Case presentation
Achalasia

Reporter:R4梁美娟

Supervisor:VS.連漢仲

Basic data

- Age:74
- Sex:female
- BW:53.8 Kg
- Cigarette smoking :denied
- Alcohol consumption : denied

Past History

- Achalasia s/p myotomy in 1985 in TCVGH
  Antral gastritis with Hp infection
- 910924 EGD: Esophageal dilatation, gastritis
  1010315 EGD: Esophageal dilation, gastritis
- Pneumatic dilatation for once

Chief complaint (110.04)

- Intermittent dysphagia to solid meal for 1-2year
- Epigastralgia in recently.

Physical examination

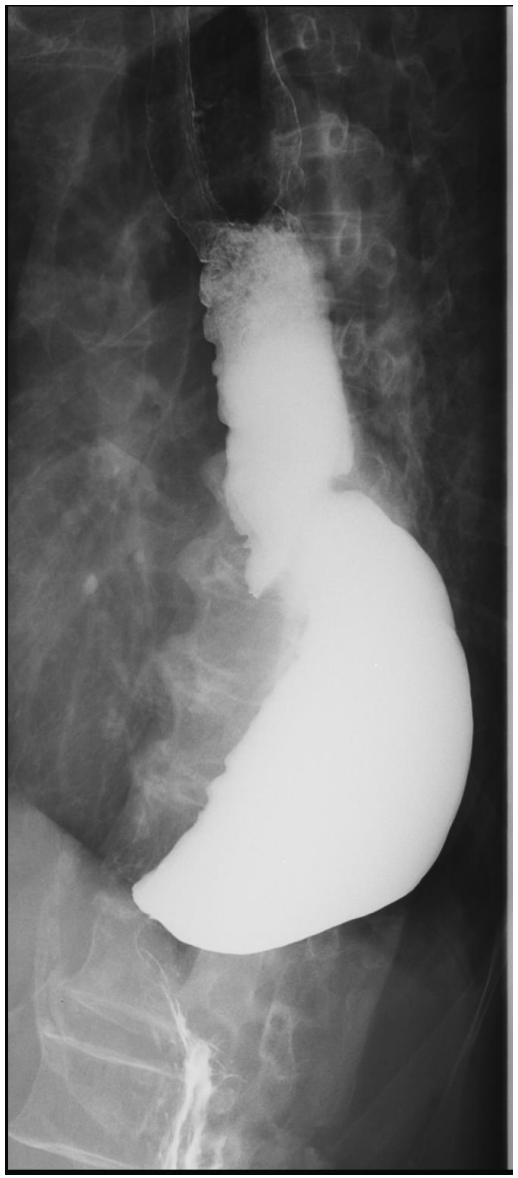
- NO BWL, BW 53.8 KG
- RSI: 6, ECKARDT SCORE: 2

ESOPHAGOGRAPHY on 110.04.30

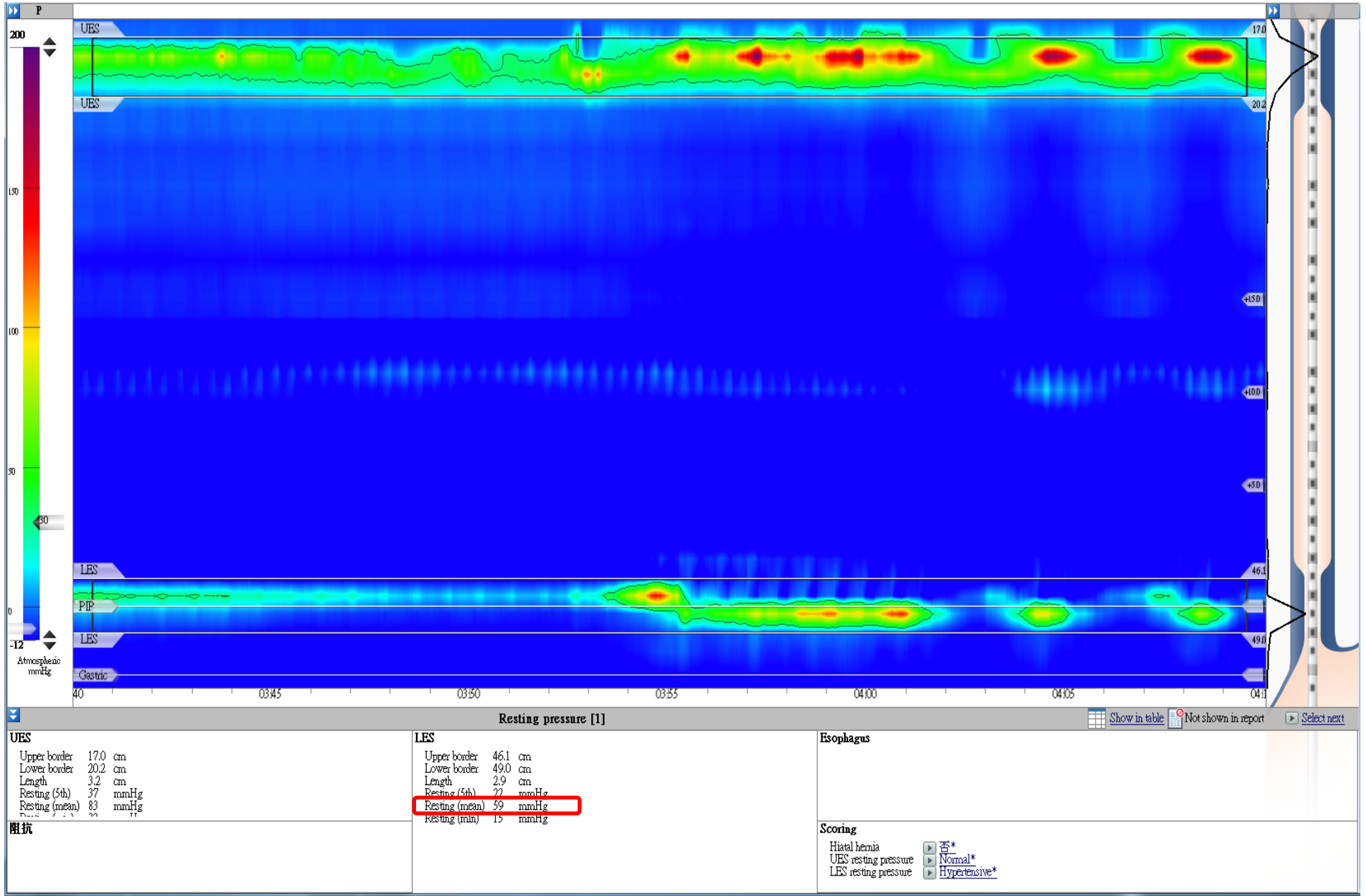
Finding:
Esophagography was performed under fluoroscopic control with swallowing of effervescent powder and barium meal by the patient.

The findings are:
> A case of achalasia s/p balloon dilatation. For survey.
> Preliminary film shows calcifications of thoracic aorta
> Under fluoroscopy, the deglutition, pharyngeal function, coordination of larynx and epiglottis are not unusual.
> Bilateral valleculae and pyriform sinuses appear symmetric.
> Obvious stasis of contrast medium from esophagus into stomach and dilatation and tortuous of esophagus are seen. Post-treatment appearance of achalasia is favored.
> The EG junction is widening.
> Except for normal indentation of aorta and left bronschus on esophagus, no obvious mass effect over esophagus.
> No choking noted during study.

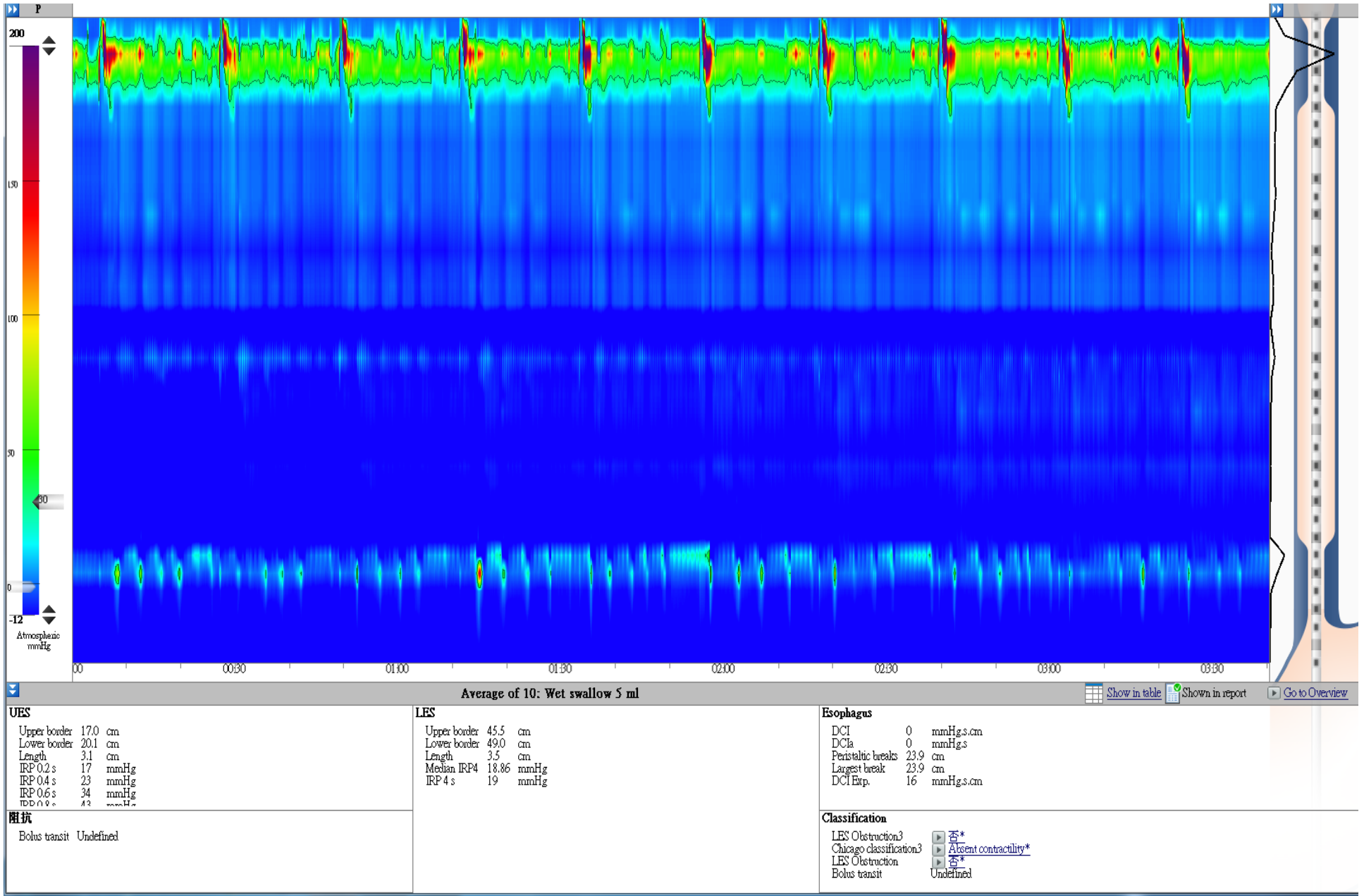
Impression
Achalasia s/p balloon dilatation with contrast media stasis



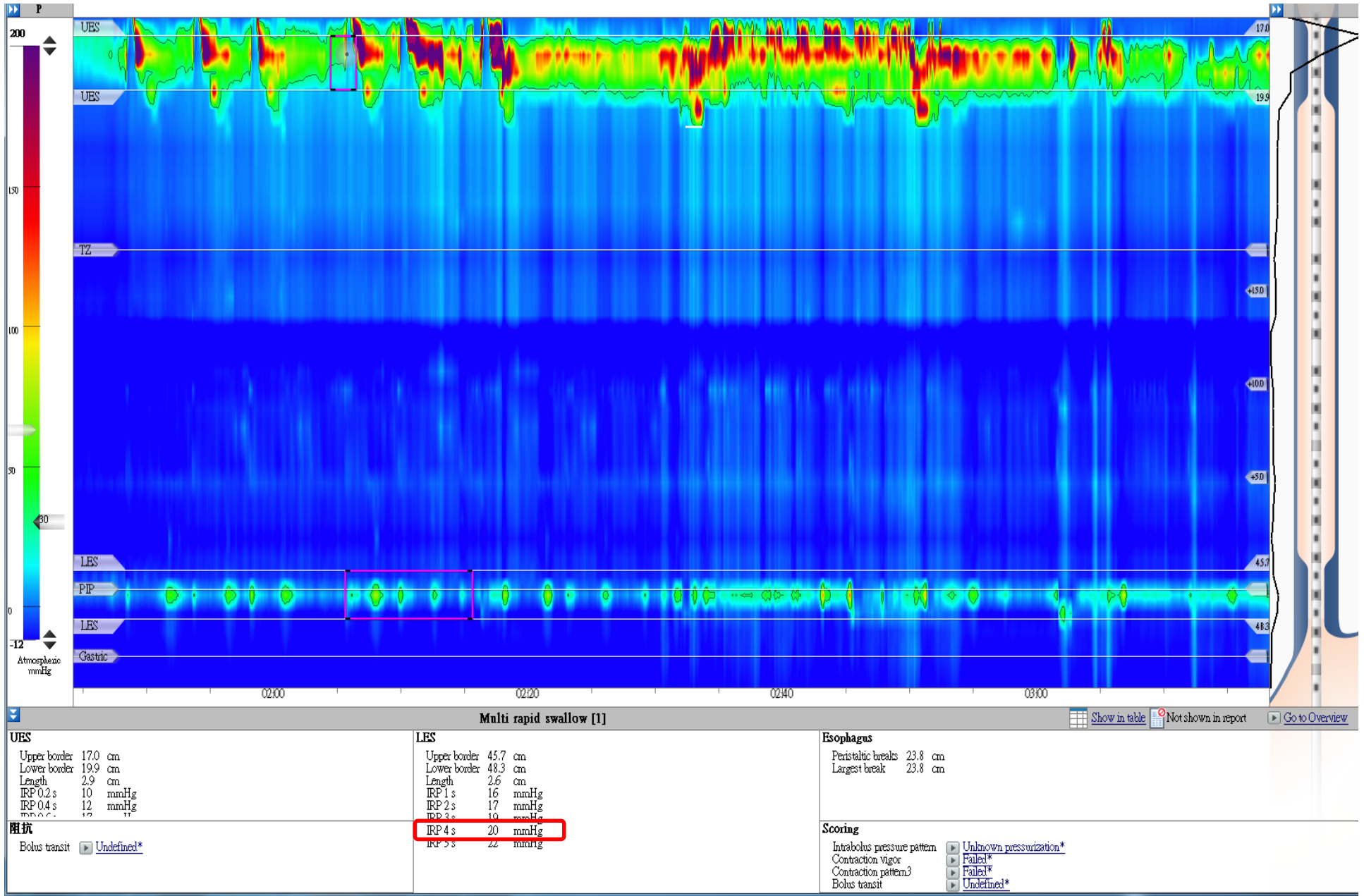
HRIM on 110.08.09



HRIM on 110.08.09



HRIM on 110.08.09



110.08.09 HRIM Report

申請序號： B6160190　　檢查項目： High Resolution Esophageal Manometry

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檢查項目： High Resolution Esophageal Manometry

判讀醫師： 連漢仲 內專３７９８ 消專６２９ 內鏡專（內）８８５８９８

HEIGHT: 166 WEIGHT: 53	VALUE	NORMAL RANGE
Resting pressure of UES	83	33-180mmHg
Location of UES	17	
Resting pressure of LES	59	10-45mmHG
Location of LES	46	
Length of LES	2.9	2.4-5.5cm
Incomplete relaxation of LES	N/A	~90% or residual pressure>5
Amplitude	N/A	13cm: 70(+/-) 32mmHg
	N/A	8cm: 90(+/-) 41mmHg
	N/A	3cm:109(+/-) 45mmHg
Simultaneous peristalsis	0	<10%
Non-transmitted peristalsis	100%	<20%
Ineffective esophageal motility (IEM)	100%	>=50%
IRP	18.86	<21mmHg
DCI	N/A	450-8000mmHg.s.cm
DL	N/A	>4.5s

MRS: 100% Failed,aperistalsis,DCI:N/A

RDC: Failed,aperistalsis,IRP:20/18.86=1.06 (Normal<1)

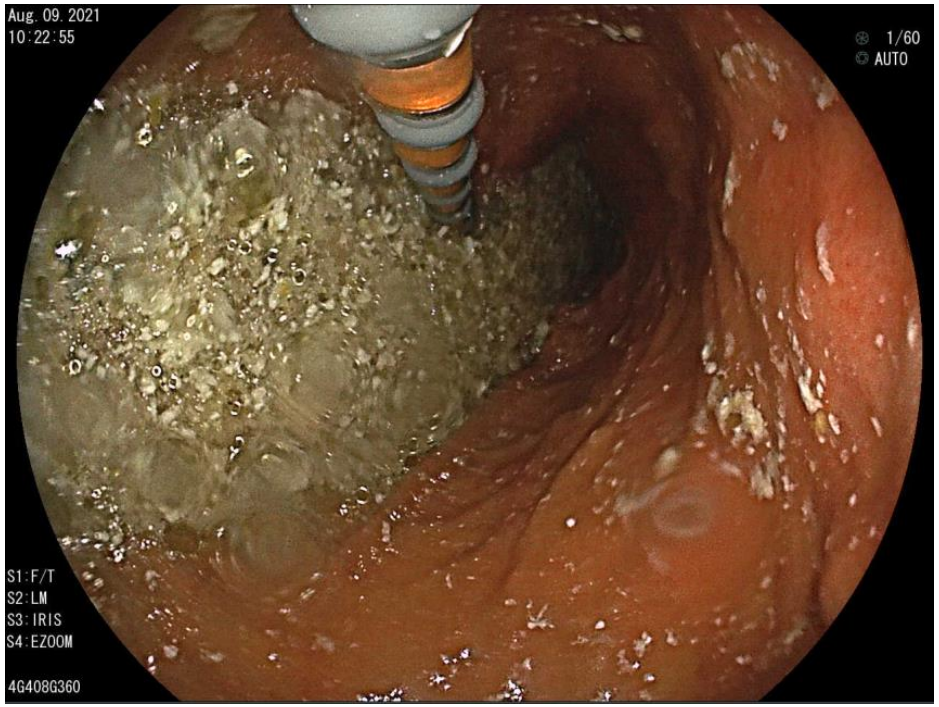
Upright WS 5 times:Aperistalsis,100% Failed contraction.100% incomplete bolus clearance,but IRP:18.93 >15 Cutoff. (by MMS HRIM/C.Cv4.0)
Given elevated IRP in upright position and abnormal RDC, normal-IRP achalasia is most likely based on Cv3.0.
Limitations: The test tube was inserted into the stomach with the gastroscope-assistance and tube was calibrated in vivo, which may account for the possible errors of the IRP.

IMPRESSION: C/W Achalasia, Type I

Impression
c/w Type 1 Achcalasia

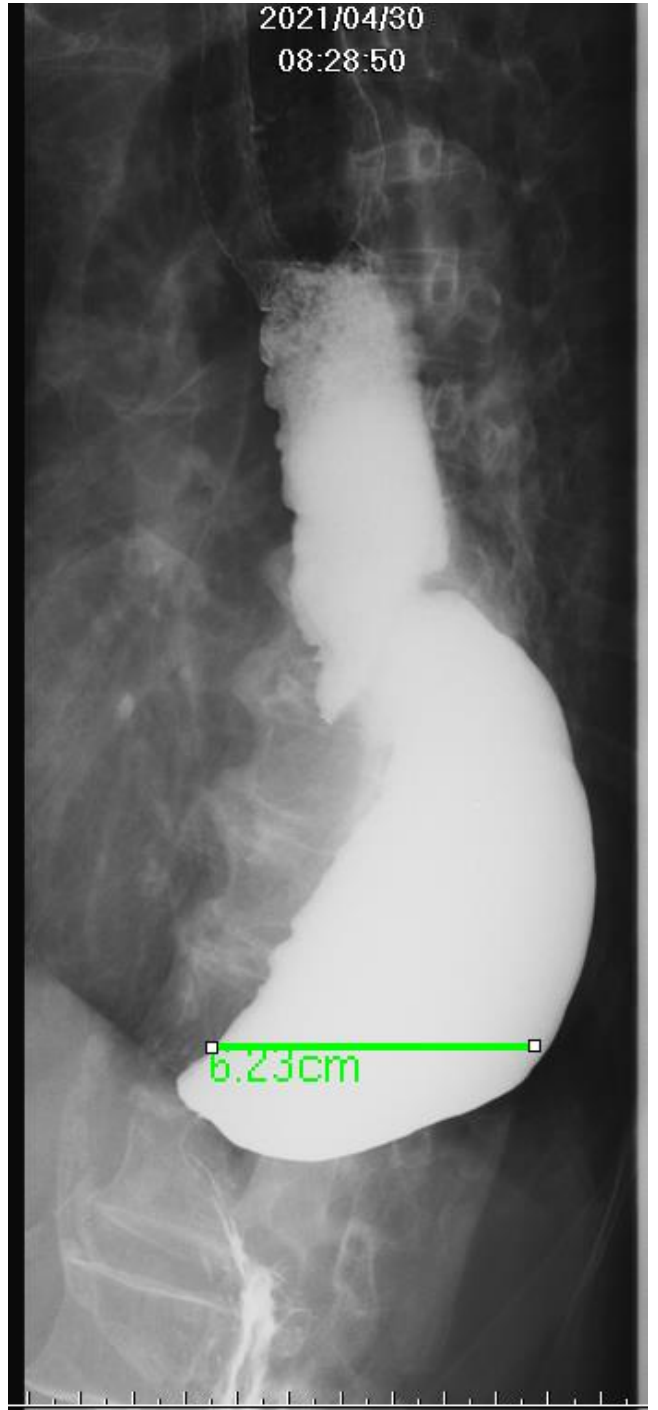
110.08.09
EsophagoGastroDuodenoscopy

ENDOSCOPY FINDINGS:
Esophagus: dilatation w/ food; s/p unisensor
EC junction: mucosa break < 5 mm
Fundus: normal appearance
Body: s/p unisensor
Angularis: normal appearance
Antrum: hyperemia, s/p H.p plate test
Pylorus: normal appearance
Duodenum: normal appearance
DIAGNOSIS/IMPRESSION:
1. Esophagus dilatation w/ food
2. Antral gastritis, s/p H.p plate test
3. s/p unisensor
ADDITIONAL PROCEDURE: biopsy for H.p plate test
COMPLICATIONS: There were no complications associated with
the procedure.
RECOMMENDATION: Follow up
CLINICAL DIAGNOSIS/SYMPTOMS: achalasia
ENDOSCOPE CODE NO: FG8
PREMEDICATION: Xylocain spray;gascon



Diagnosis

- Achalasia, type1, Eckardt score 2, Aperistalsis,100% Failed contraction.100% incomplete bolus clearance



Surgical intervention on 110.10.29

- Thoracoscopic subtotal esophagectomy, Laparoscopic reconstruction with gastric tube via substernal route and Jejunostomy on 2021/10/29.

臺中榮民總醫院

出院病歷摘要

(9)入院日期	110/10/27 (2021) 胸腔外科 W55 - 082
(10)轉科日期	
(11)出院日期	110/11/05 (2021) 住院天數計 9 日

診斷

入院：1. Achalasia, type1, Eckardt score 2, Aperistalsis,100% Failed contraction.100% incomplete bolus clearance, IRP:18.93 >15 Cutoff
- s/p myotomy in 1985
- s/p pneumatic dilatation.

- 2. Hypertension under bisoprolol and amlodipine.

出院：1. Achalasia, type1, Eckardt score 2, Aperistalsis,100% Failed contraction.100% incomplete bolus clearance, IRP:18.93 >15 Cutoff
- s/p myotomy in 1985
- s/p pneumatic dilatation
- s/p Thoracoscopic subtotal esophagectomy, Laparoscopic reconstruction with gastric tube via substernal route and Jejunostomy on 2021/10/29.

- 2. Hypertension under bisoprolol and amlodipine.

主訴

- Intermittnet dysphagia to solid meal for 1-2 years without weight loss.

病史

This is a 72 year old female with
1. Achalasia, type1, Eckardt score 2, Aperistalsis,100% Failed contraction.100% incomplete bolus clearance, IRP:18.93 >15 Cutoff
s/p myotomy in 1985
s/p balloon dilatation
2. Hypertension under bisoprolol and amlodipine
This year, she mentioned symptoms of intermittnet dysphagia to solid meal for 1-2 years without weight loss. Esophagography on 2021/04/30 showed marked distal esophageal dilatation with barium stasis. HRM (high resolution esophageal manometry) on 2021/08/03 showed Aperistalsis,100% Failed contraction.100% incomplete bolus clearance,but IRP:18.93 >15 Cutoff, c/w achalasia type 1. Duodenoscopy showed Esophagus dilatation w/ food, Antral gastritis, s/p H.p plate test (+).
Surgical intervention was suggested. The patient was refered to CS for evaluation. Treatment choice include balloon dilatation, endoscopic myotomy, esophagectomy were explained. The patient decided to receive esophagectomy. This time, due to the diagnosis of Achalasia, type 1 , Eckardt score 2, she was admitted for VATS

- esophagectomy, laparoscopic reconstruction with gastric tubing.

體檢發現

Gastroscope:Esophagus dilatation with food
Antral gastritis with Hp infection

COUGH RECENTLY
RHINORRHEA
UE(910924): esophageal dilatation, gastritis
1010315 UGI: esophageal dilation; gastritis

- Performance status(ECOG)： 1(Symptoms but ambulatory)
- General appearance :
  - easy looking , conscious clear , without cardiopulmonary distress
  - body weight : 64 kg
  - height : 164.4 cm
  - Vital signs date 體溫(BT) 脈搏(P) 呼吸(R) 血壓(BP) 身高(BH) 體重(BW) BMI
  - 20211027 36.9℃ 90次/分 18次/分 179/105mmHg 164.4cm 54.2kg 20.1
  - Vital signs: B.P. 179/105 mmHg , P.R. 90 /min , R.R. 18 /min , B.T. 36.9 ℃
- HEENT :
  - Head: grossly normal, no deformity
  - Eyes: no anemic conjunctiva, no icteric sclera, normal EOM
  - ENT: no otorrhea, no rhinorrhea, no erythematous mucosa, no injected tonsil, no deformity
- Neck: supple, freely movable, trachea midline,no deformity, no engorged jugular vein , no palpable thyroid or lymph node enlargement

手術日期及方法

手術日期及方法20211029 1.Thoracoscopic subtotal esophagectomy.
2.Laparoscopic reconstruction with gastric tube via substernal route.
3.Jejunostomy.
4.Indocyanine Green Fluorescence Real-Time Overlay Imaging.

住院治療經過

After admission, thoracoscopic subtotal esophagectomy, laparoscopic reconstruction with gastric tube via substernal route and jejunostomy were smoothly done on 2021/10/29. She was transferred to SICU for post-op care and jejunostomy feeding with D5W was tried on 10/30. Then, fair digestion from jejunostomy feeding was found. However, low-grade fever was noted and flomoxef was prescribed as empirical used. No discomfort was noted after feeding. Chest tube and neck Penrose drainage was removed on 11/03. CXR after that showed mild left pneumothorax. We rechecked CXR next day and no progression was noted. Under the stable condition, the patient

- was discharged on 11/05.

診斷醫師:文美卿 報告日期:110/11/05
Pathologic diagnosis:
1. Esophagus and stomach, esophagectomy and gastric tube reconstruction ---- Achalasia with lymphocytic esophagitis, multifocal ulcers and esophageal dilation.
2. Lymph node, group 3p, dissection ---- Unremarkable fibroadipose tissue. No lymph node seen.
Ancillary study for diagnosis:
IHC stain for PHOX2B done on section H, no ganglion cell is detected.
Gross description:
The specimen consists of 1) the distal part of esophagus, 9.5x6x1.8 cm, the cardia, 5.4x5x1.4 cm. There is area of irregular mucosa, significant lumen dilation with wall thinning over the distal part of esophagus, with the distance of 0.5 cm to the gastroesophageal junction. On serial section, no mass lesion is identified grossly. 2) "LN3p", a piece of tan brown soft tissue, 0.3x0.2x0.1 cm.
Representative sections taken: A) proximal cut end B) distal cut end C) cardioesophageal junction D-H) esophagus lesion J) para-esophageal LN K) peri-gastric LN L) specimen 2.
Microscopic description:
The esophagus show diffuse, almost complete loss of myenteric ganglion cells. Few ganglion cell preserved in the proximal cut end. Chronic inflammation, admixed eosinophils, few plasma cells, mast cells, around myenteric (Auerbach) plexus nerve fascicles are observed. There are also hypertrophy, eosinophilia of inner circular layer; the mucosa show squamous and basal cell hyperplasia, intraepithelial lymphocytosis; lymphocytic infiltrate, especially around ductal glands Prominent follicles and even germinal centers in the submucosa.
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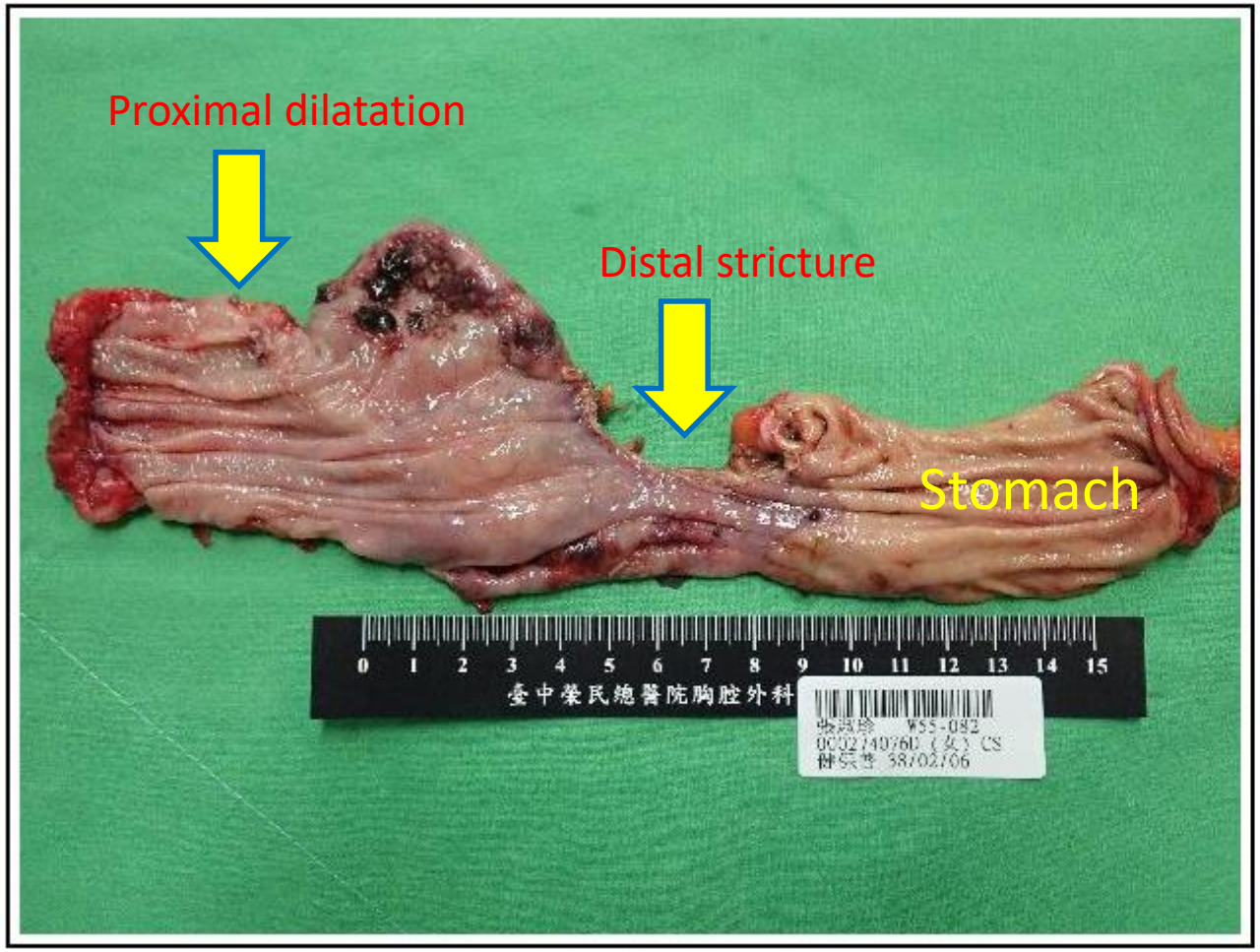
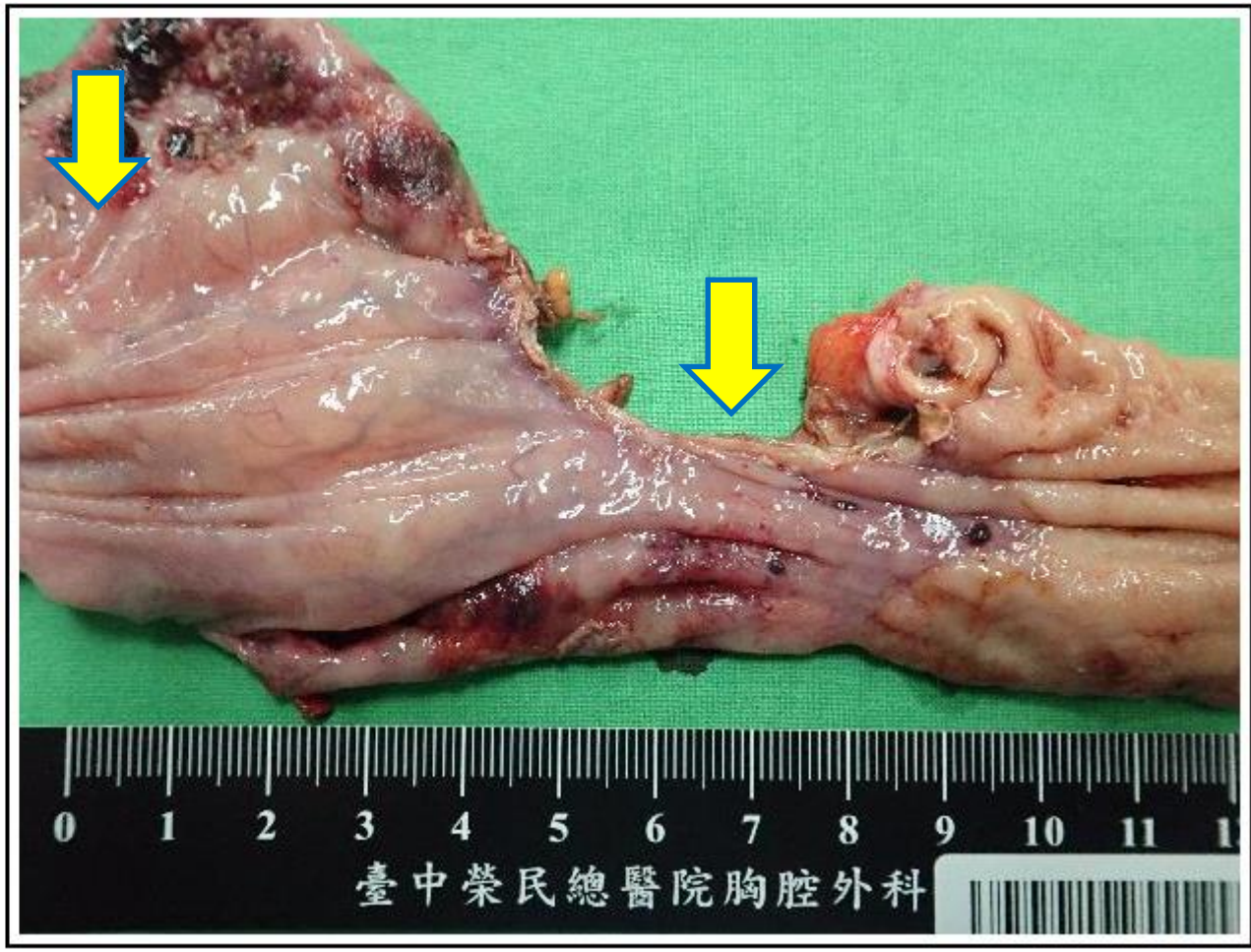
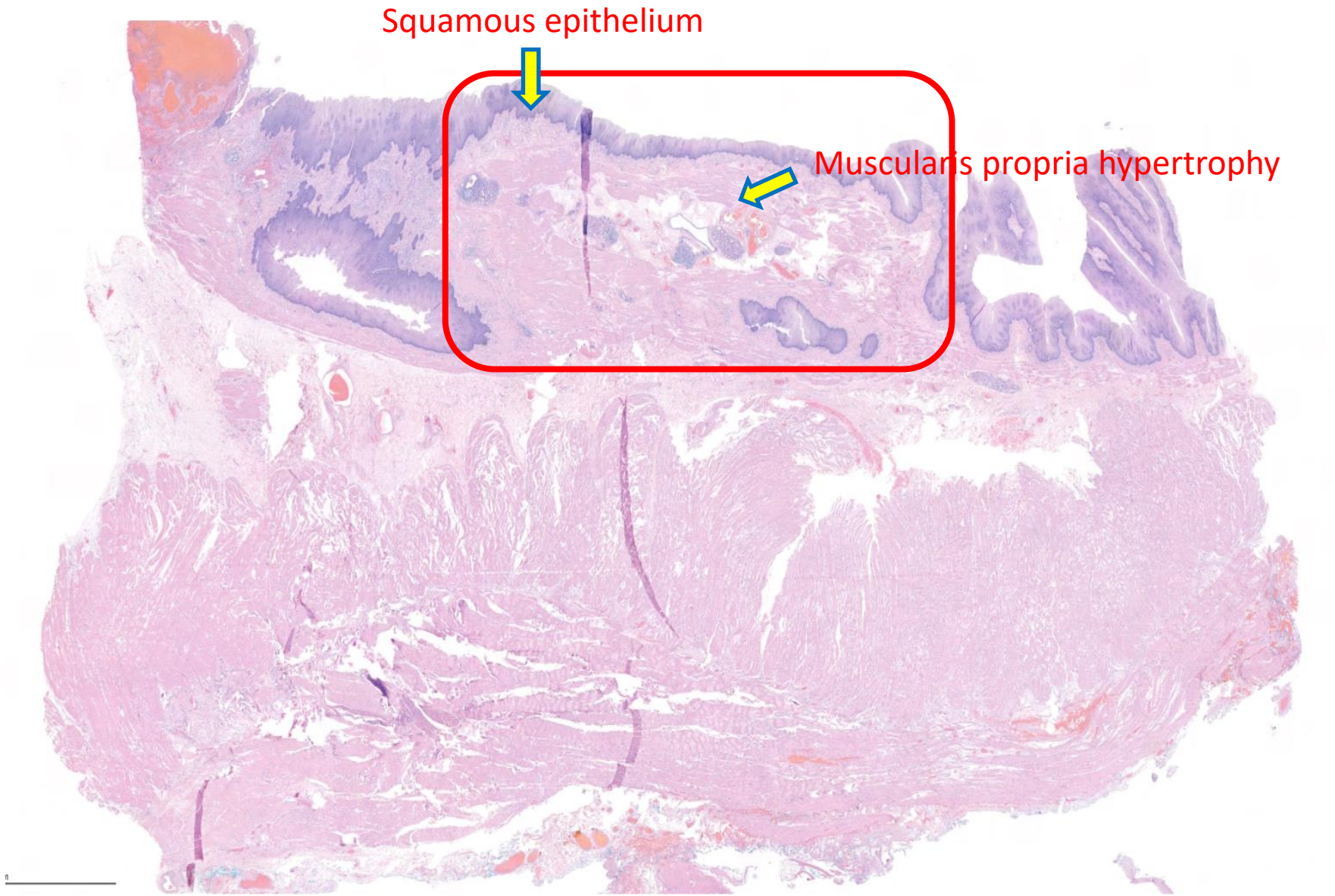
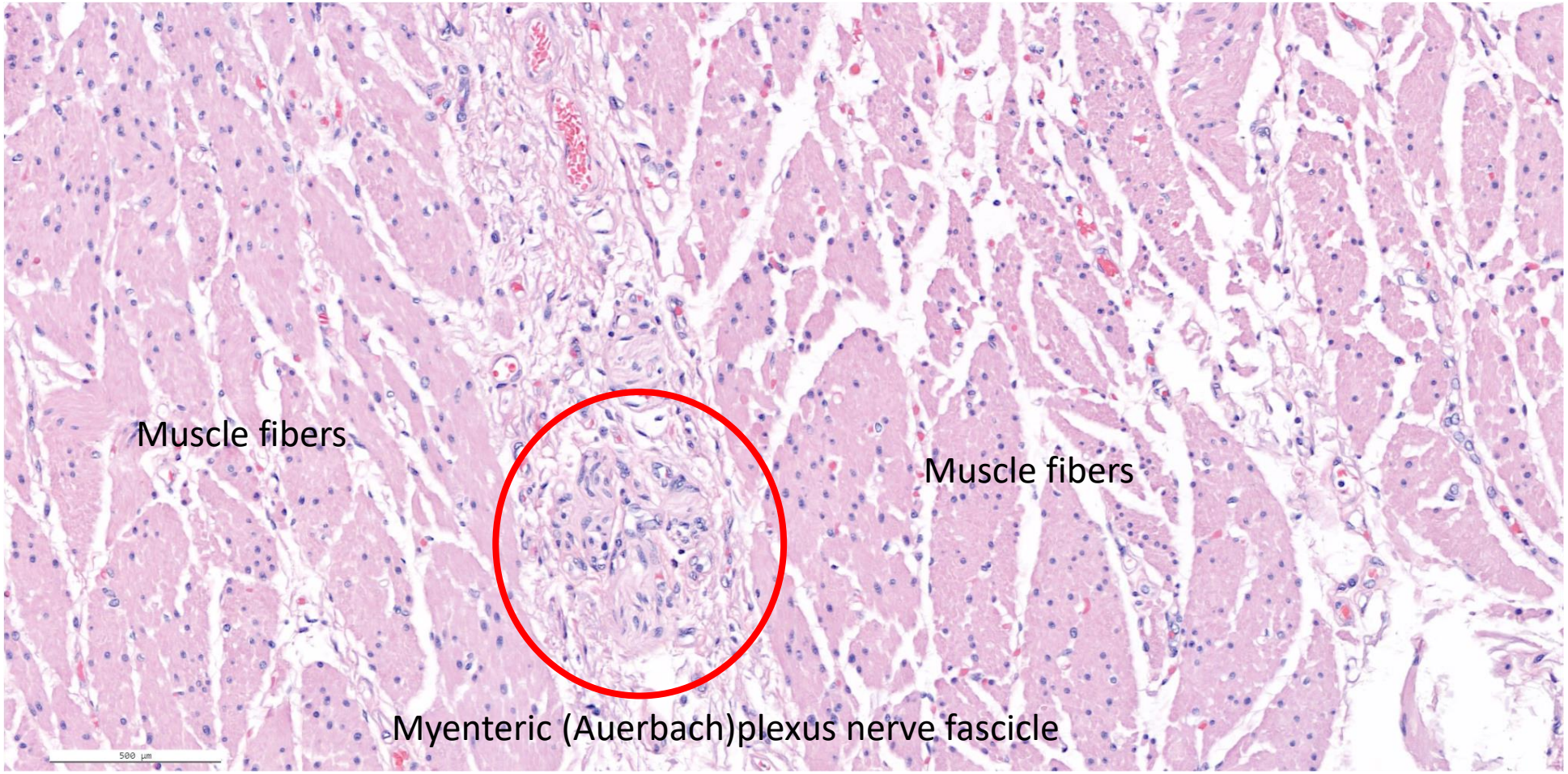
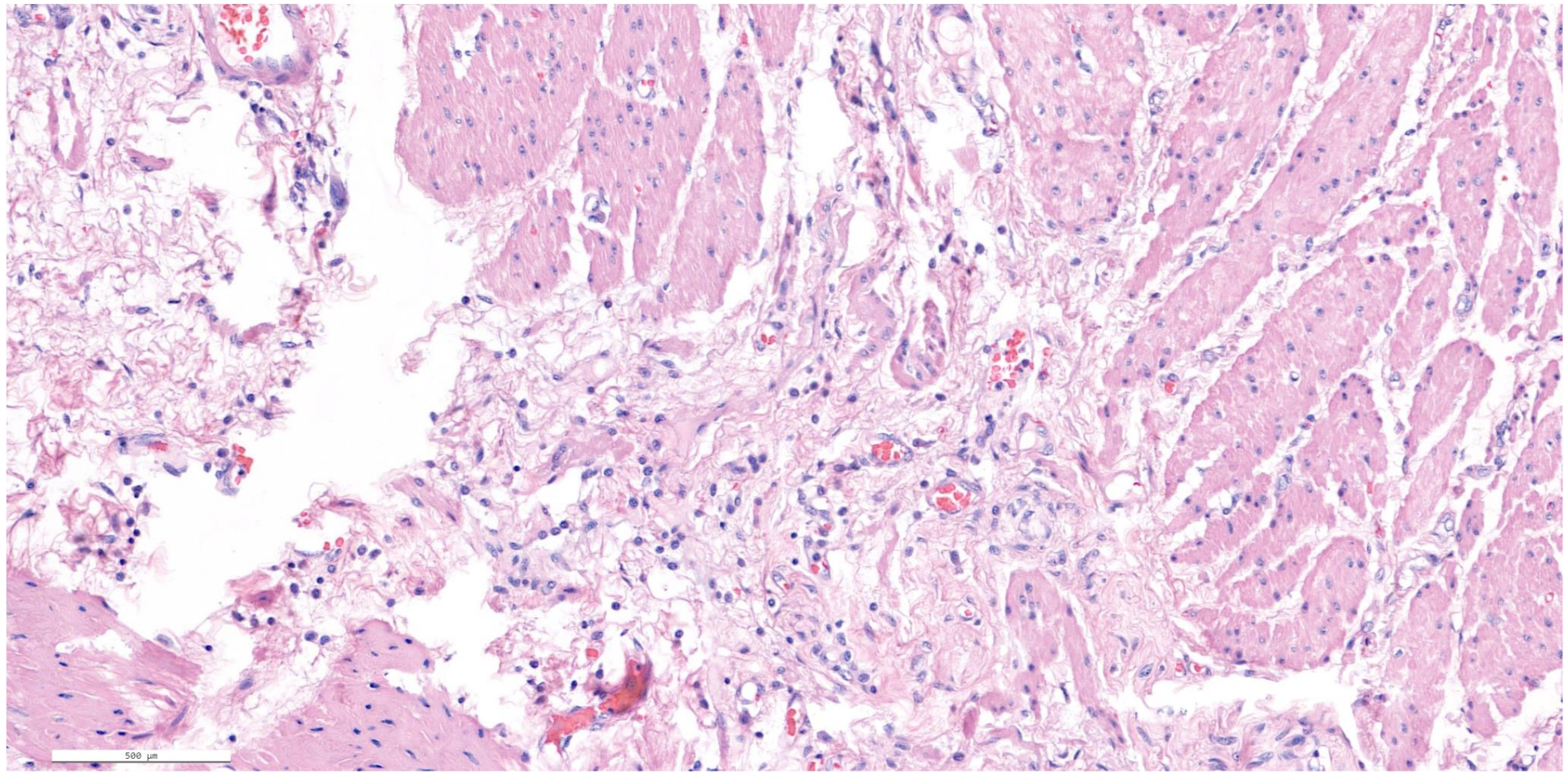

Proximal dilatation
Distal stricture
Stomach
0 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15
臺中榮民總醫院胸腔外科

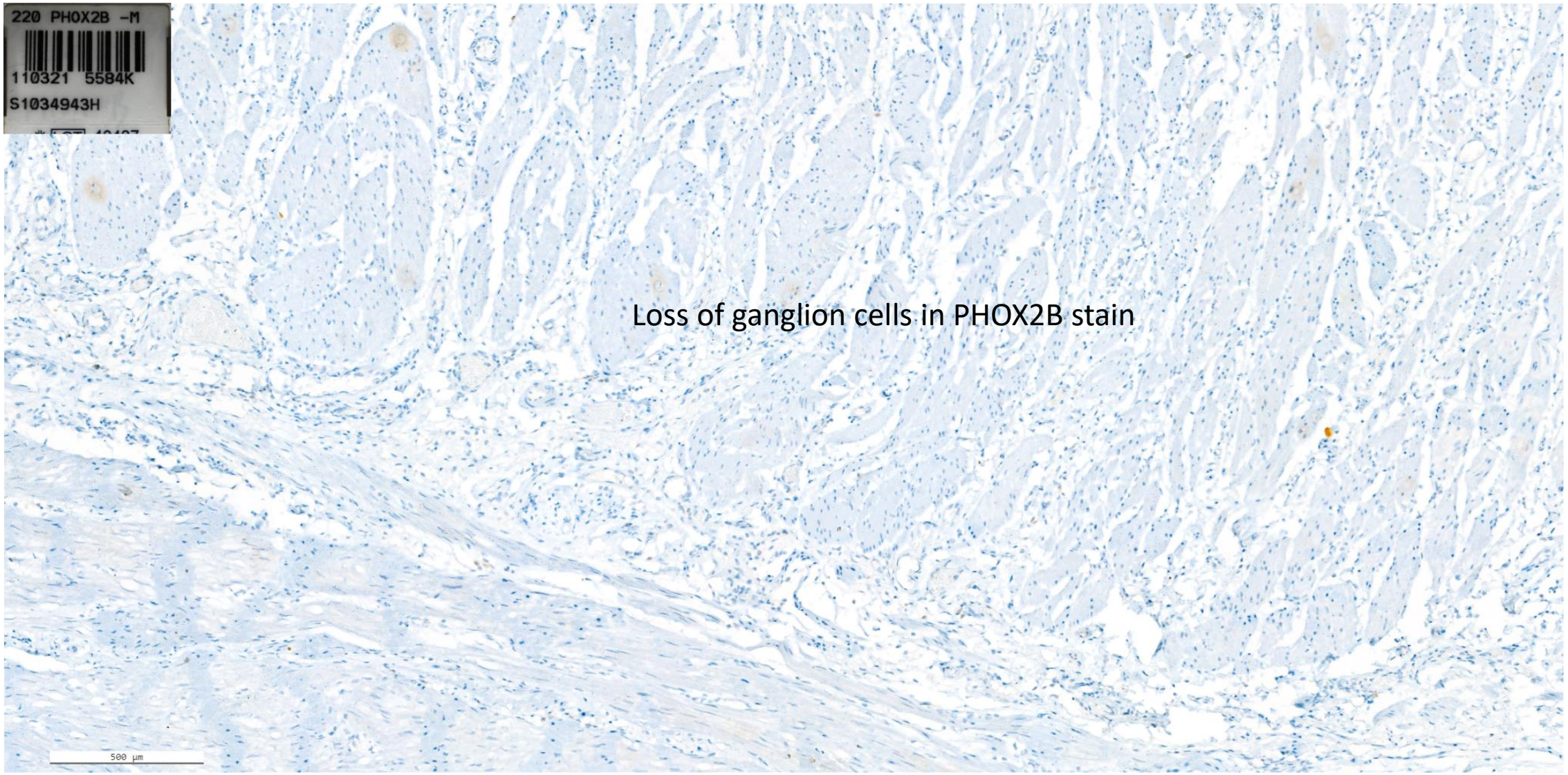

0 1 2 3 4 5 6 7 8 9 10 11
臺中榮民總醫院胸腔外科


Squamous epithelium
Muscularis propria hypertrophy


Muscle fibers
Muscle fibers
Myenteric (Auerbach)plexus nerve fascicle
500 µm


500 µm

PHOX2B (染ganglion cell)



Discussion

Achalasia Natural history and prognosis

Disease course — Without treatment, patients with achalasia can develop progressive dilation of the esophagus. Late- or end-stage achalasia is characterized by esophageal tortuosity, angulation, and severe dilation or megaesophagus (diameter >6 cm). Approximately 10 to 15 percent of patients who have undergone treatment for achalasia will develop late- or end-stage achalasia, and up to 5 percent of patients in some series require esophagectomy [54,55].

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Review

Oesophagectomy in the management of end-stage achalasia – Case reports and a review of the literature

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Department of Surgery, St. James's Hospital, Dublin 8, Ireland

Received 24 July 2010, Revised 4 November 2010, Accepted 17 November 2010, Available online 25 November 2010.

Table 2. Criteria for considering oesophagectomy in patients with end-stage achalasia.

End stage achalasia	**Failed myotomy/re-do myotomy as evidenced by:**
Clinical	Eckhardt[10] grading score of 6–12 (“Poor Response”)
	Severe dysphagia or regurgitation
	Nutritional failure
	Poor quality of life
Radiological & manometric	Massively dilated oesophagus (>6cm)
	Highly tortuous (sigmoid) oesophagus
	Oesophageal aperistalsis

End stage Achalasia treatment

- Pneumatic dilatation
- Surgical myotomy
- Re-do myotomy or oesophagectomy in highly tortuous and dilated sigmoid oesophagus

Esophagectomy

- Relief of dysphagia
- Excellent symptom control
- Improved quality of life and satisfaction with their outcome
- 75–100% : good symptom control

Esophagectomy complication

- Mortality rate: 5-10%
- Morbidity rate: 50%
- Longer-term complications : Anastomotic stricture, Dumping syndrome

Back to our patient

- P: 74 y/o , Female, Achalasia s/p Myotomy on 1985, Recurrence achalasia , End stage Achalasia
- Myotomy, Pneumatic dilatation or Esophagectomy
- Surgical intervention with Esophagectomy : Thoracoscopic subtotal esophagectomy, Laparoscopic reconstruction with gastric tube via substernal route

題目		2023/01/20
A：生活型態		
1	您平常每天吃幾次正餐	更多次
2	您平常每天吃幾次點心或零食(不含水果	3次
3	您每週三餐外食(或在外用餐)幾次	
3-1	早餐	0次
3-2	午餐	0次
3-3	晚餐	3次
4	您外食最常去的三個地方	小吃店
5	您每天下班或下課後看電視或靜態休閒	2~3.9小時
6	靜態休閒時經常吃點心或零食嗎(不含水	是
7	過去一週，您有幾次在晚上九點後進食	5次或以上
7-1	晚上九點後，都吃些什麼食物	飯麵類
8	您平常每週運動幾個小時	0~0.5小時
8-1	通常都做什麼運動	
9	抽菸習慣	沒有抽菸
C：其他飲食習慣		
1	每週吃幾份蛋料理	4～6.9份
3	每週吃幾份乳製品	0～1.9份
3-1	乳製品種類	
5	服用(魚油、燕麥、甲殼素、紅麴食品)?	都沒有
6	您近一個月內的其他飲食習慣	
6-1	每天喝幾多少水	<1000cc
18-2	每天喝幾杯牛奶	不到1杯
18-2-1	牛奶種類	全脂
18-3	每天喝幾杯咖啡	
18-3-1	咖啡種類	黑咖啡
18-4	每週喝幾杯果汁或蔬果汁	
18-4-1	果汁取得方式	
18-4-2	果汁種類	
18-4-3	果汁甜度	
18-5	每天喝幾杯豆漿	
18-5-1	豆漿種類	
18-6	每天喝幾杯紅酒	
18-7	每天喝幾杯啤酒	
18-8	每天喝幾杯烈酒	
18-9	每天喝幾杯杏仁奶	
18-10	每天喝幾杯椰奶	
18-11	每天喝幾杯運動飲料	
18-12	每天喝幾杯茶	
18-12-1	茶甜度	
18-13	每天喝幾杯奶茶	
18-13-1	奶茶甜度	
18-14	每天喝幾杯碳酸飲料	
18-14-1	碳酸飲料甜度	
18-15	其他飲食習慣	

B：請說明您的飲食習慣（平均每天或每週）			國際	台灣
0	較常吃紅肉或白肉	紅肉	(白肉,吃素) 0	
1	每天吃幾份紅肉或加工肉	1~1.9份	(＜2) 1	
1-1	每週加工肉吃幾份	0~1.9份		
1-2	每週控肉、五花肉、滷肉、三層肉吃幾	0~1.9份		
1-3	每週吃幾份紅肉或加工肉			
2	每天吃幾份家禽肉	0~1.9份		
2-1	每週吃幾份家禽肉	0.1~1.9份		
3	每週吃幾份魚或貝類	2~4.9份	(≧5) 0	
3-1	每週吃幾份海鮮(九孔螺、干貝、章魚	0~1.9份		
3-2	每週吃幾份魚或貝類			
4	每週吃幾份市售烘焙食品或零食	0~2.9份	(＜3) 1	
5	每天吃幾份精緻澱粉	0~3.9份	(＜4) 1	
6	每天吃幾份水果	0~1.9份	(≧4) 0	(≧2) 0
6-1	您近一個月常吃的水果	芭樂、香蕉、?		
7	每天吃幾份蔬菜(國際的5份中有2份是	0~2.9份	(≧5) 0	(≧3) 0
7-1	每天吃生菜(含蔬菜汁)的習慣			
8	每天吃幾份全穀類	0~2.9份	(≧3) 0	
9	每天吃幾湯匙的橄欖油(國際：橄欖油	0~3.9匙	(≧4) 0	
10	每週吃幾份豆莢類	0~7.9份	(≧8) 0	
11	每週吃幾份堅果	0~2.9份	(≧3) 0	
12	橄欖油為您的主要烹調用油嗎(國際：?	是	(是) 0	(是) 0
12-1	您都使用什麼油	葵花油		
13	每天喝幾杯汽水或含糖飲料	每天少於100c.c	(＜1) 1	
14	每天吃幾份奶油	0~0.9份	(＜1) 1	
	地中海飲食評分*		5	5

Eckardt score

110/10/29
1. Thoracoscopic subtotal esophagectomy.
2. Laparoscopic reconstruction with gastric tube via substernal route.
3. Jejunostomy.

		110/8/3	110/8/9	110/8/31	110/12/20	111/3/11	111/11/22	112/1/17	112/4/14
1	體重減輕	1	0	0	1	0	0	0	0
2	吞嚥困難	1	1	1	1	0	0	0	0
3	胸口疼痛	0	0	0	0	0	0	0	0
4	逆流	1	0	1	1	0	0	0	0
	Total	3	1	2	3	0	0	0	0

The Reflux Symptom Index (RSI)

110/10/29
1. Thoracoscopic subtotal esophagectomy.
2. Laparoscopic reconstruction with gastric tube via substernal route.
3. Jejunostomy.

The Reflux Symptom Index (RSI)		110/8/3	110/8/31	111/3/11	111/11/22	112/1/17	112/4/14
1	你有沙啞或聲音的問題	0	0	0	0	0	0
2	清喉嚨	0	0	1	0	0	0
3	過多喉嚨黏液或鼻涕倒流	0	0	1	0	0	0
4	吞嚥食物，液體或藥丸困難	1	0	0	0	0	1
5	進食或躺下後咳嗽	0	0	0	0	0	0
6	呼吸困難或嗆到事件	0	0	1	0	0	0
7	令人討厭或惱人的咳嗽	0	0	0	0	0	0
8	有東西黏在你喉嚨或有塊狀物在你喉嚨的感覺	1	1	0	0	0	0
9	心灼熱，胸痛，消化不良或胃酸跑上來	1	1	0	0	0	1
	Total	3	2	3	0	0	2

Score range: 0-45 (normal ≤13),
the higher the score, the more severe the symptom.

Belafsky PC, 2002 J Voice.
Lien HC, 2015 Value Health

Reflux Disease Questionnaire (RDQ)

110/10/29
1. Thoracoscopic subtotal esophagectomy.
2. Laparoscopic reconstruction with gastric tube via substernal route.
3. Jejunostomy.

回想過去一個月，您認為以下症狀出現時的如何？		110/8/3		110/8/31		111/3/11		111/11/22		112/1/17		112/4/14	
程度：0-不會，5-重度 頻率：0-不會，5-每天		程度	頻率	程度	頻率	程度	頻率	程度	頻率	程度	頻率	程度	頻率
1	胸骨後方感到灼熱	0	0	0	0	0	0	0	0	0	0	0	0
2	胸骨後方感到疼痛	0	0	0	0	0	0	1	0	0	0	0	0
3	上腹中間感到灼熱	0	0	0	0	0	0	0	0	0	0	1	0
4	上腹中間感到疼痛	0	0	0	0	0	0	0	0	0	0	1	0
5	口腔內有酸味	0	0	1	1	1	1	0	0	0	0	0	0
6	有東西從胃部向上移動而感到不適	0	0	1	0	0	0	0	0	0	0	0	0
Total		0		3		2		1		0		2	

分數範圍: 0-40分；正常值<12

Shaw MJ, 2001 Am J Gastroenterol CHINESE GERD STUDY GROUP, 2004 Chin J Dig Dis

Back to our patient

- P: 74 y/o , Female, Achalasia s/p Myotomy on 1985, Recurrence achalasia , End stage Achalasia
- Myotomy, Pneumatic dilatation or Esophagectomy
- Surgical intervention with Esophagectomy : Thoracoscopic subtotal esophagectomy, Laparoscopic reconstruction with gastric tube via substernal route
- Outcome : symptom relief but Dumping syndrome with poor quality of life
- Result : Consider to Re-do myotomy at future for the same patient

~~Thanks for your Attention~~