INTRODUCTION

We present the case of a 49-year-old man with odynophagia, dysphagia, and regurgitation. Endoscopy showed a partially obstructed mass in upper esophagus with ulceration of overlying mucosa. The imaging findings indicated a heterogeneous intraluminal mass in the esophagus. The patient underwent successful total esophagectomy and gastric pull-up with laryngeal preservation.

A synovial sarcoma is a soft tissue sarcoma subtype, which is assumed to arise from mesenchymal cells. The name is derived from its microscopic resemblance to the synovium under light microscopy. Synovial sarcoma accounts for approximately 5%-10% of all soft tissue sarcomas. Synovial sarcoma of digestive system is an extremely rare phenomenon. Among sarcomas, primary synovial sarcoma of the esophagus is an uncommon tumor that may present with dysphagia, odynophagia, and loss of weight. It occurs at a male-to-female ratio of 3:2, especially in young adults and adolescents. Here, we describe a case of synovial sarcoma of the esophagus, based on immunohistochemical (IHC) staining.

CASE PRESENTATION

The patient was a 49-year-old man with odynophagia, dysphagia to liquids, and regurgitation for two years, which had worsened over the last month. He refused any loss of appetite, loss of weight, or aspiration, with no clinically significant lymphadenopathy in the head or neck. The laboratory data were within the normal ranges. The gastrointestinal (GI) endoscopy showed a partially obstructed mass in the upper esophagus with ulcerating overlying mucosa. The patient underwent successful total esophagectomy and gastric pull-up with laryngeal preservation.

The results of magnetic resonance imaging (MRI) showed a large heterogeneous intraluminal solid mass, measuring...
57 × 47 × 43 mm in the upper thoracic esophagus, which led to the anterior displacement of trachea, with low T1 and high T2 signals and marked enhancement in the post-contrast images. No fat plane was observed between the lesion and the prevertebral fascia. Therefore, esophageal sarcoma was considered, as squamous cell carcinoma (SCC) and adenocarcinoma are less probable (Figure 2). Due to the absence of a postcricoid area superior to the tumor, the patient was a candidate for esophagectomy and gastric pull-up with laryngeal preservation (Figure 3).

Written informed consent was obtained from the patient before surgery. Using the lateral pharyngotomy approach, unlikely imaging findings included prevertebral fascia involvement and simple detachment and resection of tumor from the prevertebral fascia and the retropharyngeal space with a safe margin. The superior margin of the tumor was dissected from the postcricoid region, with no evidence of erosion (negative for tumor deposits on the frozen section). Following esophagus resection, the anterior aspect of the proximal part of the stomach was stitched to the normal mucosal strip in the

**FIGURE 1** A, B, Axial view of chest CT shows a large heterogenous intraluminal mass in cervical esophagus with compression effect on posterior wall of trachea

**FIGURE 2** Pre-operative neck MRI. A-F, Sagittal and axial plane of T1, B, T1 Fat suppression, D&F T1 Post GAD, E: T2
superior part of the resected site, and the posterolateral part of the stomach was stitched to the remnant retropharyngeal mucosa with Vicryl 3-0. The left recurrent laryngeal nerve was sacrificed due to tumor involvement.

On the first postoperative day, the patient was evaluated with a flexible fiberoptic bronchoscope through the nose. The mucosal color was pink, and the laryngeal tissue was viable. Although the right recurrent laryngeal nerve was preserved, double true vocal cord (TVC) paralysis occurred. The patient successfully adhered to an oral diet after no leaking confirmation, based on the gastrografin study. He was discharged from the hospital with a jejunostomy tube and a tracheostomy tube, which remained for two and six months, respectively (Figure 4). At 12 months after surgery, he could talk without tracheostomy and perform activities. The specimen size was $7 \times 4.5 \times 3$ cm, consisting of a firm creamy mass, measuring $5 \times 4.5 \times 3$ cm with hemorrhagic spots, which were reported as synovial sarcoma of the esophagus (Figures 5 & 6). Also, proximal and distal margins of the specimen were free from the tumor, and vascular invasion was not identified.

## DISCUSSION

Synovial sarcoma is a rare type of soft tissue sarcoma, which is typically found in the extremities and adjacent to tendon...
A synovial sarcoma of the head and neck is extremely rare and accounts for only 3% to 9% of all sarcomas. In this regard, a review study by F. Sista et al showed that the stomach was the most common site of monophasic synovial sarcoma, followed by jejunoileal, duodenal, and colorectal regions. Although monophasic synovial sarcoma has not been reported in the esophagus, approximately 75% of biphasic synovial sarcomas of the digestive system occur in the esophagus.

The CT scan findings typically indicate a well-defined heterogeneous soft tissue mass. Also, CT scan is useful for detecting tumor calcifications. However, MRI is the imaging modality of choice to stage the tumor locally. The tumor is commonly hypointense to muscles and heterogeneous on T1- and T2-weighted images, respectively. The T2-weighted MRI images may demonstrate the "triple sign" due to the presence of areas with low, medium, and high signal intensities. Histologically, they are classified into three types: monophasic, biphasic, and poorly differentiated. The monophasic type consists of entirely spindle-shaped cells. The biphasic type is composed of both spindle-shaped and epithelial components, while the poorly differentiated type is characterized by a predominantly round or short spindle-cell morphology. In the present case, a poorly differentiated synovial sarcoma was reported. Overall, the characteristic IHC profile for the diagnosis of synovial sarcoma includes the expression of transducer-like enhancer of split 1 (TLE-1), CD99, partial expression of cytokeratin, and epithelial membrane antigen (EMA). In our case, CK5, CK6, CK7, TLE1, CD99, BCL-2, and CKAEl/AE3 were positive.

Cytogenetically, translocation between X and 18 chromosomes is seen in more than 90% of synovial sarcomas. Generally, a combination of surgery and adjuvant therapy is used to treat synovial sarcoma. Once an adequate clear margin is not achievable, radiotherapy is performed. Our patient underwent total esophagectomy and gastric pull-up with laryngeal preservation; however, radiotherapy was not performed because of no evidence of residual tumor. Also, the larynx was preserved in our case.

The factors indicating a poor prognosis include a large tumor size, cystic or hemorrhage components, tumor location in the head, neck, or trunk, older age, a poorly differentiated type on histology, heterogeneity, necrosis, a high nuclear grade, a high mitotic rate, and p53 mutations. The overall five-year survival is estimated at 36% to 76%. Although advanced imaging investigations, such as MRI and/or CT scan, are particularly useful in diagnosing neoplasms of the neck, synovial sarcoma of the GI tract needs to be differentiated from malignant gastrointestinal stromal tumor, leiomyosarcoma, and sarcomatoid carcinoma.

CONCLUSION

Although rare, synovial sarcoma should be considered in the differential diagnosis of esophageal lesions. In our patient, the type of surgery, that is, esophagectomy and gastric pull-up with laryngeal preservation, was selected, based on the involved tumor areas with major impacts on the patient's quality of life.

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CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Aslan Ahmadi, Mohammad Dehghani Firouzabadi, Ayda Sanaei, Hadi Ghanbari, and Maryam Roomiani: involved in conception and design of work. Aslan Ahmadi, Mohammad Dehghani Firouzabadi, Ayda Sanaei, Hadi Ghanbari, and Maryam Roomiani: critically revised the article. All the authors approved the final version and have the agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

ETHICAL APPROVAL

This study protocol was approved by the local ethics committee of the Iran University of Medical Sciences. Informed consent was obtained from the patient before the study.
DATA AVAILABILITY STATEMENT
All data generated or analyzed during this study are included in this submitted article.

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