

## Case Report

### A case report of Synovial Sarcoma of the Esophagus

Pedram Fadavi<sup>1</sup>, Helaleh Khoshbakht Ahmadi<sup>1\*</sup>, Mastaneh Sanei<sup>1</sup>, AD Kakhki<sup>2</sup>

#### Author Information

*1 Department of Radiation Oncology, Shohadaye Haft-e-Tir Hospital, Iran University of Medical Sciences, Tehran, Iran.*

*2 Department of thoracic Surgery, Shahid Beheshti University of Medical Sciences, Tehran, Iran.*

Submitted: 16.11.2019

Accepted: 01.02.2019

Published : 02.02.2020

**Keywords:** Esophagus, Synovial Sarcoma

#### Introduction

The most common pathology of esophageal cancer is carcinoma including adenocarcinoma and squamous cell carcinoma but other histologies may be seen (e.g. sarcoma, melanoma and lymphoma). synovial sarcomas are a rare entity of malignant mesenchymal tumors arising in periarticular region of the extremities. Less frequently, they have occurred in other sites such as head & neck and digestive tract. We describe a primary SS of the esophagus in a 28-year-old man.

An otherwise healthy 28-year-old man presented with dysphagia to solid foods and epigastric discomfort for about 4 months.

He had anorexia and weight loss (20 kg) from 3 month before admission. There was no abnormal finding on Physical examination.

He had no significant medical or family history.

#### Case Report

The patient underwent esophagoscopy that through the posterior mediastinum and revealed a large mass that was 19 cm from cervical esophagogastric anastomosis.

the incisors.

Biopsy was taken. Biopsy was suggestive of low grade spindle cell sarcoma.

EUS showed giant submucosal lesion from muscularis mucosa, 18 – 30 cm from incisors without any regional lymph nodes. Spiral CT scan of the Neck & Thorax revealed a large intraluminal tumoral mass in the esophagus.

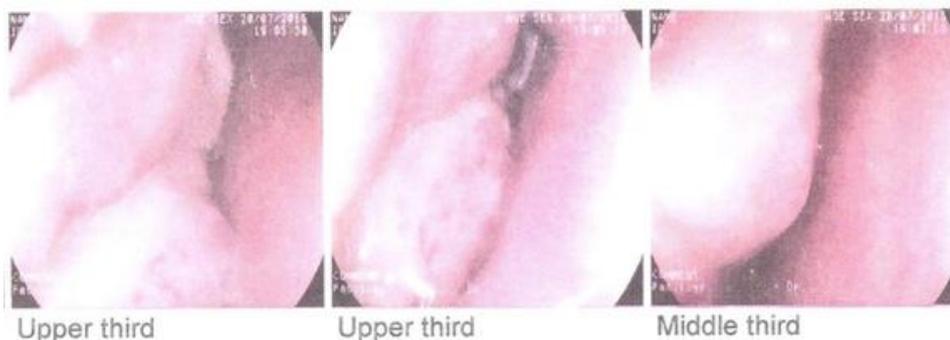
Multiple lymphadenopathies were detected on both sides of the neck.

Abdominopelvis CT scan was normal.

The patient thereafter received chemotherapy (Ifosfamide, Etoposide, Mesna every 3 week for 6 courses).

Contrary to our recommendation, he underwent radiation therapy to the mediastinum (54 Gy in 30 fr) in another institute. He remained disease-free for 4 months after radiation therapy. He died soon afterwards due to diffuse lung metastasis. This case survived about 18 months.

The patient underwent a transhiatal total esophagectomy with gastric pull-up



### Pathologic findings

Macroscopic examination: a submucosal mass of upper esophagus consists of pieces soft ,gray-white tissue totally measuring 10\*10\*5cm .

Microscopic examination: sections show biphasic neoplasm infiltrating squamous-lined mucosa. Spindle cell component is densely cellular with finely dispersed chromatin, scant cytoplasm & notable mitotic activity. The glandular elements are positive for AE1/3 & CK 19.S100,MIC 2,Ckit,CD34,Desmin,Actin,SMA are negative.Bcl2 is positive in some of cells of both components. Ki67 stained about 15% of neoplastic cells. These findings confirmed the diagnosis of synovial sarcoma of the esophagus.

Esophageal surgical margins are free of tumor.

Subcarinal lymph node biopsy is negative for malignancy.

### Discussion

Synovial sarcoma (SS) is an uncommon malignant mesenchymal tumor of soft tissue, involving mainly the periarticular region of the extremities such as tendons and bursal structures of large joints but rarely in the synovium. Sometimes their origin is in the head and neck and often as parapharyngeal or retropharyngeal masses (1). Also it has been reported in other sites such as abdominal wall and retroperitoneum(2,3).



Synovial sarcomas of the digestive tract component that forms clefts and gland-like spaces, and spindle cell region, esophagus and account for 0.1% to 1.5% therefore, diagnosis is made based on the presence of the biphasic pattern. In 70 to 90% of synovial sarcoma, t(X;18) translocation is a sensitive marker in establishing the diagnosis of synovial sarcomas [12]. Other spindle-cell sarcomas such as sarcomatoid carcinomas (carcinosarcoma), fibrosarcomas, and solitary fibrous tumors are among the differential diagnoses of biphasic synovial sarcomas of the esophagus.

Combined histological type sarcomas are more frequently seen than pure sarcomas and the most common type of the pure mesenchymal tumors of the esophagus is Leiomyosarcoma. The median age at presentation is 58 years [range from 26–76 years] [5], and male to female ratio is 2 to 1. In the esophagus the most common tumor site is the cervical to upper third of the thoracic esophagus. (22)

Their pathology may be biphasic, monophasic, or poorly differentiated. Characteristic histologic findings of synovial sarcoma is often a biphasic pattern accompanied by an epithelial

On immunohistochemistry, neuroectodermal antigens such as CD99, CD56 and CD57 are often expressed. [21]

On immunohistochemistry, neuroectodermal antigens such as CD99, CD56 and CD57 are often expressed. [21]

Their pathology may be biphasic, monophasic, or poorly differentiated.

Characteristic histologic findings of synovial sarcoma is often a biphasic pattern accompanied by an epithelial

### References

1. Enzinger FM, Weiss SW. Synovial sarcoma. In: Soft tissue tumors. 3rd ed. St. Louis: Mosby; 1995. pp. 757–86.
2. Al-Dewachi HS, Sangal BC, Zakaria MA (1981) Synovial sarcoma of the abdominal wall: a case report and study of its fine structure. J SurgOncol 18:335–344
3. Yokouchi J, Saitoh K, Shimoyamada K, Shimura Y, Itoh N, Matsuda H, Kuroda M, Wakabayashi Y, Kanesaka N, Kotake F, Abe K (1993) A case of retroperitoneal synovial sarcoma (in Japanese). Rinshogazou (ClinImagiol) 9:103–106

Also, tumor cells show positive staining for the epithelial elements (eg. cytokeratin (AE-1/3) and EMA). Tumoral mass may enhance on CT/MRI imaging as intramural mass [11].

### **Treatment:**

However, their specificity and sensitivity of all of these markers are limited for diagnosing synovial sarcoma. It seems TLE-1 is the best marker because expression being associated with a sensitivity and specificity of 75 and 96 %, respectively [22]. The mainstay of treatment is surgery provided that it is feasible [13]. The best choice is esophagectomy/esophagogastrectomy. Therefore for resectable tumors, surgery would be the first step of treatment while The functional outcome is acceptable.

### **Clinical Features:**

Tumoral mass can cause compression or infiltrate the adjacent structures; therefore progressive dysphagia is the most common complaint. Weight loss, chest discomfort, burning retrosternal pain, nausea, and vomiting are other symptoms present in patients. In the adjuvant setting, radiotherapy and chemotherapy have a controversial role [13]. Resection with adequate margins is a goal of surgery. So in cases of uncertain margins, postoperative radiotherapy would be considered.

### **Diagnosis:**

These tumors are often presented with polypoid and exophytic masses on Endoscopy [9] and rarely as ulcerating tumours [10]. Stenting as a Palliative procedure can relieve dysphagia and improve quality of life [14].

### **References**

4. Perch SJ, Soffen EM, Whittington R, et al: Esophageal sarcomas. *J Surg Oncol*48:194-198, 1991
5. G. Rocco, V. F. Trastek, C. Deschamps, M. S. Allen, D. L. Miller, and P. C. Pairolero, "Leiomyosarcoma of the esophagus: results of surgical treatment," *Annals of Thoracic Surgery*, vol. 66, no. 3, pp. 894-896, 1998.
6. K. S. Kwatra, B. R. Prabhakar, S. Jain, and J. S. Grewal, "Sarcomatoid carcinoma (carcinosarcoma) of the esophagus with extensive areas of osseous differentiation: a case report," *Indian Journal of Pathology and Microbiology*, vol. 46, no. 1, pp. 49-51, 2003.

Occasionally, neoadjuvant therapy is also dacarbazine, either alone or in combination. considered based on functional outcomes.

and other factors. Today, a number of targeted therapies have

For unresectable lesions, treatment options been considered for adding to include radiation therapy, chemotherapy, or chemotherapy regimen and they have had chemoradiotherapy. promising results. For example Pazopanib

It seems synovial sarcoma is relatively significantly prolonged median progression chemosensitive according to existing data -free survival and tended to improve [24], and chemotherapy has improved overall survival, although this latter trend survival in primary synovial sarcoma of was not significant [25].

the extremities [23].

The primary chemo therapy regimen consists of doxorubicin, ifosfamide, and

## References

7. S. Kubota, T.Morita, K.Murakawa et al., "A case or pseudosarcoma associated with type 3 squamous cell carcinoma of the esophagus: report of a case," *Surgery Today*, vol. 29, no. 8, pp. 1999, 781-777.
8. T. D. Liakakos, T. G. Troupis, C. Tzathas et al., "Primary liposarcoma of esophagus: a case report," *World Journal of Gastroenterology*, vol. 12, no. 7, pp. 1149–1152, 2006.
9. F. Kayaselcuk, I. Tuncer, Y. Toyganozu, N. Bal, and G. O'zgu'r, "Carcinosarcoma of the stomach," *Pathology and Oncology Research*, vol. 8, no. 4, pp. 275–277, 2002.
10. O. Chino, H. Kijima, H. Shimada et al., "Clinicopathological studies of esophageal carcinosarcoma: analyses of its morphological characteristics using endoscopic, histological, and immunohistochemical procedures," *Endoscopy*, vol. 32, no. 9, pp. 706–711, 2000.
11. C. S. Pramesh, G. H. Pantvaidya, M. T. Moonim, N. A. Jambhekar, S. Sharma, and R. K. Deshpande, "Leiomyosarcoma of the esophagus," *Diseases of the Esophagus*, vol. 16, no. 2, pp. 142–144, 2003.
12. S. D. Billings, L. F. Meisner, O. W. Cummings, and E. Tejada, "Synovial sarcoma of the upper digestive tract: a report of two cases with demonstration of the X;18 translocation by fluorescence in situ hybridization," *Modern Pathology*, vol. 13, no. 1, pp. 68–76, 2000.
13. C. S. Pramesh, G. H. Pantvaidya, M. T. Moonim, N. A. Jambhekar, S. Sharma, and R. K. Deshpande, "Leiomyosarcoma of the esophagus," *Diseases of the Esophagus*, vol. 16, no. 2, pp. 142–144, 2003.

*The Present Case in the Context of the* than 5 cm in maximum diameter may be curative and that adjuvant chemotherapy

*Literature:* Amr et al. reported a polypoid synovial may not be necessary for such patients, sarcoma arising in the upper third of the because the value of chemotherapy esophagus in a 25-year-old man and did a for synovial sarcoma is still controversial. total esophagectomy followed by [17].

radiotherapy. This approach was effective On the other hand, Pappo et al. concluded [15]. that adjuvant chemotherapy is meritted in

Bloch et al. reported a case that was treated children with resected synovial sarcoma with a local resection and postoperative (clinical group I or II) who present with radiation therapy and there was no unfavorable clinicopathologic features evidence of disease after 36 months [16]. such as large, invasive, or grade 3

Yokoyama et al. did a clinicopathologic lesions. [18]

study of 18 cases and concluded that complete surgical resection of tumors less

## References

14. M. Conio, S. Bianchi, R. Filiberti, and A. De Ceglie, "Self-expanding plastic stent to palliate symptomatic tissue in/overgrowth after self-expanding metal stent placement for esophageal cancer," *Diseases of the Esophagus*, vol. 23, no. 7, pp. 590–596, 2010.
15. Amr SS, Shihabi NK, Hajj HA (1984) Synovial sarcoma of the esophagus. *Am J Otolaryngol* 5:266–269
16. Bloch MJ, Iozzo RV, Edmunds LH, Brooks JJ (1987) Polypoid synovial sarcoma of the esophagus. *Gastroenterology* 92:229–233
17. Yokoyama K, Shinohara N, Kondo M, Mashima T (1995) Prognostic factors in synovial sarcoma: a clinicopathologic study of 18 cases. *Jpn J Clin Oncol* 25:131–134
18. Pappo AS, Fontanesi J, Luo X, Rao BN, Parham DM, Hurwitz C, Avery L, Pratt CB (1994) Synovial sarcoma in children and adolescents: the St Jude Children's Research Hospital experience. *J Clin Oncol* 12:2360–2366
19. Holtz F, Magielski JE (1985) Synovial sarcoma of tongue base. The seventh reported case. *Arch Otolaryngol* 111:271–272
20. Shusaku Habu, Eizo Okamoto, Akihiro Toyosaka, Yoshiyuki Nakai, and Masaharu Takeuchi (1998) Synovial Sarcoma of the Esophagus: Report of a Case *Jpn J Surg* (1998) 28:401–404

Holtz et al. reported A 16-year-old boy with Synovial Sarcomas of the S Habo et.al chose a local excision and Tongue Base. His tumor was postoperative radiation and resected and Four weeks after chemotherapy(selected minimally surgery, a course of radiation invasive surgical management thus totaling 6,700 rad to the tumor bed avoiding a total esophagectomy) ; was given over eight weeks. The patient has shown no evidence of recurrence or metastasis 20 months initial surgery and 40 months after postoperatively. They believe that the initial symptom with clinical and Postoperative radiation and chemotherapy roentgenographic evidence of decreases the rate of local metastases to lungs, liver, bone, recurrence and metastasis [20]. retroperitoneum, and mediastinum despite receiving multi chemotherapy agents. Holtz believes that the size of the primary tumor is the most critical determinant of the

## References

21. Catherine Butori & Véronique Hofman & Rita Attias & Jérôme Mouroux & Florence Pedeutour & Paul Hofman(2006) Diagnosis of primary esophageal synovial sarcoma by demonstration of t(X;18) translocation: a case report *Virchows Arch* (2006) 449:262–267. DOI 10.1007/s00428-006-0234-1
22. Masahiro Niihara • Hiroshi Sato • Kimihide Kusafuka • Satoshi Kamiya • Ryo Ashida • Takashi Nakajima • Yasuhiro Tsubosa; Biphasic synovial sarcoma of the cervical esophagus confirmed by the presence of SYT-SSX1 fusion transcripts(2014) *Esophagus* DOI 10.1007/s10388-014-0419-7
23. Eilber FC, Eilber FR, Eckardt J, et al. The impact of chemotherapy on the survival of patients with high-grade primary extremity liposarcoma. *Ann Surg.* 2004;240:686–95.
24. Canter RJ, Qin LX, Maki RG, Brennan MF, Ladanyi M, Singer S, et al. A synovial sarcoma-specific preoperative nomogram supports a survival benefit to ifosfamide-based chemotherapy and improves risk stratification for patients. *Clin Cancer Res.* 2008; 14(24): 8191-7

It has not definitely shown that these determination of prognosis are under systemic therapies for synovial sarcoma of consideration. Definitely, these finding are the esophagus are effective. Unfortunately contributory factors for selecting the best The 2-year survival of esophageal synovial treatment approach for synovial sarcoma of sarcoma is not very good according to the the esophagus previous studies.

Genetic features and its role in

---

### References

25. [van der Graaf WT, Blay JY, Chawla SP, Kim DW, Bui-Nguyen B, et al. Pazopanib for metastatic soft-tissue sarcoma \(PALETTE\): a randomised, double-blind, placebo-controlled phase 3 trial. Lancet. 2012;379 \(9829\):1879–86.](#)