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Communicating oesophageal duplication: a case report

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SUMMARY: **Communicating oesophageal duplication: a case report.**

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Duplications of the oesophagus are rare congenital abnormalities and rarely comunicate with the oesophageal lumen. They are commonly associated with other congenital malformations, such as spinal deformities, congenital heart disease, vertebral anomalies, malrotation of the bowel, Meckel's diverticulum.

During a percutaneous endoscopic gastrostomy, performed because of a neurological dysphagia, the endoscopy revealed a very rare case of a 26-year old man affected by Klippel-Trenaunay syndrome, with an asymptomatic oesophageal duplication that communicated proximally and distally.

RIASSUNTO: **Duplicazione dell'esofago: descrizione di un caso clinico.**

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Le duplicazioni esofagee sono rare anomalie congenite che spesso non comunicano con il lume esofageo. Sono comunemente associate con altre malformazioni congenite, come deformità spinali, malattie cardiache congenite, anomalie vertebrali, malrotazioni intestinali, diverticolo di Meckel.

Durante l'esecuzione di una gastrostomia endoscopica percutanea per una disfagia neurologica, l'endoscopia metteva in evidenza un rarissimo caso di sindrome di Klippel-Trenaunay in un giovane uomo di 26 anni, con una asintomatica duplicazione esofagea comunicante sia prossimamente che distalmente.

KEY WORDS: Oesophageal duplication - Klippel-Trenaunay syndrome - Percutaneous endoscopic gastrostomy.
Duplicazione dell'esofago - Sindrome di Klippel-Trenaunay - Gastrostomia endoscopica percutanea .

Introduction

Duplications of the oesophagus are rare congenital abnormalities of obscure origin which account for 10-15% of all duplications of the alimentary tract. They usually manifest during the first year of life (5, 6, 10).

They may involve the thoracic oesophagus but can extend below the diaphragm (3). Oesophageal duplications, either spherical or tubular, rarely communicate with the oesophageal lumen, are usually lined by gastric mucosa and have a smooth muscle coating. They may lie centrally but more usually lie to one side (3, 6, 11).

Duplications are commonly associated with other congenital malformations, such as spinal deformities, congenital heart disease, vertebral anomalies, malrotation of the bowel, Meckel's diverticulum (10).

A very rare case of a 26-year old man affected by Klippel-Trenaunay syndrome, with an asymptomatic oesophageal duplication communicating proximally and distally, is reported.

Case report

A 26-year-old man, born with a port-wine hemangioma involving the neck, presented with dizziness and progressive weakness in the left extremities.

At physical examination, the left arm was longer than the right and showed clear hypertrophy of the soft tissue. There was moderate left hemiparesis, paresis of the soft palate with absence of the pharyngeal reflex, resulting in a high grade of neurological dysphagia.

A percutaneous endoscopic gastrostomy was performed. During this procedure, endoscopy of the upper gastrointestinal tract revealed division of the oesophagus into two lumens at 28 cm. with a narrow mucosal bridge 2 cm. above. It was possible to pass the endoscope through one lumen only for few cm. because of the progressive narrowing of the channel. The principal lumen communicated with the stomach through a normal oesophagogastric junction whereas the accessory lumen communicated distally with a small orifice just below that.

Histologic examination showed normal oesophageal epithelium.

An upright contrast study of the oesophagus with Gastrografin showed a narrow tubular channel projecting from the left posterolateral aspect of the lower third of the oesophagus, slowly emptying into the stomach just below the oesophagogastric junction (Fig. 1).

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Abbreviations: Klippel-Trenaunay syndrome (KTS), percutaneous endoscopic gastrostomy (PEG).

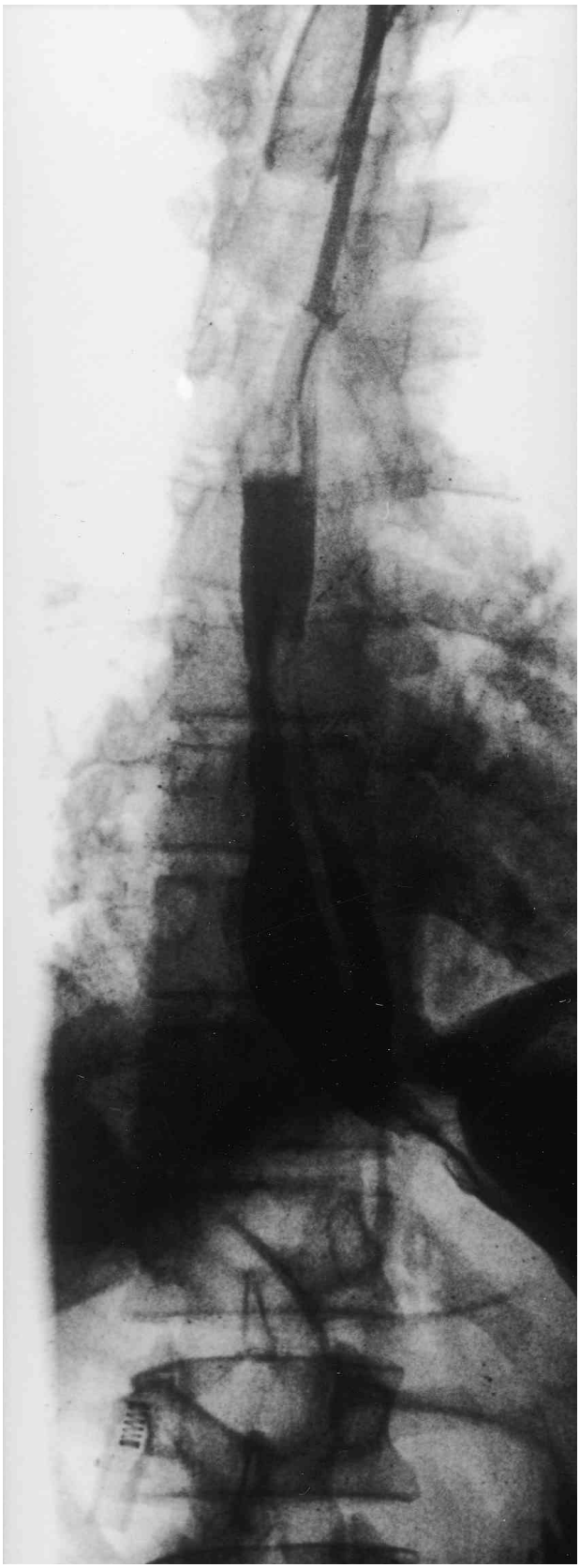

Fig. 1 - Oesophageal X-ray after ingestion of Gastrografin. In the third distal is evident the lumer duplication.

The patient was submitted to magnetic resonance imaging and angiography that revealed a fusiform aneurysm of the right vertebral artery, some aneurysmatic dilatation of the distal tract of the left vertebral artery and a giant sack-like basilar artery aneurysm (Fig. 2).

To reduce the strong compression on the brain stem, occlusion of both the vertebral arteries was accomplished in two steps. The patient tolerated both treatments and the neurological signs remained unchanged.

He subsequently followed a rehabilitation program.

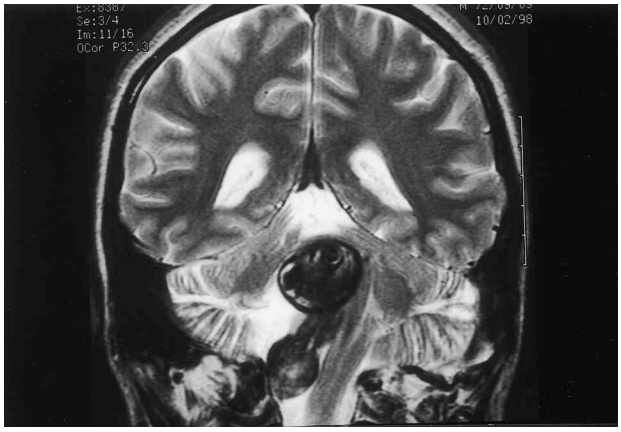

Fig. 2 - Coronal RM: voluminous aneurysm of basilar artery.

Discussion

The criteria suggested by Ladd and Gross in 1944 for diagnosis of duplication are: adherence to some part of the alimentary tract; the presence of a smooth muscle wall, usually in two layers; and an internal lining corresponding to some part of the alimentary tract (10). The particular features of this case are the absence of symptoms due to the double communication, proximal and distal, of the accessory lumen with the alimentary tract, the normal lining with epidermoid epithelium and the association with multiple vascular abnormalities typical of Klipper-Trenaunay syndrome (KTS) (7-9).

In this syndrome, venous varicosities can also involve intra-abdominal and intra-pelvic organs: gastrointestinal bleedings, basically from colonic vascular malformations or oesophageal varices caused by hypoplasia of the portal vein, are also reported (2).

The association of arterial and venous anomalies could be explained by the hypothesis of a congenital disorder resulting from a mesodermal abnormality occuring during fetal development (1). KTS had been suggested to result from the action of a mosaic gene abnormality that is lethal to the gamete when present in all cells of the embryo (4).

This abnormality can also interfere with the development of the oesophagus, either preventing the regression of embryological diverticula (according to the diverticulum theory of Lewis and Thyng) or, in the stage of vacuolation, favouring the isolation of some of these vacuoles that coalesce to form a parallel tube (according to Sir Arthur Keith's vacuolation theory) (10). The variety of clinical features is probably due to the stage of embryogenesis when the noxae operate.

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