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Case Report

A case report of Synovial Sarcoma of the Esophagus

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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.

Case Report

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.

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The patient underwent esophagoscopy that revealed a large mass that was 19 cm from 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.

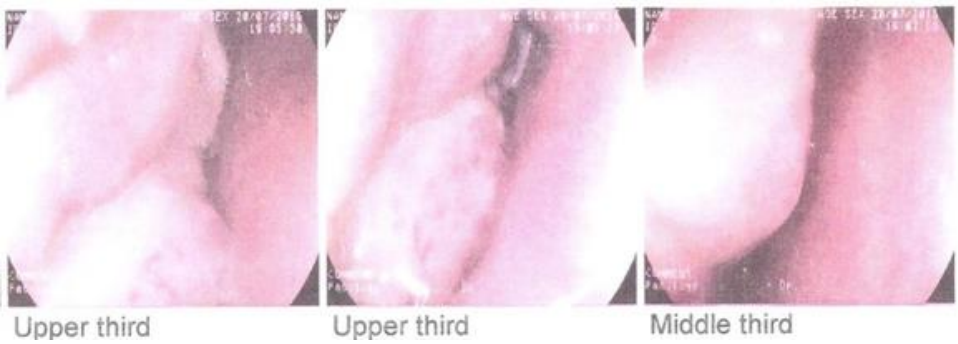
Spiral CT scan of the Neck & Thorax revealed a large intraluminal tumoral mass in the esophagus.

Multiple lymphadenopathies were detected on both sides of the neck.

Abdominopelvis CT scan was normal.

The patient underwent a transhiatal total esophagectomy with gastric pull-up through the posterior mediastinum and cervical esophagogastric anastomosis.

The patient thereafter recieved 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.



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.

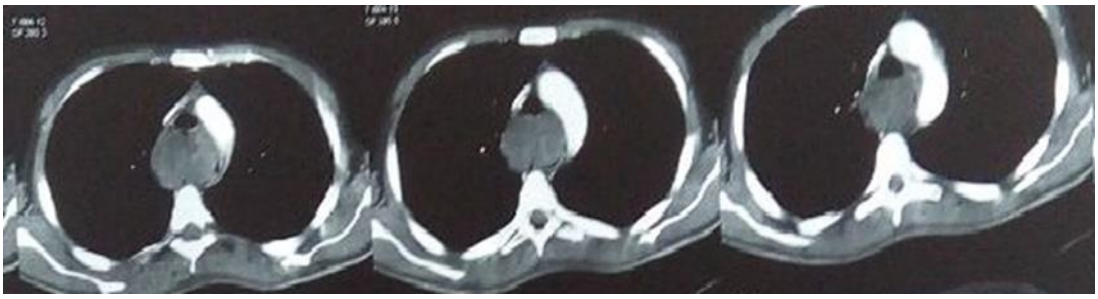
Subcarinal lymph node biopsy is negative for malignancy.

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 are rare neoplasms, particularly in the esophagus and account for 0 .l% to 1.5% of all esophageal tumors (4).

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 component that forms clefts and gland-like spaces, and spindle cell region, 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.

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

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Also, tumor cells show positive staining for the epithelial elements (eg. cytokeratin (AE-1/3) and EMA).

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].

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.

Diagnosis:

These tumors are often presented with polypoid and exophytic masses on Endoscopy [9] and rarely as ulcerating tumours [10].

Tumoral mass may enhance on CT/MRI imaging as intramural mass [11].

Treatment:

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.

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.

Stenting as a Palliative procedure can relieve dysphagia and improve quality of life [14].

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Occasionally, neoadjuvant therapy is also considered based on functional outcomes and other factors.

For unresectable lesions, treatment options include radiation therapy, chemotherapy, or chemoradiotherapy.

It seems synovial sarcoma is relatively chemosensitive according to existing data [24], and chemotherapy has improved survival in primary synovial sarcoma of the extremities [23].

The primary chemo therapy regimen consists of doxorubicin, ifosfamide, and dacarbazine, either alone or in combination.

Today, a number of targeted therapies have been considered for adding to chemotherapy regimen and they have had promising results. For example Pazopanib significantly prolonged median progression -free survival and tended to improve overall survival, although this latter trend was not significant [25].

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The Present Case in the Context of the Literature:

Amr et al. reported a polypoid synovial sarcoma arising in the upper third of the esophagus in a 25-year-old man and did a total esophagectomy followed by radiotherapy. This approach was effective [15].

Bloch et al. reported a case that was treated with a local resection and postoperative radiation therapy and there was no evidence of disease after 36 months [16]. Yokoyama et al. did a clinicopathologic study of 18 cases and concluded that complete surgical resection of tumors less than 5 cm in maximum diameter may be curative and that adjuvant chemotherapy may not be necessary for such patients, because the value of chemotherapy for synovial sarcoma is still controversial. [17].

On the other hand, Pappo et al. concluded that adjuvant chemotherapy is meritted in children with resected synovial sarcoma (clinical group I or II) who present with unfavorable clinicopathologic features such as large, invasive, or grade 3 lesions. [18]

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Holtz et al. reported A 16-year-old boy with Synovial Sarcomas of the Tongue Base. His tumor was resected and Four weeks after surgery, a course of radiation totaling 6,700 rad to the tumor bed was given over eight weeks. The patient died 37 months after the initial surgery and 40 months after the initial symptom with clinical and roentgenographic evidence of metastases to lungs, liver, bone, retroperitoneum, and mediastinum despite receiving multi chemotherapy agents. Holtz believes that the size of the primary tumor is the most critical determinant of the outcome.[19].

S Habo et.al chose a local excision and postoperative radiation and chemotherapy(selected minimally invasive surgical management thus avoiding a total esophagectomy) ; patient has shown no evidence of recurrence or metastasis 20 months postoperatively. They believe that Postoperative radiation and chemotherapy decreases the rate of local recurrence and metastasis [20].

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It has not definitely shown that these systemic therapies for synovial sarcoma of the esophagus are effective. Unfortunately The 2-year survival of esophageal synovial sarcoma is not very good according to the previous studies.

Genetic features and its role in determination of prognosis are under consideration. Definitely, these finding are contributory factors for selecting the best treatment approach for synovial sarcoma of the esophagus

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