.534	0.066	0.107	0.085	0.011	2.00	0.105		100.00	yes	
NERF OP G	0.543	0.061	0.102	0.078	0.011	2.00	0.102		100.00	yes	
OEIL DRT	9.024	0.018	0.090	0.062					100.00	yes	
OEIL G	9.789	0.026	0.083	0.058					100.00	yes	
PAROTIDE DRT	18.663	0.074	0.536	0.195					100.00	yes	
PAROTIDE G	21.138	0.069	0.693	0.221					100.00	yes	
PTV 50	60.414	51.551	69.390	65.686	60.414	100.00	47.500		100.00	yes	

Prescription | Dose Reference Points | DVH Statistics | Beam Visibility

Figure 4: DVH statistics showing the dose distribution across the target volumes and organs at risk (OARs) in the VMAT planning

During the weekly follow-up consultation conducted during treatment, the patient developed grade I radiomucositis according to the Common Terminology Criteria for Adverse Events (CTCAE) version 4.0.

At the end-of-treatment, the patient reported complete resolution of dysphagia along with an overall improvement in general condition.

IV. DISCUSSION

Esophageal sarcomas represent an exceedingly rare subset of esophageal malignancies [8].

Myxofibrosarcoma, a distinct entity within myxoid mesenchymal sarcomas, is most commonly seen in elderly patients and typically arises in the extremities rather than the gastrointestinal tract.

Myxofibrosarcoma was first proposed by Angervall et al. [9], who described it as a group of fibroblastic lesions with varying degrees of cellular distribution, nuclear pleomorphism, and mitotic activity ranging from a sparsely cellular lesion with minimal cytological atypia to a more cellular lesion with features resembling those of pleomorphic-storiform malignant fibrous histiocytoma (MFH).

The latest WHO classification recognizes MFS as a separate pathological entity, with key diagnostic features including:

1. Multinodular infiltrative architecture.
2. Variably pleomorphic fibroblastic cells.
3. Distinctive curvilinear vasculature.
4. Hypercellular foci in higher-grade variants.
5. Myxoid stroma (10).

MFS is extremely rare in the head and neck region, with only a few reported cases in the sinus sphénoïdal (11), thyrid gland (12), mandible, (13), hypopharynx (14) and maxillary sinus (15).

The clinical presentation of esophageal sarcomas can be nonspecific, with symptoms such as progressive dysphagia, weight loss, and odynophagia, often leading to delayed diagnosis (16). In our case, the patient's history of Plummer-Vinson syndrome complicated the clinical presentation, with masking the progression of symptoms attributed to an underlying malignant process.

Plummer-Vinson syndrome is known to predispose patients to squamous cell carcinoma of the esophagus and pharynx (17), for now no association with mesenchymal tumors, including MFS, has been reported. The co-occurrence in this case need further investigation.

The management of esophageal MFS poses unique challenges due to its rarity and the anatomical constraints of the esophagus. Surgical resection remains the cornerstone of treatment, with the goal of achieving negative margins to minimize local recurrence (6,18). Adjuvant radiotherapy is often employed to reduce the risk of local recurrence, particularly in high-grade tumors or where wide surgical margins cannot be achieved (19). Systemic chemotherapy may be considered for metastatic or unresectable cases, although its efficacy in MFS is limited. Emerging therapies, such as immune checkpoint inhibitors and targeted agents, are currently under investigation and may offer new hope for patients with advanced disease (20)

Radiotherapy alone in the management of MFS is less commonly reported but has shown promise in

inoperable cases or in patients unfit for surgery with soft tissue sarcoma ([21]). Intensity-modulated radiation therapy (IMRT) or volumetric modulated arc therapy (VMAT) techniques offer the advantage of delivering high-dose conformal radiation while minimizing toxicity to surrounding structures.

In our case and due to the tumor's unresectability, the patient received definitive radiotherapy using a VMAT technique. A total dose of 66 Gy was delivered in 2 Gy fractions, five days per week, without concurrent chemotherapy. During weekly follow-up consultations, the patient reported gradual improvement in symptoms, and by the end of treatment, there was a marked relief of dysphagia.

V. CONCLUSION

Primary myxofibrosarcoma of the esophagus is an extremely rare diagnosis. This case highlights the importance of considering mesenchymal tumors in atypical esophageal lesions. Histopathological and immunohistochemical evaluation is essential for diagnosis. In the absence of a surgical option, definitive radiotherapy can offer effective local control. However, due to the rarity of this presentation, standardized treatment guidelines are lacking, and management must be individualized based on tumor characteristics, patient factors, and available resources. Long-term follow-up remains essential to monitor for local recurrence and distant metastases.

([1]) Bray, F · Laversanne, M · Sung, H · et al: Global cancer statistics 2022: GLOBOCAN estimates of incidence and mortality worldwide for 36 cancers in 185 countries
CA Cancer J Clin. 2024; 74:229-263, PMID: 38572751,DOI: 10.3322/caac.21834
([2]) Zhang BH, et al. Sarcomas of the Esophagus: A Review of the Literature. World J Surg Oncol. 2013.
([3]) A. J. Ruppert-Kohlmayr, J. Raith, G. Friedrich, S. Regauer, K. W. Preidler, and D. H. Szolar, "Giant liposarcoma of the esophagus: radiological findings," *Journal of Thoracic Imaging*, vol. 14, no. 4 I, pp. 316–319, 1999.
([4]) Y. K. Batoroev and G. K. Nguyen, "Esophageal rhabdomyosarcoma: report of a case diagnosed by imprint cytology," *Acta Cytologica*, vol. 50, no. 2, pp. 213–216, 2006.
([5]) Yaghmai I, Ghahremani GG. Chondrosarcoma of the esophagus. Am J Roentgenol. 1976;126:1175-7.
([6]) Lazaros GA, Matsakas EP, Madas JS, Toli DI, Nikas DJ, Kershaw MA, Alpert MA. Primary myxofibrosarcoma of the left atrium: case report and review of the literature. Angiology. 2008;59:632-635.
([7]) Wang XS, Zhao CG, Wang HM, Wang XY. Giant myxofibrosarcoma of the esophagus treated by endoscopic submucosal dissection: A case report. *World J Clin Cases* 2023; 11(5): 1094-1098 [PMID: 36874423 DOI: 10.12998/wjcc.v11.i5.1094].
([8]) Y. Akishima, Y. Akasaka, G. Yih-Chang et al., "Histiocytic sarcoma with fatal duodenal ulcers," *Pathology Research and Practice*, vol. 200, no. 6, pp. 473–478, 2004.
([9]) Angervall L, Kindblom LG, Merck C. Myxofibrosarcoma. A study of 30 cases. Acta Pathol Microbiol Scand 1977;85A(2):127–40.
([10]) Huang HY, Mentzel TDW, Shibata T. Myxofibrosarcoma.. In: WHO Classification of Tumours Editorial Board. WHO Classification of Tumours. Soft Tissue and Bone Tumours. 5th ed. Lyon, France: IARC; 2020:124e126.
([11]) Lam PK, Trendell-Smith N, Li JH, Fan YW, Yuen AP. Myxofibrosarcoma of the sphenoid sinus. J Laryngol Otol 2002;116(6):464–6.
(12) Y Darouassi 1, H Attifi 2, M Zalagh 2, I Rharrassi 3, F Benariba Myxofibrosarcoma of the thyroid gland, DOI: 10.1016/j.anorl.2013.09.004
([13]) Kummoona R. Central myxofibrosarcoma of the mandible treated by radical resection. Oral Surg Oral Med Oral Pathol 1975;39(5):713–7
([14]) Nishimura G, Sano D, Hanashi M, *et al.,* Myxofibrosarcoma of the hypopharynx. Auris Nasus Larynx 2006;33:93–6.
([15]) Pomerantz JM, Sanfacon DG, Dougherty TP, Hanson S. Myxofibrosarcoma of the maxillary sinus. Del Med J 1982;54(3):147–52.
([16]) Brierley J, *et al.,* TNM Classification of Malignant Tumours, 8th Edition. Wiley-Blackwell.
([17]) Goel A, *et al.,* Plummer-Vinson syndrome: A review. World J Gastroenterol. 2015.
([18]) Huang HY, *et al.,* Prognostic Factors in Myxofibrosarcoma: A Clinicopathologic Study of 175 Cases. Am J Surg Pathol. 2004.
([19]) O'Sullivan B, *et al.,* Preoperative Versus Postoperative Radiotherapy in Soft-Tissue Sarcoma of the Limbs. Lancet. 2002.
([20]) Pan, M., Zhou, M., Xie, L. *et al.,* Recent advances in sarcoma therapy: new agents, strategies and predictive biomarkers. *J Hematol Oncol* 17, 124 (2024). https://doi.org/10.1186/s13045-024-01650-6
([21]) Guadagnolo BA, *et al.,* The Role of Radiotherapy in Soft Tissue Sarcoma Management. Clin Orthop Relat Res. 2007.