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Esophageal Duplication Cyst Complicated with Intramural Hematoma

– Case Report –

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Esophageal duplication cysts account for a very small percentage of benign esophageal tumors and are infrequently symptomatic. Esophageal duplication cysts result from aberrant alignment of the normal vacuolization process that produce the esophageal lumen in the 5th to 8th week of embryonic life. Complications most often are bleeding into or infection of cysts. Recently, we experienced a case of esophageal duplication cyst complicated with intramural huge hematoma and the cause of hematoma could not be identified. We report it with a review of literatures.

Key Words : *Esophageal duplication cyst, intramural hematoma.*

INTRODUCTION

Esophageal cystic lesions of foregut origin account for approximately 10% of lesions presenting as mediastinal tumor(Morrison, 1958 ; Kirwan et al., 1973). This lesion is classified into three main categories based on embryology, location and histopathologic composition – esophageal duplication cyst, bronchogenic cyst, enteric cyst(Kirwan et al., 1973 ; Arbona et al., 1984 ; Bremer, 1942). Esophageal duplication cysts are a quite uncommon form of congenital developmental cyst which was first reported by Blassium in 1711. About 50 cases had been reported in the English literatures(Arbona et al., 1984 ; Rhee et al., 1988 ; Dresler et al., 1990 ; Kaneko et al., 1989 ; Nakahara et al., 1990 ; Rafal et al., 1991). In Korea, three cases have been reported in the literature(Kim et al., 1989 ; Hur et al., 1988 ; Hong et al., 1981). Generally these cysts are asymptomatic and are usually discovered as mediastinal tumors in routine chest X-Rays or incidentally found on esophagograms(Kirwan, 1973 ; Arbona et al., 1984).

We experienced a case with esophageal duplication cyst complicated with intramural hematoma in a 49-year-old man.

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CASE REPORT

A 49-year-old man was admitted to hospital via the emergency room because of retrosternal pain and dysphagia. He had been treated by oral hypoglycemic agent due to diabetes mellitus since 2 years previously. He had been relatively well until 1 day before admission, when he suffered acutely from retrosternal pain and dysphagia two hours after dinner. These were not associated with hematemesis or vomiting and not radiated. The family medical history was not contributory. On admission, he was alert but in acute distress. Blood pressure was 150/90 mmHg, pulse rate 82/mim,

respiratory rate 24/mim, temperature 36℃. Physical examination was completely unremarkable. Laboratory studies included hemoglobin 15.1 gm/dl, hematocrit 44.6%, WBC 16,100/mm^3 with 83% neutrophils and 13% lymphocytes, platelets 198,000/mm^3, ESR 58 mm/hr, normal bleeding time, normal prothrombin time, normal aPTT, ALT 59 IU/L, AST 20 IU/L, alkaline phosphatase 59 IU/L, total protein 6.7 gm/dl, albumin 3.8 mg/dl, total bilirubin 0.9 mg/dl, direct bilirubin 0.1 mg/dl, total cholesterol 139 mg/dl, BUN 12 mg/dl, creatinine 1.2 mg/dl, HbA_1C 5%, fasting blood sugar 94 mg/dl, pp2hr glucose 146 mg/dl. Urinalysis and stool examination were normal. EKG and echocardiogram were completely normal.

Routine chest radiography showed normal lungs but mediastinal widening(Fig. 1). An esophagogram revealed an extrinsic mass effect on the esophagus from 3cm below the carina to the esophagogastric junction, causing compression of its right posterolateral aspect and leftward displacement. No evidence of obstruction was found(Fig. 2). On chest CT, an elongated mediastinal mass was found inseperable from the esophagus from the aortic arch level to the esophagogastric junction. A cystic mass was located in the lower esophagus, lateral side of the elongated mediastinal mass(Fig. 3). On admission, endoscopic examination revealed a bulging lesion into the lumen of the esophagus 20cm from the incisor to the gastroesophageal junction. The bulging side mucosa was smooth, normal pinkish color, not eroded but some portion of the oral side showed a slightly bluish coloring(Fig. 4). The endoscope was advanced carefully into the stomach, which was completly normal.

Three day later endoscopic examination was followed up. The bulging lesion was not changed in size or location, but mucosal color was deeply bluish as bruise(Fig. 5). Five days later, we performed an endoscopic examination for a biopsy, which revealed more blue colored mucosa. Blood was not aspirated in needle aspiration of the bulging lesion. Endoscopic ultrasonography revealed a low inhomogeneous echo density mass inseperable from the esophagus and did not show precise relation with three walls of the esophagus(Fig. 6). It was assumed that the patient had an esophageal cyst with intramural hematoma. A right posterolateral thoracotomy was performed through the 5th intercostal space. Pleural cavity and lung parenchyme were normal except for some adhesion of the pulmonary apex. Bluish discoloration was noted in mediastinal pleura, which was adherent extensively to the esophagus. A cyst measuring approximately

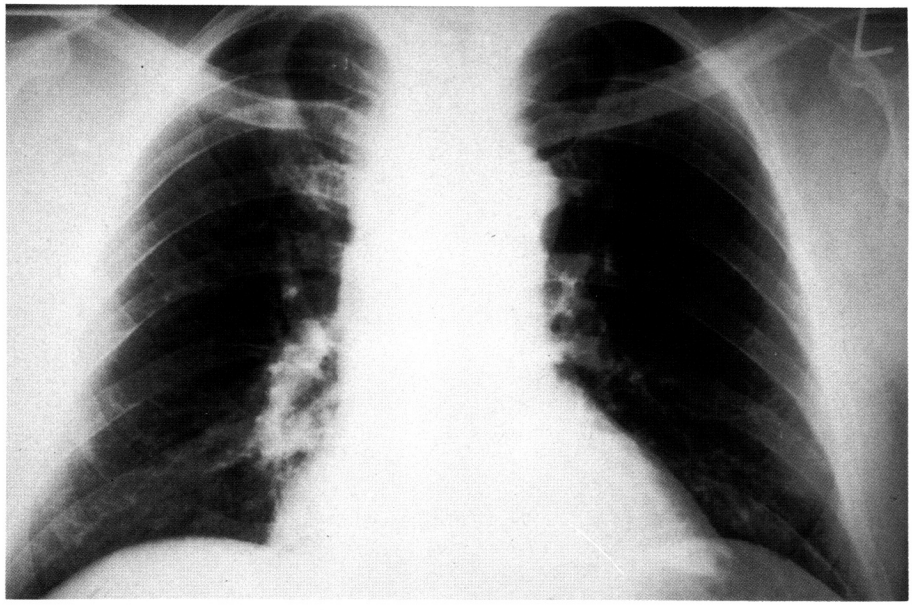

Fig. 1. Chest PA shows mediastinal widening.

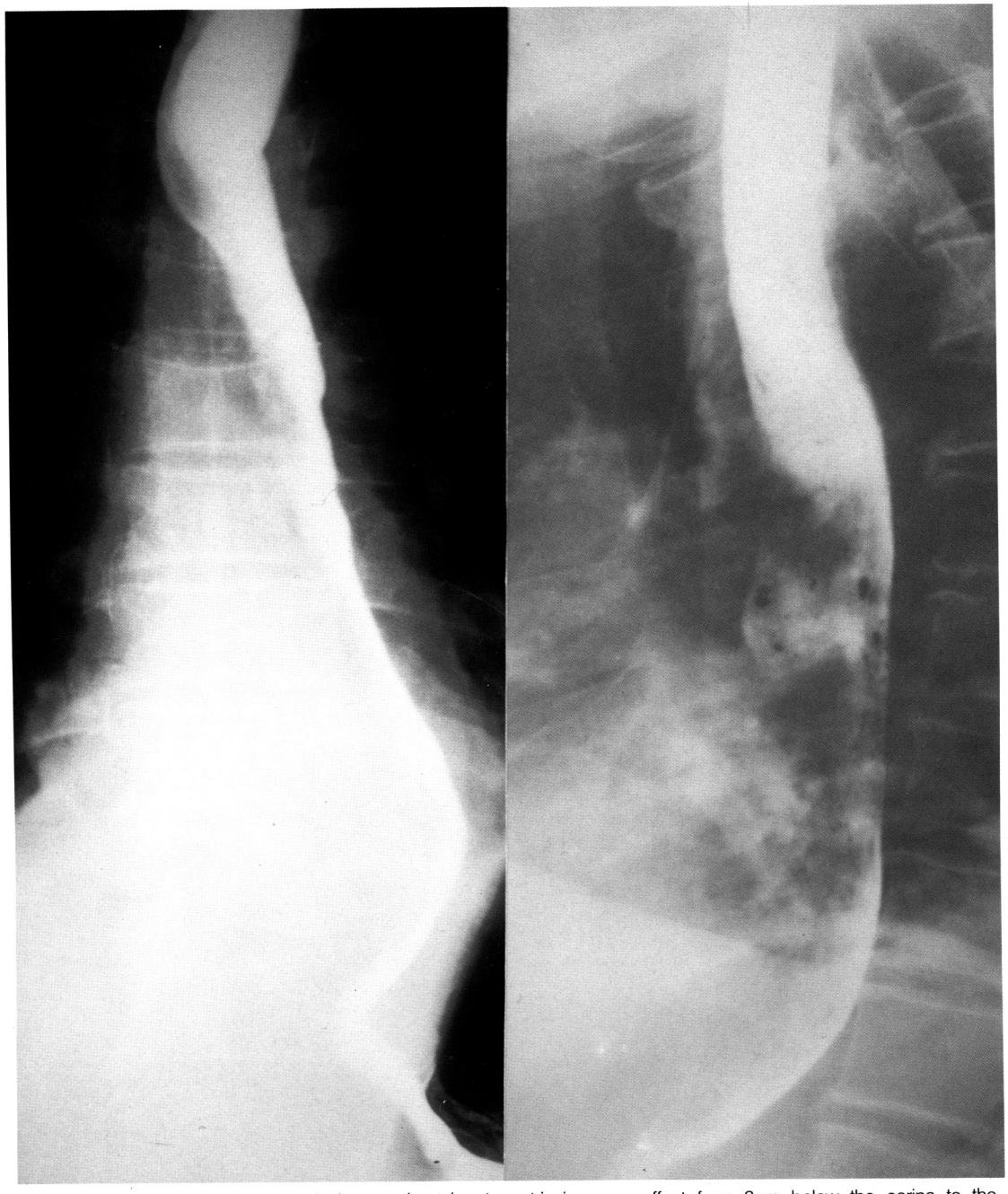

Fig. 2. Esophagogram shows luminal narrowing due to extrinsic mass effect from 3cm below the carina to the esophagogastric junction,causing compression of the posterior right lateral aspect.

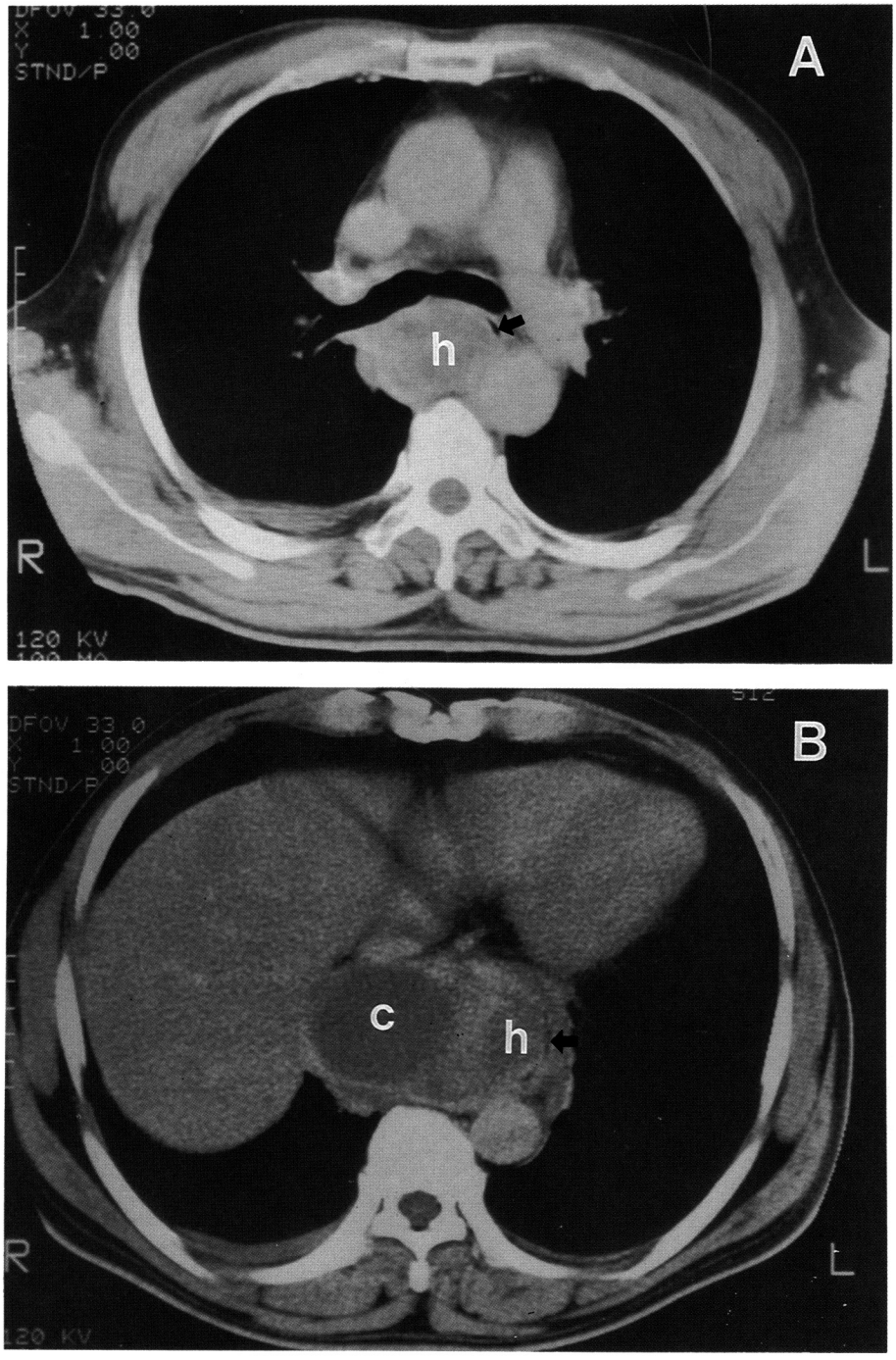

Fig. 3. (A) Chest CT shows a round elongated inhomogeous mediastinal mass(h) inseparable from the esophageal wall that compresses the esophageal lumen(black arrow)
(B) In the lower esophagus, a homogenous cystic mass(c) is found adjacent to the lateral side of the elongated mediatinal mass(h)

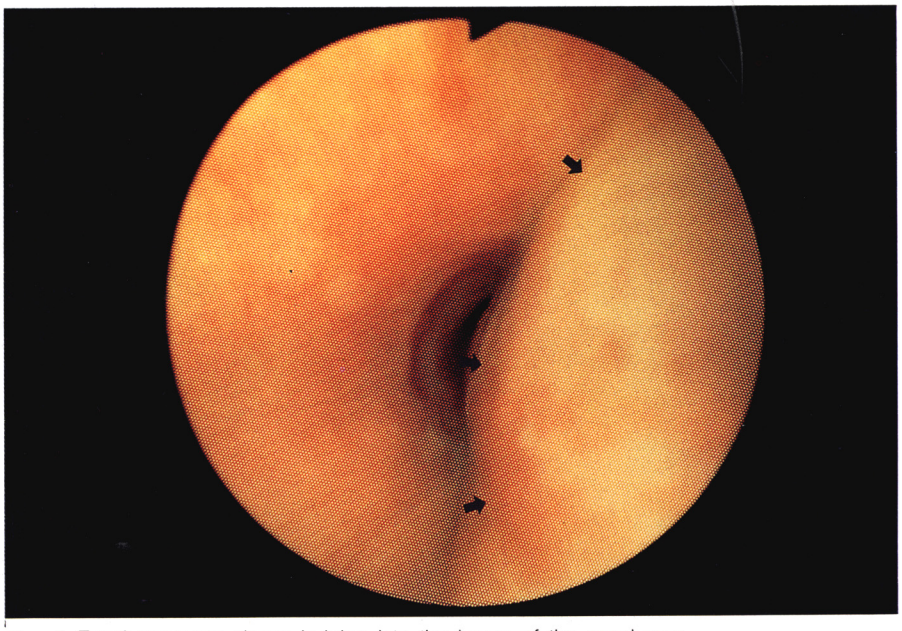

Fig. 4. Esophagoscopy shows bulging into the lumen of the esophagus. Bulging mucosa was smooth, normal pinkish color, not eroded.

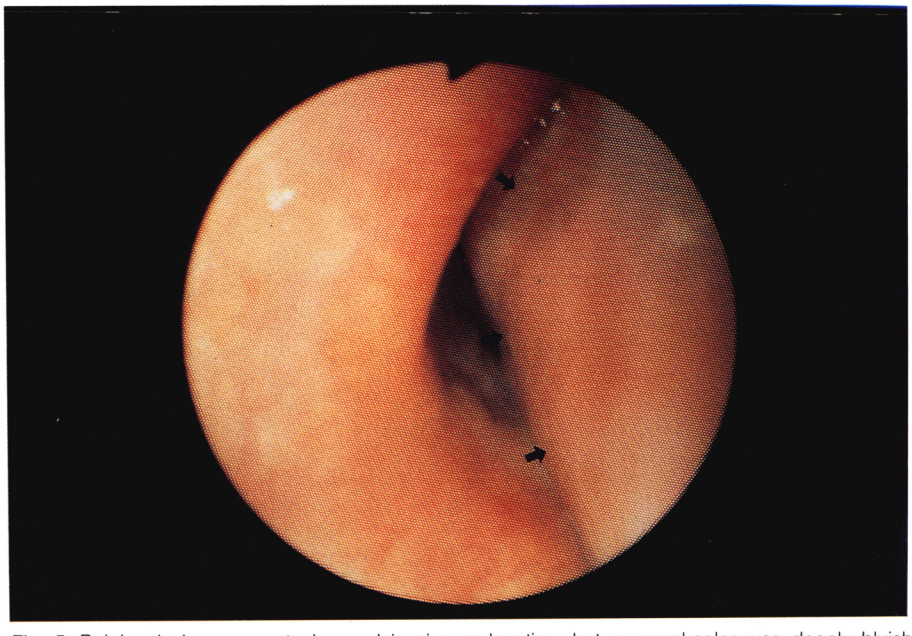

Fig. 5. Bulging lesion was not changed in size or location, but mucosal color was deeply bluish.

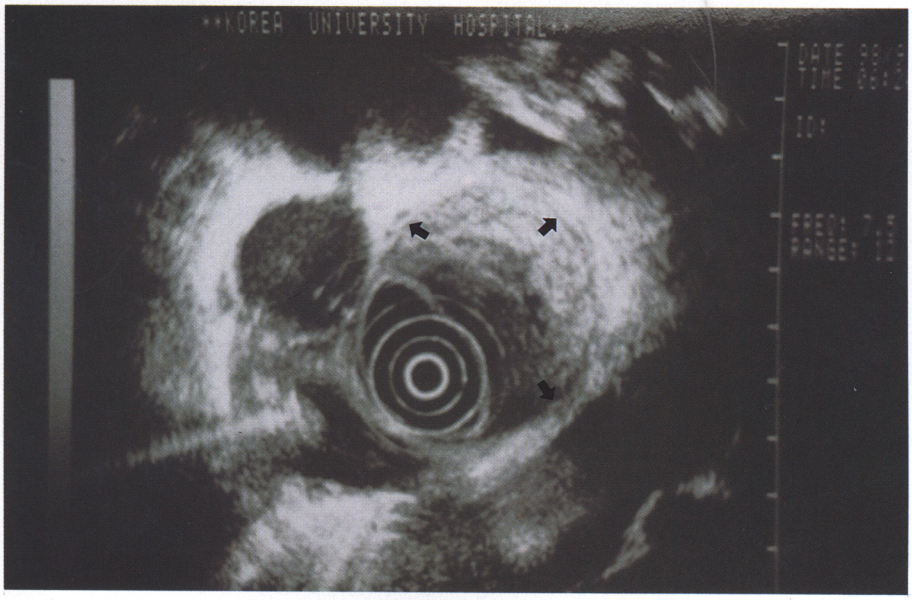

Fig. 6. Endoscopic ultrasonography shows a low inhomogenous echo-density mass in the esophageal wall, that was a hematoma(arrow).

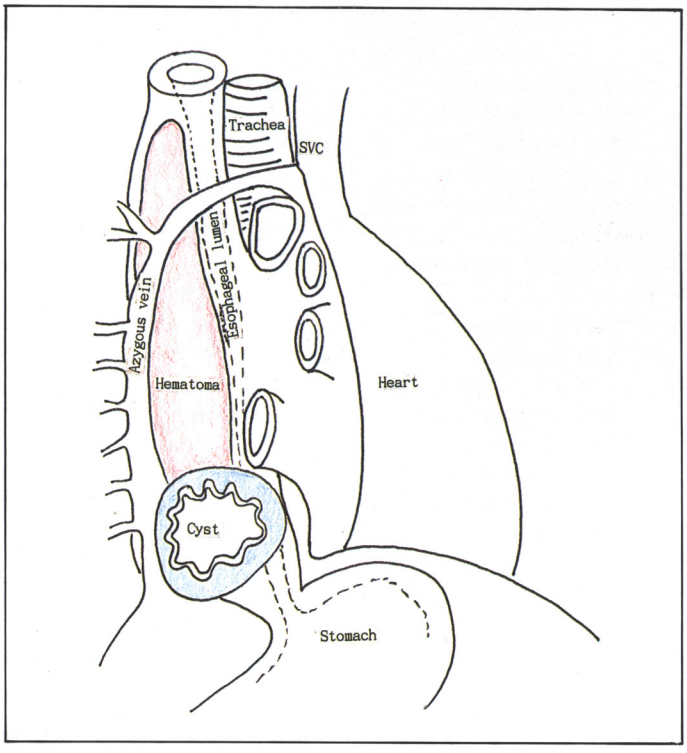

Fig. 7. Schematic drawing of the esophageal cyst and intramural hematoma.

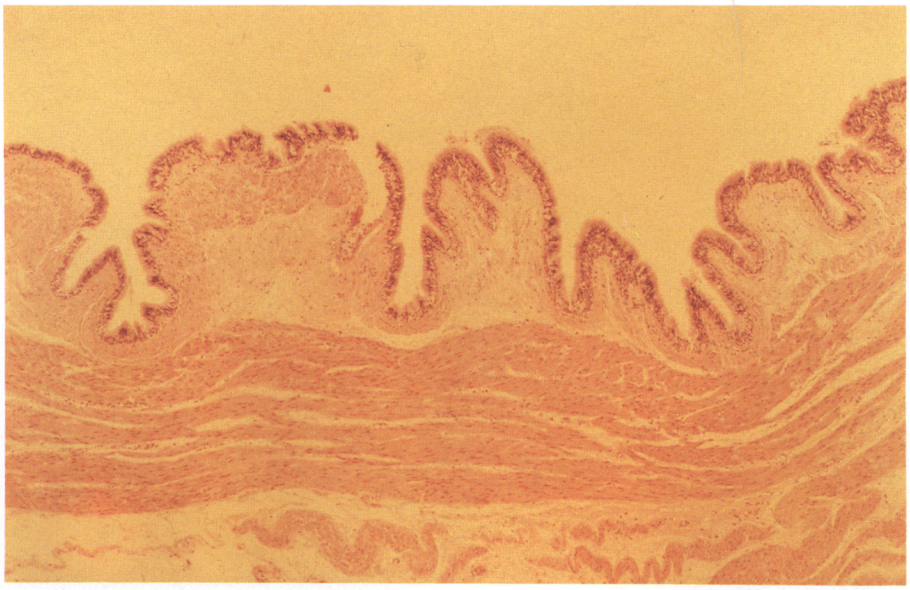

Fig. 8. Microscopic examinations show that the cyst was lined by pseudostratified ciliated columnar epithelium surrounded by smooth muscle.

5X4X3cm was noted intramurally just above the diaphragmatic hiatus(Fig. 7). On aspiration, it revealed approximately 30cc of chocolate colored viscid fluid. The esophagus below the lower margin of the cyst was normal, but above the upper margin of the cyst was bluish bulging out upto the thoracic inlet. The muscle layer was incised longitudinally, and the hematoma was evacuated. The cyst was shelled out from the esophageal wall without opening to the esophageal mucosa.

Histopathologically the cyst was lined by pseudostratified ciliated columnar epithelium surrounded by smooth muscle without cartilage. So it was diagnosed as an complicated esophageal duplication cyst with intramural hematoma(Fig. 8).

Postoperatively, a follow up esophagogram was unrevealing and The patient discharged on postoperative day 12 without complication.

DISCUSSION

Esophageal cysts represent rare benign tumor of congenital foregut anomalies(Morrison, 1958 ; Kirwan et al., 1973).

The classification and terminology used to describe esophageal cysts has been sometimes confused according to anatomical site, embryological origin and histopathologic composition, but usually they were classified as esophageal duplication cysts, bronchogenic cysts and enteric cysts(Kirwan et al., 1973 ; Arbona et al., 1984 ; Bremer, 1942). Embryologically, after the fourth weeks the embryonic foregut elongated fairly rapidly. The lining epithelium proliferates, converting the esophagus into an almost solid tube. At about six weeks, vacuoles develop within this solid tube. These vacuoles gradually coalesce to form the esophageal lumen. One vacuole may persist, however, giving rise to an esophageal duplication cyst. Due to elongation of the intrathoracic viscera and dextrorotation of the stomach, these cysts are frequently found in the inferior portion of the esophagus and on its right side.(Kirwan et al., 1973 ; Arbona et al., 1984).

Esophageal cysts are usually asymptomatic but some clinical manifestations are represented by size and location(Kirwan et al., 1973 ; Arbona et al., 1984 ; Vithespongse et al., 1971 ; Whitaker et al., 1980 ; Desforges et al., 1960). Arbona et al(1984) mentioned that 35% of esophageal duplications were noted to be asymptomatic, but dysphagia, epigastric discomfort, retrosternal pain, dyspnea,

regurgitation and cough have also been reported. When symptoms occur, they are secondary to inflammation or infection, at which time they increase in size and cause pain, distortion and malfunction of the esophagus at the level of the cysts(Vithespongse et al., 1971). In our patient, sudden onset of retrosternal pain and dysphagia might have been due to dissection of esophageal wall by hemorrhage in the vicinity of the cyst.

The location of cysts were as follows : 60% were located in the lower third of the esophagus, 17% in the middle third, and 23% in the upper third(Arbona et al., 1984). Duplication cysts in the upper third are generally diagnosed in infancy, due to the respiratory symptoms, dysphagia and impairment of growth(Arbona et al., 1984 ; Rhee et al., 1988).

A male predominance in a ratio of 2 : 1 was mentioned by Arbona et al.(1984) in case review up to 1984 and some male predominance has been noted in recent literature reviews(Rhee et al., 1988 ; Dresler et al., 1990 ; Kaneko et al., 1989 ; Nakahara et al., 1990 ; Rafal et al., 1991).

Diagnosis was usually incidental. Radiological studies of the chest and esophagus are helpful in making a diagnosis. On chest X-rays, they show as a mediastinal mass. Esophagograms often reveal a round, soft, filling defect covered by apparently normal mucosa(Arbona et al., 1984 ; Whitaker et al., 1980). Schatiki et al.(1942) emphasized that an esophagogram could not be used to differentiate a cyst of the esophagus from other benign intramural tumors. However, Mansour et al.(1977) suggest that the absence of a sharp "step" effect of the superior and inferior margins on the barium swallow is more characteristic of the duplication cyst as opposed to a leiomyoma.

Esophagoscopy shows the smooth round shape of the soft compressible submucosal tumor with no mucosal alteration. Biopsy through endoscopic examination in most cases does not advised since it may not reach the lesion and might cause mucosal ulceration and initiates an inflammatory process(Arbona et al., 1984 ; Vithespongse et al., 1971 ; Mansour et al., 1977 ; McHardy et al., 1971). In our case, endoscopic examination revealed an elongated bulging lesion in which mucosal color became bluish as bruise with time, so that bulging lesion was suspected as esophageal hematoma. The CT scans provide accurate assessment of the location, size of the lesion and neighboring anatomic structure(Kim et al., 1989 ; Rhee et al., 1988 ; Dresler et al., 1990 ; Rafal et al., 1991). The CT scan is very useful in confirming the underlying cyst in the complicated cyst with overlaid intramural hematoma as this case.

Recently, magnetic resonance image has proven to be of value in predicting the nature of cystic contents as well as outlining the anatomic relationship(Rhee et al., 1988 ; Rafal et al., 1991). 99m-sodium pertechnetate is beneficial in diagnosis of duplication with ectopic gastric mucosa(Kim et al., 1989 ; Whitaker et al., 1980). The average size was 4cm in diameter(Arbona et al., 1984). Esophageal duplication is usually in cyst form but may be of tubular structure(Arbona et al., 1984 ; Rafal et al., 1991). The majority of these duplications are closed, but less than 10% communicate with the normal alimentary tract(Dresler et al., 1990).

Complications may be infections, rupture or bleeding(Kirwan et al., 1973 ; Whitaker et al., 1980 ; Desforges et al., 1960 ; Nakahara et al., 1990 ; Gantzinsky et al., 1978). Kirwan et al.(1973) reported that seven cases developed complications, 1 peptic ulceration, 5 infections, 1 tracheal obstruction. Infection is the most common complication. One infected cyst ruptured into the pericardial space, which came to cardiac tamponade. Whitaker et al.(1980) reported an intracystic hemorrhage case due to infection. Gatzinsky et al.(1978) reported an esophageal cyst with massive mediastinal hemorrhage which was managed by esophagectomy but they emphasized that esophagectomy was not necessary in management of esophageal cyst. In our case hemorrhage confined within esophageal wall in vicinity of the cyst. The pathogenesis of the bleeding in the present case is obscure. Malignant degeneration of these cysts is extremely rare and has been reported only two in the literature(Arbona et al., 1984).

The only definite treatment for esophageal cyst is surgical excision. Surgery must be considered in every case even though it may be asymptomatic due to the potential for ulceration and perforation(Arbona et al., 1984 ; Vithespongse et al., 1971 ; Dresler et al., 1990). It is rarely necessary to open the mucosa, and the esophageal wall can be satisfactorily reconstructed without resection(Arbona et al., 1984).

In Korea, 3 cases have been reported, and all cases were asymptomatic men, found incidentally on routine chest radiography as a mediastinal mass without complication. They were located in the up-

per two thirds of the esophagus, and treated by surgery(Kim et al., 1989 ; Hur et al., 1988 ; Hong et al., 1981).

This report is a case of an esophageal duplication cyst complicated with intramural hematoma and the definitive cause of hematoma could not be identified. Our case may be the first report of an intramural hematoma with a duplication cyst.

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