FIEBRE RECURRENTE
Y
SÍNDROME GENERAL
DE MESES DE EVOLUCIÓN

Sesión Conjunta Radiología y Medicina Interna

18 de Diciembre de 2019

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Resumen del Ingreso:

Paciente con antecedente de proceso psiquiátrico y cuadro de varios meses de evolución consistente en episodios febriles repetidos, la mayoría no constatados, de muy breve duración, sin clara repercusión funcional, sin alteración analítica puntual ni a lo largo del tiempo, sin evidencia radiológica de enfermedad (Tórax, Senos nasales, Abdomen) y acompañada de intensa pérdida de peso

Dos ingresos previos, numerosas consultas hospitalarias y múltiples visitas a Urgencias sin diagnóstico concreto de enfermedad orgánica.

Y ahora que?

- TAC Tóraco-Abdominal: Patrón en vidrio deslustrado
- Ese mismo día fiebre de 38,5ºC, sin clínica, se extraen hemocultivos, analítica sin cambios, salvo discreto aumento de la PCR
- Azitromicina
- Afebril

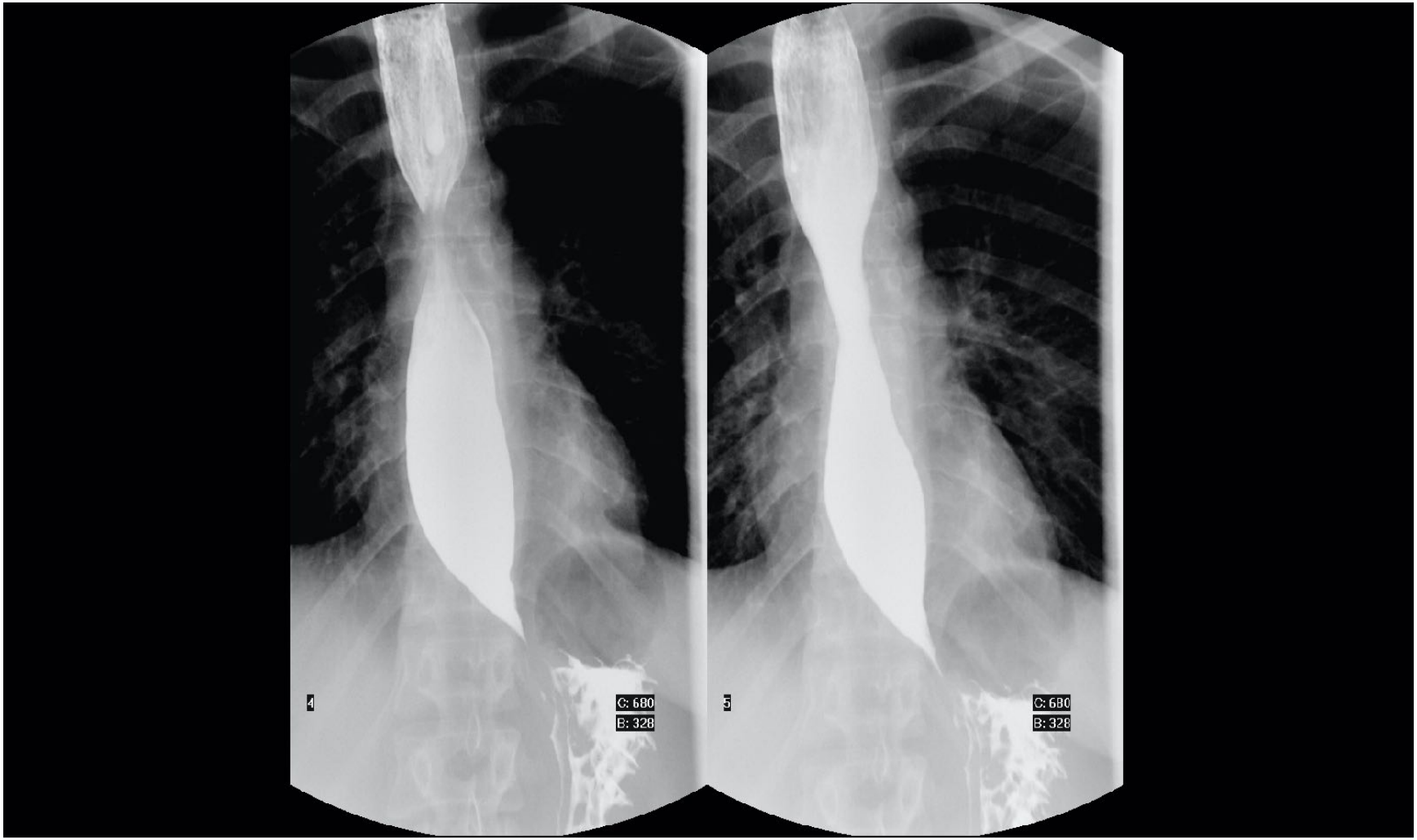
Gastroscopia:

Esofagitis péptica Vs infecciosa¿?

Resistencia al paso del endoscopio en cardias.

Gastritis crónica antral

Fiebre recurrente y síndrome general de meses de evolución



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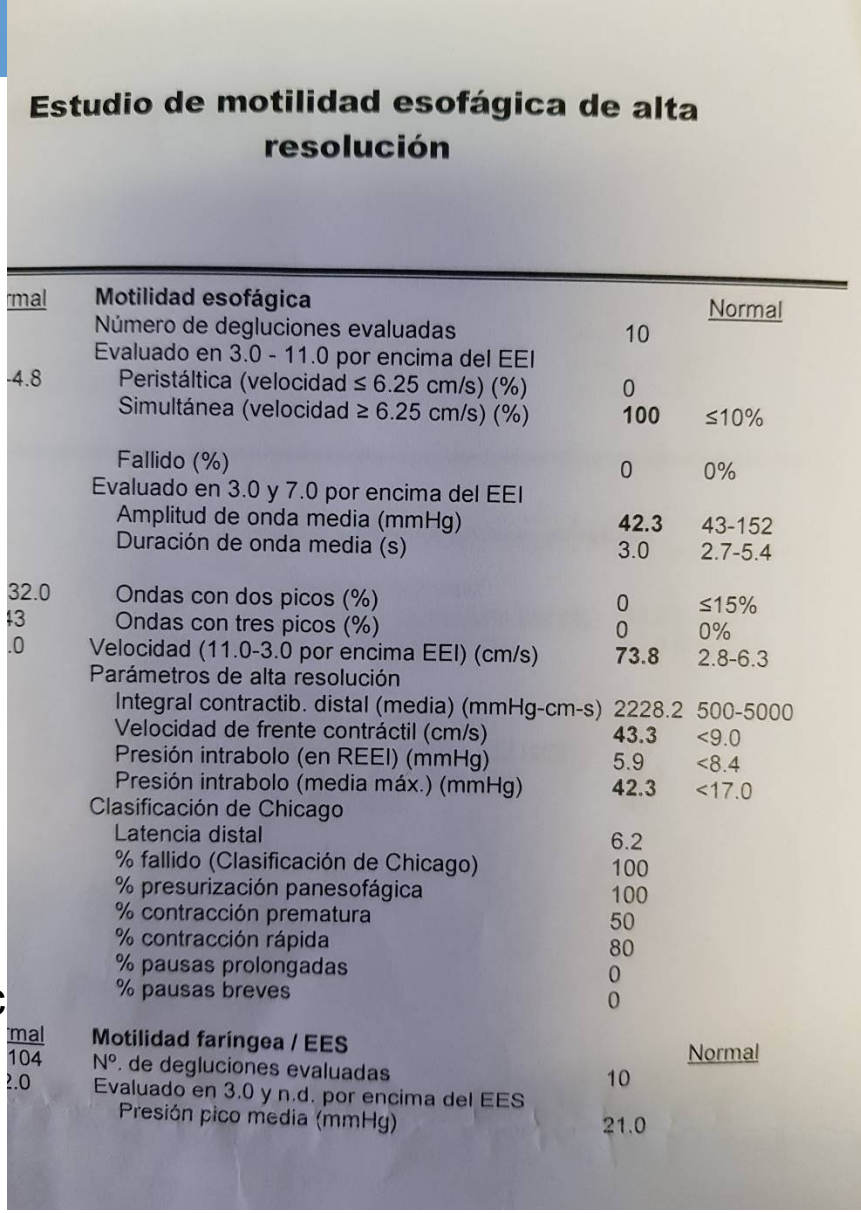
Manometría de Alta Resolución:
Unión esofagogástrica de 3,7 cm

con **presión de reposo elevada** 41 mm Hg

y **presión residual también elevada**
que traduce falta de relajación,

en cuerpo **100% simultaneas siendo**
todas ellas con panpresurización esofágic



Estudio de motilidad esofágica de alta resolución

		Normal
Motilidad esofágica		
Número de degluciones evaluadas	10	
Evaluado en 3.0 - 11.0 por encima del EEI		
Peristáltica (velocidad ≤ 6.25 cm/s) (%)	0	
Simultánea (velocidad ≥ 6.25 cm/s) (%)	**100**	≤10%
Fallido (%)	0	0%
Evaluado en 3.0 y 7.0 por encima del EEI		
Amplitud de onda media (mmHg)	**42.3**	43-152
Duración de onda media (s)	3.0	2.7-5.4
Ondas con dos picos (%)	0	≤15%
Ondas con tres picos (%)	0	0%
Velocidad (11.0-3.0 por encima EEI) (cm/s)	**73.8**	2.8-6.3
Parámetros de alta resolución		
Integral contractib. distal (media) (mmHg-cm-s)	2228.2	500-5000
Velocidad de frente contráctil (cm/s)	**43.3**	<9.0
Presión intrabolo (en REEI) (mmHg)	5.9	<8.4
Presión intrabolo (media máx.) (mmHg)	**42.3**	<17.0
Clasificación de Chicago		
Latencia distal	6.2	
% fallido (Clasificación de Chicago)	100	
% presurización panesofágica	100	
% contracción prematura	50	
% contracción rápida	80	
% pausas prolongadas	0	
% pausas breves	0	
Motilidad faríngea / EES		Normal
Nº. de degluciones evaluadas	10	
Evaluado en 3.0 y n.d. por encima del EES		
Presión pico media (mmHg)	21.0	

- Acalasia tipo II

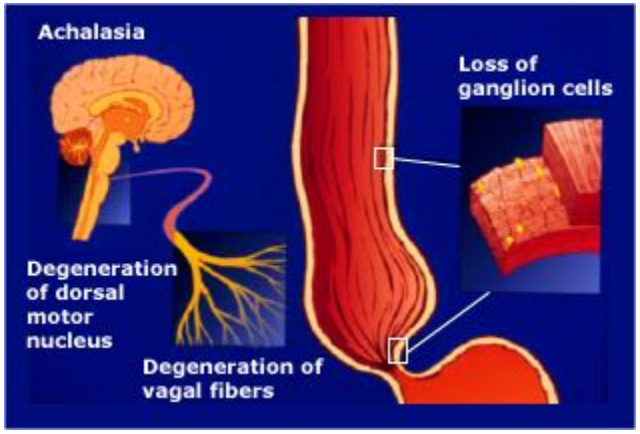
- **incidence** of approximately **1.6** cases per 100,000 individuals and **prevalence** of **10** cases per 100,000 individuals
- Men and women are affected with equal frequency
- can occur at any age, but onset before adolescence is rare. Achalasia is **usually** diagnosed in patients between the ages of **25 and 60** years.
- The etiology of primary or idiopathic achalasia is unknown

Diseases with manometric findings of achalasia

Malignancy, especially gastric carcinoma
Chagas disease
Amyloidosis
Sarcoidosis
Neurofibromatosis
Eosinophilic esophagitis
Multiple endocrine neoplasia, type 2B
Juvenile Sjögren's syndrome with achalasia and gastric hypersecretion
Chronic idiopathic intestinal pseudo-obstruction
Anderson-Fabry disease

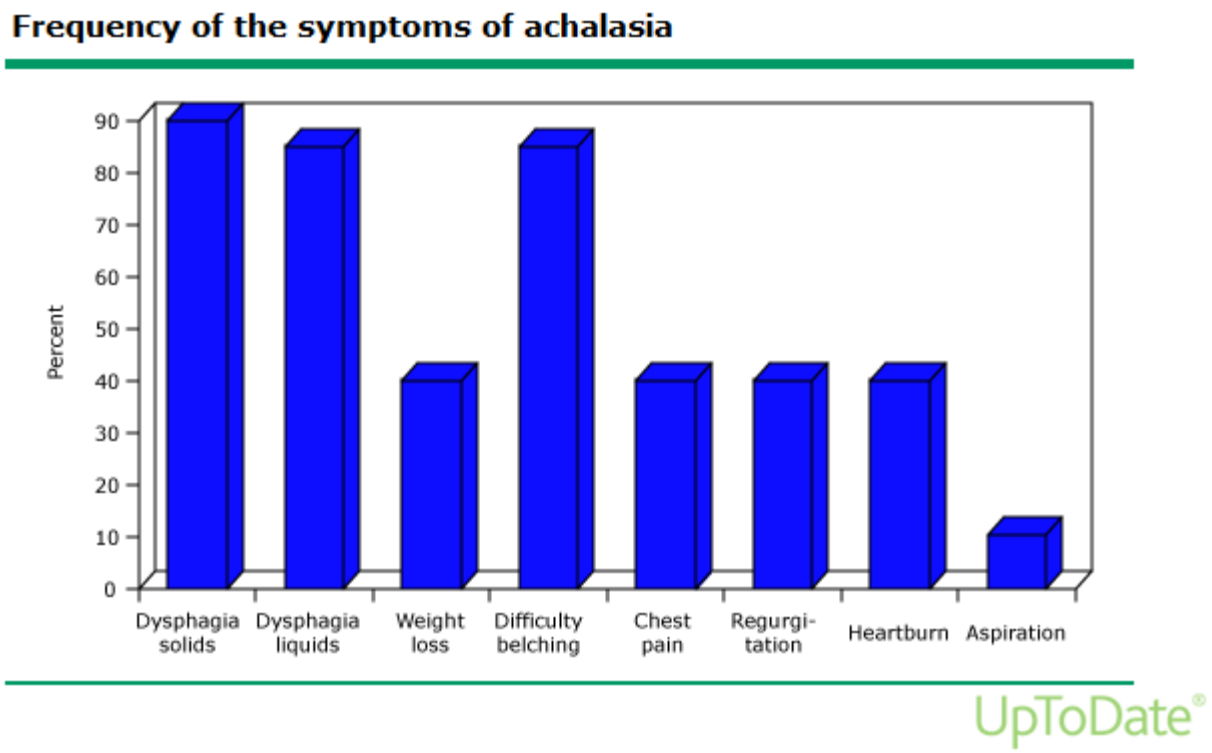
UpToDate®

- Achalasia results from **inflammation and degeneration of neurons in the esophageal wall** The **cause** of the inflammatory degeneration of neurons in primary achalasia is **not known**
- is associated with **variants in the HLA-DQ** region and that affected patients **often have circulating antibodies to enteric neurons** suggest that achalasia is an **autoimmune** disorder (herpes zoster, measles viruses, HSV-1)



- **Histologic: decreased numbers of neurons (ganglion cells) in the myenteric plexuses, and the ganglion cells** that remain often are surrounded by lymphocytes and, less prominently, by eosinophils. This inflammatory degeneration preferentially **involves the nitric oxide-producing, inhibitory neurons that affect the relaxation of esophageal smooth muscle**; the cholinergic neurons that contribute to lower esophageal sphincter (LES) tone by **causing smooth muscle contraction** may be relatively spared

Fiebre recurrente y síndrome general de meses de evolución



- **Diagnostic approach** — Achalasia should be suspected in the following patients:
- **Dysphagia** to solids and liquids

- **Heartburn** unresponsive to a trial of proton pump inhibitor therapy

- **Retained food** in the esophagus on upper endoscopy

- Unusually **increased resistance** to passage of an **endoscope** through the esophagogastric junction (EGJ)

- **Endoscopic** evaluation with upper gastrointestinal endoscopy should be performed in patients with suspected achalasia **to exclude a malignancy** at the EGJ that can mimic achalasia

- **Esophageal manometry** is required to establish the **diagnosis**.

- In patients with **equivocal esophageal manometry** results (eg, incomplete LES relaxation but some preserved peristalsis; some complete LES relaxation with aperistalsis), **barium esophagram** should be performed to assess esophageal emptying and EGJ morphology.

- **Typical conventional manometric findings:**

- **•Aperistalsis in the distal two-thirds of the esophagus**

- **•Incomplete LES relaxation**

- **•Elevated resting LES pressure**

Chicago Classification

- **Type I (classic achalasia)** – Swallowing results in *no significant change in esophageal pressurization*. By CC-3 criteria, type I achalasia has 100 percent failed peristalsis with a distal contractile integral (DCI, an index of the strength of distal esophageal contraction) <100 mmHg.

- **Type II** – Swallowing results in *simultaneous pressurization that spans the entire length of the esophagus*. According to CC-3, type II achalasia has 100 percent failed peristalsis and *pan-esophageal pressurization with ≥20 percent of swallows.*

- **Type III (spastic achalasia)** – Swallowing results in *abnormal, lumen-obliterating contractions or spasms*. By CC-3 criteria, type III achalasia has no normal peristalsis and premature (spastic) contractions with DCI >450 mmHg·s·cm with ≥20 percent of swallows.

Tratamiento

- **pneumatic dilation** (Patients with the best outcomes after pneumatic dilation are those older than 40 years, women, those with narrow esophageal diameter, and those with a type II pattern by high-resolution manometry) **or**

- **laparoscopic Heller myotomy** with a partial fundoplication, for patients with type I or type II achalasia

- peroral **endoscopic myotomy (POEM)** of choice for type III achalasia because POEM can deliver a longer myotomy that is generally not possible with pneumatic dilation or the Heller . Short-term results of POEM for types I and II achalasia are at least as effective (and perhaps even better) compared with pneumatic dilation and surgical myotomy, data on long-term outcomes of POEM are limited

- For patients who are **not candidates** for surgery, pneumatic dilation or POEM, or who are unwilling to undergo these procedures, we suggest a trial of **botulinum toxin** injection as this may improve swallowing

- For patients who are unwilling or unable to tolerate surgery, pneumatic dilation, or POEM, and have failed botulinum toxin injections, we suggest pharmacological therapy with **nitrates**, isosorbide dinitratae, nitroglycerin

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