Case Report

Chondromatous hamartoma of cervical esophagus: a case report and literature review

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Abstract: Esophageal chondromatous hamartomas are very rare tumors, characterized by proliferating hyaline cartilage cells. We described the case of a 64-year-old woman, with a progressive foreign body sensation in the laryngopharynx caused by an intraluminal pedicled mass. Transcervical esophagectomy was performed and intraoperative biopsy revealed the mass was a cervical esophageal chondromatous hamartoma. The patient's postoperative course was uneventful and she recovered well. A review of the literature was conducted, and the symptoms, locations, histopathology findings, treatment methods and follow up results of esophageal hamartoma cases were summarized. We found that pre-operative endoscopic ultrasonography-guided fine-needle aspiration (EUS-FNA) can aid in diagnosis and that aggressive surgical treatment should be recommended for cervical esophageal hamartomas.

Keywords: Chondromatous hamartoma; cervical esophagus; surgical treatment; endoscopic ultrasonography-guided fine-needle aspiration (EUS-FNA)

Submitted Oct 16, 2016. Accepted for publication Jan 11, 2017.
doi: 10.21037/jtd.2017.02.79
View this article at: http://dx.doi.org/10.21037/jtd.2017.02.79

Introduction

Hamartomas are congenital tumor-like malformations that contain a range of heterogeneous structures (1). Esophageal hamartomas are rare and account for approximately 6% of large resected hypopharyngeal and esophageal polyps (2). Esophageal chondromatous hamartomas, which are histologically composed of nodules of proliferating hyaline cartilage and spindle cells, are very rare (3). Only several studies have reported that esophageal hamartomas were chondromatous or contained cartilaginous (4-9) or bony tissue (10,11). We herein present a very rare case of an elderly women with a chondromatous hamartoma in the cervical esophagus.

A literature review was conducted using the PubMed database (up to August 2016). The search terms "esophageal hamartoma" (71 studies) and "chondromatous hamartoma" (102 studies) were used in the search process. The references in the relevant articles and reviews were also scanned for potentially eligible studies.

Case presentation

A 64-year-old woman who had a progressive foreign body sensation in the laryngopharynx for 3 years was admitted to our department. She denied dysphagia, nausea and vomiting, anorexia, weight loss, belching and acid reflux, chest pain, and hemoptysis. The patient had a history of hypertension, and she was receiving antihypertensive treatment. An echocardiogram indicated mild regurgitation in the aortic, mitral, and tricuspid valves, left ventricular diastolic dysfunction, and arrhythmia. Tumor markers and other blood tests showed no abnormalities. The patient had no other significant medical background or family history. A barium test revealed a stricture in the upper esophagus for approximately 2.5 vertebral bodies (*Figure 1A*). Computed tomography (CT) and enhanced CT scans suggested a 2.5-cm soft-tissue mass in the upper esophagus with

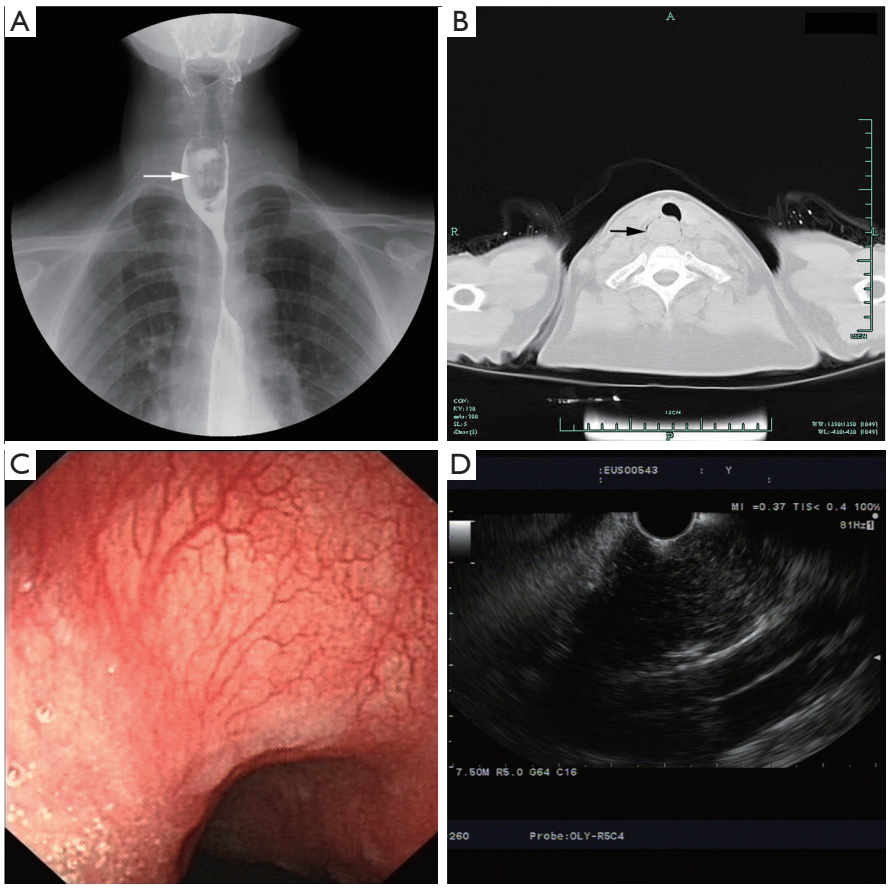

Figure 1 Imaging findings suggest stricture in upper esophagus for approximately 2.5 vertebral bodies. (A) Barium test image; (B) CT image; (C,D) endoscopic ultrasonography suggests prominent lesions and heterogeneous hypoechoic of mass.

inhomogeneous mild enhancement and no obvious lymph node enlargement (*Figure 1B*). The patient underwent endoscopic ultrasonography (EUS), which revealed a giant bulging lesions in the upper esophagus with smooth surface mucosa. The lesion presented with inhomogeneous low-echo areas and multiple high-echo points (*Figure 1C*,*D*). EUS-guided fine-needle aspiration (EUS-FNA) was conducted and the pathology report suggested that the lesion consisted of small pieces of cartilaginous tissue. The multidisciplinary team (MDT) devised a further treatment plan, and the patient underwent trans-cervical esophagectomy after comprehensive evaluation (*Figure 2A*). The patient was placed in a supine position, and general endotracheal anesthesia was induced without complications. The neck was then prepared and draped in the usual sterile fashion. A 15-cm U-shaped incision was made, and the platysma was incised to free the upper esophagus. A longitudinal myotomy was performed, and an intraluminal hard mass was exposed without breaking through the mucosa. A 4.5 cm × 2 cm × 2 cm intraluminal pedicled mass was resected (*Figure 2B*). The incision was sutured layer by layer with 4-0 non-invasive sutures and a negative-pressure drainage ball was placed. An intraoperative biopsy revealed it a cervical esophageal chondromatous hamartoma composed of cartilage cells (*Figure 3*). The patient was stable after surgery, had no obvious complications and was discharged 12 days later. She was regularly followed up for 4 months and had no recurrence of the foreign body sensation.

Discussion

Esophageal hamartoma, which was first described in 1963, is rare (12). To the best of our knowledge, twenty studies with twenty esophageal hamartoma cases (1,4-22) have been reported and eight of these studies described

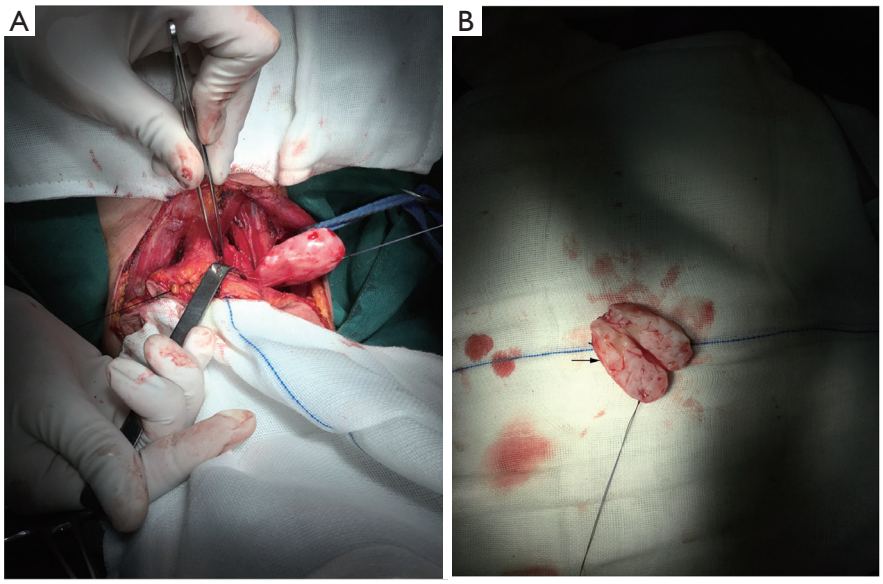

Figure 2 Intraoperative findings. (A) Cervical incision to reveal mass; (B) gross appearance of resected mass (4.5 cm × 2 cm× 2 cm).

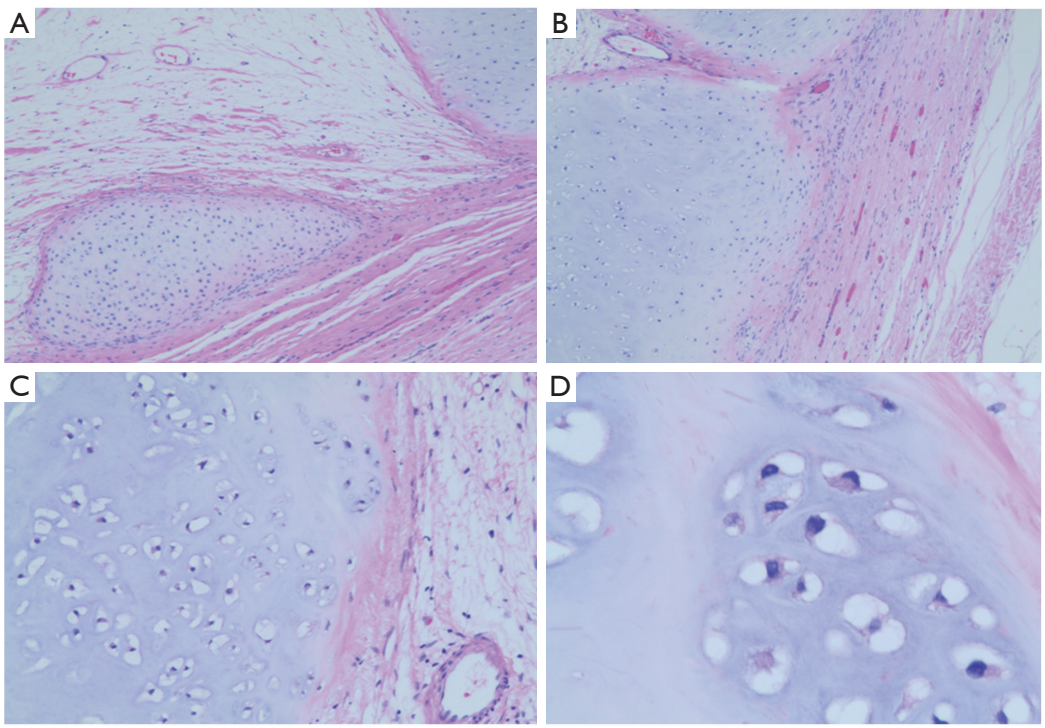

Figure 3 Biopsy results. Chondromatous hamartoma comprised nodules of proliferating hyaline cartilage admixed with areas of spindle cells associated with woven bone. (A,B) Low magnification (50×); (C) medium magnification (100×); (D) high magnification (400×).

Table 1 Study characteristics for esophageal hamartomas.

No.	Author	Year	No. of cases	Age	Sex	Initial symptoms	Location	Histopathology finding	Treatment	Further treatment	Follow up
1	Fuller (12)	1963	1	61 y	Male	Difficulty in swallowing solids	Upper esophagus/ intraluminal	Fibro-adipose connective tissue intermingled with mucous glands and mucous retention cysts	Right lateral esophagectomy	NR	Uneventful (no detailed information)
2	Dieter (13)	1970	1	6 y	Female	Breathing and swallowing difficulty and wheezing on exertion	Lower cervical esophagus	Islets of cartilage and tubules lined by respiratory epithelium, gastric mucosa, ciliated respiratory epithelium and metaplastic respiratory epithelium	Several surgeries were performed. The last surgery: right thoracotomy and thoracic esophagectomy and right cervical esophagectomy	Infection and pneumonia and subsequently healed	With no pulmonary problem in 4 months
3	Shah (4)	1975	1	60 y	Male	Vague mid-chest pain	Cervical esophagus/ intraluminal	Cartilaginous tissue, glandular structures and adipose tissue	Esophagoscope was failed and then left cervical esophagectomy was performed	NR	Uneventful at the time of discharge
4	Smith (5)	1976	1	3.5 y	Female	Stricture of distal esophagus	Distal one third of the Esophagus /Intramural	Small mature lymphocytes, acidophilic histiocytes, cartilage, bronchial-like mucus glands.	Left thoracotomy and end-to-end anastomosis	NR	Uneventful 1 year post-operation
5	Kafai (14)*	1977	1	Children	Male	NR	NR	NR	NR	NR	NR
6	Beckerman (6)	1980	1	11 m	Male	Dysphagia drooling and airway obstruction	Cervical esophagus/ Intraluminal	Skeletal muscle fibers, fibrous connective tissue, and hyaline cartilage.	Neck incision to get biopsy, tracheostomy and gastrostomy to relieve airway obstruction, and completely resected tumor surgically	Removed gastrostomy tube 4 weeks after surgery	Uneventful post-surgery

Table 1 *(continued)*

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No.	Author	Year	No. of cases	Age	Sex	Initial symptoms	Location	Histopathology finding	Treatment	Further treatment	Follow up
7	Everett (7)	1980	2	3.5 y	Female	Episodes of vomiting, intolerance of solid foods, lack of weight	Distal esophagus/ intraluminal	Cartilage, glandular elements, lymphoid tissue, smooth muscle and stromal elements	Surgically resected and end-to-end esophageal anastomosis	NR	NR
				8 m	Male	Vomiting and weight loss	Distal esophagus/ intraluminal	Cartilage, glandular elements, smooth muscle and stromal elements	Esophageal myotomy	NR	NR
8	Venn (15)	1985	1	52 y	Male	Dry cough and progressive dysphagia.	Upper esophagus/ intraluminal	Polypoid esophageal hamartoma	Thoracotomy and esophagectomy	NR	Uneventful at the time of discharge
9	Gupta (16)	1987	1	10 y	Male	Increasing difficulty in respiration and dysphagia	Upper end of esophagus/ Intraluminal	Adipose tissue, fibres of skeletal tissue, collagen tissue and moderate vascularity.	Cervical esophagectomy	NR	Uneventful at the time of discharge and no recurrence at 1 year follow up
10	Saitoh (8)	1990	1	40 y	Female	Persistent bloody sputum	Cervical esophagus/ Intraluminal	Osseous, cartilaginous, fibrous, adipose tissue and glandular structure.	Transcervical esophagotomy	NR	Uneventful
11	Lakhkar (17)	1991	1	30 y	Female	Progressive dysphagia	Upper esophagus/ Intraluminal	Squamous epithelium, hyperplastic and cystic glands with abundant well vascularized stroma.	Esophagotomy	NR	Uneventful and recovered

Table 1 *(continued)*

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No.	Author	Year	No. of cases	Age	Sex	Initial symptoms	Location	Histopathology finding	Treatment	Further treatment	Follow up
12	Halfhide (10) [Ginai,1998 (11)][#]	1995	1	41 y	Male	Progressive dysphagia	Upper esophagus/ Intraluminal	Fat cells, fibrous tissue, atypical blood vessels and a large solitary island of bone enclosed by periosteum; lipomatous, hemangiomatous, and bony structure of the hamartoma	Endoscopic resection by snare coagulation.	NR	Uneventful and recovered
13	Song (18)	2004	1	30 y	Male	Dysphagia	Middle esophagus/ intraluminal	Fibrous connective tissue, smooth muscle tissue and fat tissue	Surgically removed	NR	NR
14	Xu (19)	2008	1	45 y	Male	Foreign body sensation	Middle-lower esophagus/ intraluminal	Esophageal hamartoma characterized in hemangioma	Endoscopic resection	NR	Uneventful at 13 months follow-up
15	Coury (20)	2010	1	8 d	Female	Apnea and cyanosis	Cervical esophageal	Hamartomatous polyp	Microdirect laryngoscopy and esophagoscopy	NR	Uneventful at 6-year follow-up
16	Hou (9)	2012	1	33 y	Male	Dysphagia and Abdominal distention	Lower esophagus	Cartilage like tissue, Fibrous connective tissue and glands	Surgically removed	NR	NR
17	Wu (1)	2014	1	40 y	Male	Epigastric pain after drinking	Lower esophagus	NR	Refused treatment	NR	NR
18	Yorita (21)	2015	1	51 y	Male	Dysphagia	Lower esophagus	Submucosal gland duct	Endoscopically removed	NR	NR
19	Zhao (22)	2015	1	60 y	Male	Dysphagia	Upper esophagus	Fibers, blood vessels, lymphatic and striated muscles and nerves	Open surgery	NR	NR

*, the full text was not available; [#], the case reported by the two authors was the same. NR, not reported.

nine cases (4-11) of esophageal chondromatous hamartomas or that contained cartilaginous or bony tissue. Four cases (4,6,8,10,11) were cervical or upper esophagus hamartomas [Halfhide *et al.* (10) and Ginai *et al.* (11) reported the same case], and five cases (5,7,9,13) were in distal or lower esophagus [one (9) was in Chinese, *Table 1*]. Several other studies (23-32) also reported congenital esophageal stenosis caused by tracheobronchial remnants. Generally, most chondromatous hamartomas are located in the lung parenchyma (33), and although this type of tumor is very rare in the esophagus, it is the most common benign lung tumor and third most common pulmonary nodule (34). Esophageal chondromatous hamartomas are very rare in terms of both their anatomical location and their pathological components. In this case, the clinical manifestation and auxiliary examination were not specific, and EUS-FNA played an important role in preoperative diagnosis of the hamartoma. Xu *et al.* (19) reported that EUS was superior to other imaging techniques, including barium esophagogram, contrast-enhanced CT image and conventional gastroscopy, in the diagnosis and treatment of hamartoma. Indeed, EUS can provide helpful diagnostic accuracy and comprehensive pre-operative assessment of submucosal tumors of the esophagus, including more detailed information about the tumor size, layer of origin, morphologic features, and even histopathological components (35). Thus, clinicians could design further treatment plans for patients with submucosal tumors of esophagus based on the EUS-FNA results.

The distinctive histologic feature of a chondromatous hamartoma is the hyaline cartilage, which is located in cartilage lacunae and surrounded by a thin fibrous perichondrium. However, highly differentiated chondrosarcoma, which featured with lobulated tumor cells with less atypia, should be taken in to consideration for different diagnosis. In our case study, the mass was covered with an integrated capsule and was composed of cartilage cells that were well differentiated, and our hospital's pathology team had taken chondromatous hamartoma into prior consideration.

There are no standard treatment instructions for esophageal hamartoma. Most of the previously reported patients underwent surgical treatment, including esophagectomy and thoracotomy, while one underwent endoscopic resection by snare coagulation (10), two underwent routine endoscopic resection (19,20) and one refused operation (1). Ginai *et al.* (11) suggested that the resection method depends on the polyp's volume and site of origin. When the pedicle is thin and the polyp is relatively small, endoscopic ligation is preferred. If there is an increased risk of bleeding, cervical esophagectomy should be performed. However, Xu *et al.* (19) suggested that gastroscopic resection is more feasible and the selection of different treatment methods should be based on the depth of tumor invasion. In our case, the patient was carefully evaluated and transferred to our department from the department of gastroenterology, and it was discussed during the MDT consultation before surgery that the tumor might have malignant potential. For conservative considerations and to avoid perforation and bleeding from an endoscopic resection, we performed transcervical esophagectomy. According to the limited report information, no further post-surgical treatment was given for these esophageal hamartoma cases, and all patients were discharged uneventfully and recovered with no recurrence.

Hamartomas are uncommon congenital abnormalities that are usually asymptomatic and grow slowly. In this case, the patient complained of a progressive foreign-body sensation in the laryngopharynx for 3 years. However, it can be speculated that the hamartoma might have existed for many years and grown slowly until it was large enough to cause symptoms. Previous studies suggested that esophageal hamartoma was benign, however, obstruction and pulmonary complications can cause fatal events. Thus, aggressive surgical treatment should be recommended because it can significantly improve the patients' status and is nearly curative (5).

In conclusion, we report a very rare chondromatous hamartoma of the cervical esophagus. Pre-operative EUS-FNA can aid in diagnosis and surgical treatment should be recommended for cervical esophageal hamartomas.

Acknowledgements

Funding: This work was funded by Major science and technology projects of Zhejiang province (2014C03032) and General research program of medical and health in Zhejiang Province (2015KYB142).

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

Informed Consent: Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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Cite this article as: Zhang C, Xu J, Wang Z, He Z, Yang H, Hu J. Chondromatous hamartoma of cervical esophagus: a case report and literature review. J Thorac Dis 2017;9(3):E236-E244. doi: 10.21037/jtd.2017.02.79