International Surgery Journal
Puvvada P et al. Int Surg J. 2024 Sep;11(9):1559-1562
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20242462

Case Report

Giant rare esophageal malignancy in adolescent boy: a case report

Prashanth Puvvada*, Sugaprakash S., Prabhakaran Raju

Department of Surgical Gastroenterology, Madras Medical College, Chennai, Tamil Nadu, India

Received: 24 June 2024
Revised: 17 July 2024
Accepted: 03 August 2024

***Correspondence:**
Dr. Prashanth Puvvada,
E-mail: puvvadaprashanth@yahoo.com

ABSTRACT

Synovial sarcoma is rare soft tissue tumour affecting mostly extremities and less commonly head and neck, lungs, heart and digestive system in adult age group. Only about 20 such cases arising from oesophagus have been reported in literature but none have been reported in adolescent age group with large size in Indian population. So here we present a case report of synovial sarcoma of oesophagus seen in a 14-year-old boy and successful management. Our patient was a 14-year boy who came to our outpatient department (OPD) department with complaints of dysphagia to and retrosternal pain from past 2 months. A contrast enhanced computed tomography (CT) scan showed vertically oriented long and large hypodense lesion arising from right anterolateral wall of pharyngoesophageal junction to upper two-thirds of esophagus from C6 to D8 causing >50% narrowing of trachea. Upper gastrointestinal (GI) endoscopy was done which showed linear submucosal lesion from 17 to 28 cm from incisors. After anaesthetic clearance patient was planned and taken up for minimally invasive esophagectomy. Intraoperatively bulky submucosal growth extending from just cricopharynx to lower thoracic esophagus till inferior pulmonary ligament. Histopathology report came out as synovial sarcoma spindle cell type with Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) grade 2. After tumor board discussion, adjuvant therapy with doxorubicin and ifosfamide was given for five courses. After Six months follow up, the boy is free from tumor recurrence. Synovial sarcoma in esophagus is very rare with nearly 20 reported cases, of which most were polypoid and located in upper and mid esophagus and biphasic histology. The treatment for patients with localized disease is surgery with an adequate wide margin combined with adjuvant chemotherapy and/or radiation is regarded as the preferable treatment. Esophagectomy mostly with minimally invasive approach is preferred surgical option. Robotic minimally invasive surgery is also a safe and feasible option. Clinical follow-up has been limited, and synovial sarcoma is widely known to recur or metastasize after long disease-free intervals.

Keywords: Esophageal sarcoma, Synovial sarcoma, VATS esophagectomy, Minimally invasive esophagectomy

INTRODUCTION

Synovial sarcoma is rare soft tissue tumor affecting mostly extremities and less commonly head and neck, lungs, heart and digestive system in adult age group.[1] Differential diagnosis includes other submucosal tumors which are more common like leiomyoma and gastrointestinal stromal tumor (GIST). Only about 20 such cases of synovial sarcoma affecting esophagus have been reported in literature but none have been reported in adolescent age group with large tumor size in Indian population.[2] So, here we present a case report of synovial sarcoma of esophagus seen in a 14-year-old boy and successful management.

CASE REPORT

Our patient was a 14-year boy who came to our outpatient department (OPD) department with complaints of dysphagia to both solid and liquids and mild retrosternal pain from past 2 months. Physical examination was unremarkable. Routine blood investigations were within normal limits. A contrast enhanced computed tomography

(CT) scan showed vertically oriented long and large hypodense lesion arising from right anterolateral wall of pharyngoesophageal junction to upper two-thirds of esophagus from C6 to D8 causing >50% narrowing of trachea (Figure 1). Upper gastrointestinal (GI) endoscopy was done which showed linear submucosal lesion from 17 to 28 cm from incisors. Barium swallow showed dilated esophagus (Figure 2). Our preoperative diagnosis was giant leiomyoma. Bronchoscopy was also done which was normal.

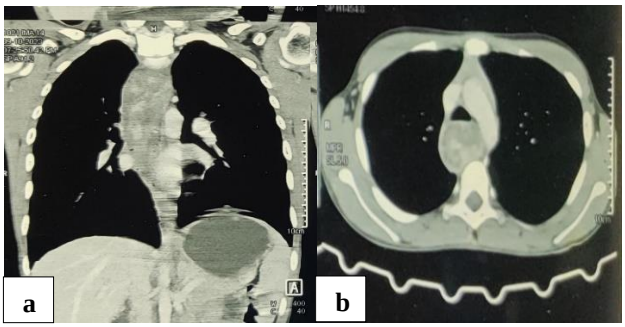

Figure 1 (a and b): CECT showing giant esophageal growth.

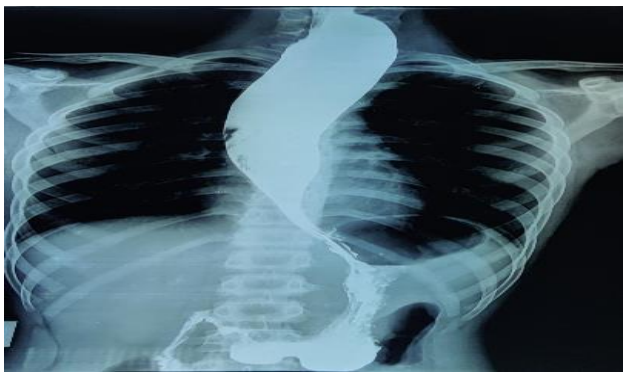

Figure 2: Barium swallow showing dilated and bulky esophagus.

After anaesthetic clearance patient was planned and taken up for minimally invasive esophagectomy. Due to long segment growth esophagectomy was considered instead of enucleation. Intraoperatively bulky submucosal growth extending from just cricopharynx to lower thoracic esophagus till inferior pulmonary ligament (Figure 3). Stomach and rest of abdomen was normal. First thoracoscopic mobilisation of esophagus was done in prone position followed by laparoscopic retrieval of specimen and creation of gastric conduit with posterior mediastinal pullup of gastric conduit in supine position (Figures 4 and 5). Finally, hand-sewn neck anastomosis was done. Patient was gradually started on oral diet after taking oral contrast X-ray and was discharged on post-operative day (POD) 15. Histopathology report came out as synovial sarcoma spindle cell type with FNLCC grade 2 (tumor differentiation-3, mitosis-1, and necrosis-1). Mitotic rate 7-8/10 HPF with 30% necrosis. Margins were free. Immunohistochemistry (IHC) staining showed DOG1 positive in 50% tumor cells, CD 117 negative, CD 34 negative and Ki67 80%. After tumor board discussion, adjuvant therapy with doxorubicin and ifosfamide was then started and planned to be given for five courses. After six months follow up, the boy is free from tumor recurrence.

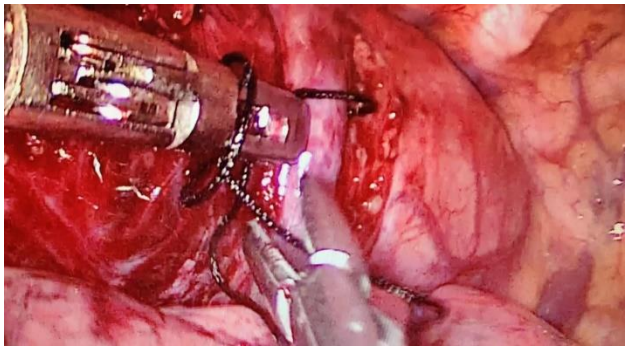

Figure 3: Thoracoscopic picture with azygous vein being ligated for esophageal mobilization.

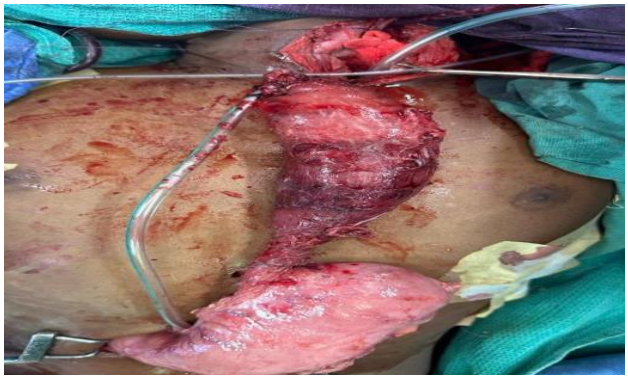

Figure 4: Completion of thoracolaparoscopic mobilization.

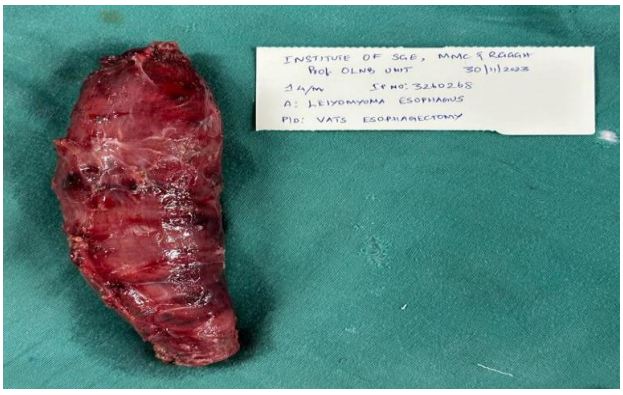

Figure 5: Resected specimen.

DISCUSSION

Fewer than 2% of primary esophageal tumors are mesenchymal in origin; the majority of esophageal sarcomas are leiomyosarcoma.[3] No cellular origin has yet been proven; however, current research suggests that it

might develop from primitive mesenchymal cells or myoblasts.[4,5] Synovial sarcoma in esophagus is very rare with nearly 20 reported cases, of which most were polypoid and located in upper and mid esophagus and biphasic histology.[2] Esophageal sarcoma is usually reported in adult age group. Genetic study has shown that translocation of t (X; 18) (p11.2; q11.2) results in the fusion gene product SYT-SSX which is responsible for pathogenesis of synovial sarcoma.[6] They can be intraluminal as a polypoidal growth and may progress extraluminally into the mediastinum. Symptoms include dysphagia related to intraluminal obstruction, substernal pain, heartburn, or weight loss. Cough, dyspnoea, or other respiratory symptoms may rarely develop from large lesions causing airway obstruction.

Contrast enhanced CT and upper GI endoscopy will usually pick up the submucosal lesion. EUS can also be helpful to identify the layer involved, extent of growth, margins, size. Biopsy preferably EUS guided is indicated in lesions with a diameter greater than 2 cm, or PET avidity to help guide management.

Synovial sarcoma of esophagus has two histological subtypes. The monophasic type consists of only spindle cells alone. The biphasic subtype, which is more common consists of epithelial cells along with spindle cells. Immunohistochemistry is necessary to differentiate from other spindle cell tumors and cells are positive for vimentin, epithelial (EMA, CK7, AE1/3), bcl-2, and neuroectodermal (CD56, CD57, CD99) markers.[6] Also, molecular markers like t (X:18) is a sensitive marker and is demonstrated in 70 to 90% of synovial sarcomas. Novel SS18-SSX fusion-specific antibody provide high sensitivity and specificity for the diagnosis of synovial sarcoma.[6]

Coming to the treatment for patients with localized disease, surgery with an adequate wide margin combined with adjuvant chemotherapy and/or radiation is regarded as the preferable treatment.[7] Esophagectomy mostly with minimally invasive approach is preferred surgical option. Robotic minimally invasive surgery is also a safe and feasible option. The length of stay for minimally invasive surgery is about half that expected for open thoracic surgery. Our case being a giant growth extending from cervical till distal thoracic esophagus, we performed minimally invasive thoracolaparoscopic esophagectomy with gastric pull through followed by neck anastomosis and successfully managed without any complication. Stretched azygous and tracheal narrowing also make minimally invasive esophagectomy a safer option compared to transhiatal esophagectomy.

Adjuvant chemotherapy using doxorubicin and/or ifosfamide is the best treatment option in terms of 5-year overall and disease-free survival in cases of soft tissue sarcoma.[7] For advanced patients with unresectable tumor and metastasis, doxorubicin and ifosfamide may be the front-line chemotherapy of choice.

Clinical follow-up has been limited, and synovial sarcoma is widely known to recur or metastasize after long disease-free intervals. The 5- and 10-year overall survival of 60% and 34%, respectively, for synovial sarcoma.[8] Several factors indicative of a favourable outcome have been reported, including a patient age less than 25 years old, a tumor size less than 5 cm, and the absence of a poorly differentiated component.[9] Singer et al found that synovial sarcomas smaller than 5 cm had 100% 10-year survival.[8]

CONCLUSION

Our patient belonging to adolescent age with synovial sarcoma extending from cervical to distal thoracic esophagus makes it an interesting case report and very rarely reported in literature. Such patients can be managed by minimally invasive esophagectomy especially in minimally invasive era without any complications. As many lesions are misdiagnosed as GIST or leiomyosarcoma and with availability of cytogenetic and molecular techniques, the accurate diagnosis of synovial sarcoma in the digestive tract may increase in the coming years.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Siegel HJ, Sessions W, Casillas MA, Said-Al-Naief N, Lander PH, Lopez-Ben R. Synovial sarcoma: clinicopathologic features, treatment, and prognosis. Orthopedics. 2007;30:1020-5.
2. Sasaki K, Noda M, Tsuruda Y, Uchikado Y, Omoto I, Kita Y, et al. Primary monophasic synovial sarcoma of the cervical esophagus confirmed by detection of the SS18-SSX2 fusion transcripts: case report and literature review. Surg Case Rep. 2020;6:176.
3. Sons HU, Borchard F. Esophageal cancer: autopsy findings in 171 cases. Arch Pathol Lab Med. 1984;108:983-8.
4. Eizinger F. Soft Tissue Tumors. St. Louis. Year Books, Inc: Mosby. 1993.
5. Haldar M, Hancock JD, Coffin CM, Lessnick SL, Capecchi MR. A conditional mouse model of synovial sarcoma: insights into a myogenic origin. Cancer Cell. 2007;11:375-88.
6. Sekaran A, Shinde P, Vanere V, Ramchandani M, Reddy DN. Rare case of primary esophageal synovial sarcoma with (x;18) Translocation presenting as dysphagia. Int J Res Med Sci. 2021;9:1782-6.
7. O'Connor JM, Chacón M, Petracci FE, Chacón RD. Adjuvant chemotherapy in soft tissue sarcoma (STS): a meta-analysis of published data. J Clin Oncol. 2008;26:10526.
8. Singer S, Baldini EH, Demetri GD, Fletcher JA, Corson JM. Synovial sarcoma: prognostic significance of tumor size, margin of resection, and

mitotic activity for survival. J Clin Oncol. 1996;14:1201-8.
9. Garcia-Ortega D, Alvarez-Cano A, Martinez-Said H, Luna-Ortiz K, CaroSanchez C, Cuellar-Hubbe M. Prognostic factors in synovial sarcoma. Experience of 173 cases in a high volume sarcoma center. Eur J Surg Oncol. 2019;45:e156.

Cite this article as: Puvvada P, Sugaprakash S, Raju P. Giant rare esophageal malignancy in adolescent boy: a case report. Int Surg J 2024;11:1559-62.