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BRIEF REPORT

Incidental diffuse esophageal intramural pseudodiverticulosis. A rare autopsy finding

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KEYWORDS
Esophagus;
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Dysphagia

Abstract Esophageal intramural pseudodiverticulosis (EIPD) is a rare condition of uncertain etiopathogenesis which usually presents with either intermittent or progressive dysphagia. A review of the literature revealed very few studies of its histopathology. We report the case of a 70-year-old diabetic man who died after a road traffic accident. The patient had no history of dysphagia. The entity was discovered incidentally during the medico-legal autopsy. The esophagus showed numerous dilated pseudodiverticula confined to the submucosa and lined by both stratified squamous and cuboidal epithelium. EIPD can remain stable and asymptomatic. This case demonstrates the importance of a thorough investigation during a forensic autopsy.

PALABRAS CLAVE
Esófago;
Pseudodiverticulosis intramural;
Pseudodiverticulitis;
Disfagia

Pseudodiverticulosis intramural difusa esofágica incidental. Un raro hallazgo de autopsia

Resumen La pseudodiverticulosis intramural esofágica (PDIE) es un raro proceso de etiopatogénesis incierta que generalmente se manifiesta con disfagia intermitente o progresiva. Una revisión de la literatura ha revelado muy pocos casos publicados mostrando el aspecto histológico de esta condición. Presentamos el caso de un varón de 70 años de edad, diabético, que falleció tras un accidente de tráfico. El paciente no tenía historia de disfagia. La entidad fue descubierta incidentalmente al practicar la autopsia médico-legal. El esófago mostraba numerosos pseudodivertículos confinados a la submucosa y tapizados por epitelio estratificado

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escamoso y cuboideo. La PDIE puede permanecer estable y asintomática. Este caso demuestra la importancia de una exhaustiva investigación en la autopsia forense.

Introduction

Esophageal intramural pseudodiverticulosis (EIPD) is a rare disease characterized by the development of multiple, small, flask-shaped, epithelium-lined outpouchings within the esophageal wall. These cysts consist of dilated excretory ducts of submucosal mucous glands connecting to the esophageal lumen via narrow openings.[1] The etiopathogenesis remains uncertain. This process has been associated with diverse risk factors, such as reflux esophagitis, achalasia, diabetes mellitus and chronic alcoholism. EIPD shows a bimodal peak incidence in teens and over the fifth decade of life. The predominant symptom is dysphagia that may be accompanied by esophageal stricture.[2] Esophageal contrast radiography is the most sensitive diagnostic method. Because lesions are intramural, endoscopic biopsies usually show acute or chronic esophagitis. Thus, in most cases histological diagnosis is only possible from esophagectomy specimens or on autopsy. Since most cases are clinically diagnosed by radiological study there are few reports describing the histopathological features of this entity.[1,3–6]

We report a case found incidentally at postmortem.

Case report

A 70-year-old man who had died after a road traffic accident had a long clinical history of diabetes, periuretral abscesses and perineal fistulae for which he had undergone various surgical interventions.

The forensic autopsy revealed massive bilateral thoracic trauma, with fractures in almost all the costal arches and a complete fracture of the sternum. There was a chronic ischemic cardiopathy with myocardiosclerosis and coronary atherosclerosis. The esophageal mucosa was white with multiple small orifices extending outward from the upper to the lower thoracic esophageal wall (Fig. 1A). The wall of the esophagus was markedly thickened and cut section showed multiple diverticulum-like cavities (Fig. 1B). Esophageal stricture was not detected. The paraoesophageal ganglia were enlarged. The cause of death was traumatic shock.

Histopathologically, the esophagus showed submucosal fibrosis. There were multiple dilated outpouchings, confined to the submucosa, lined by both stratified squamous (Fig. 2A) and cuboidal epithelium (Fig. 3A) with transitions between these two epithelial types. Adjacent mucosal glands showed ectatic ducts lined by cuboidal epithelium that had undergone oncocytic change (Fig. 3B). A prominent neutrophilic inflammatory exudate with presence of desquamative squamous cells in the lumina of the outpouchings was seen (Fig. 2B). Some of these structures were surrounded by an intense inflammatory infiltrate composed of lymphocytes, eosinophils and plasma cells and occasional production of lymphoid follicles (Figs. 2 and 3). The inflammatory infiltrate was polyclonal with the presence of $CD3^+$ and $CD20^+$ cells. Fungal hyphae or spores were not observed. A PCR for detection of herpesvirus and enterovirus was negative.

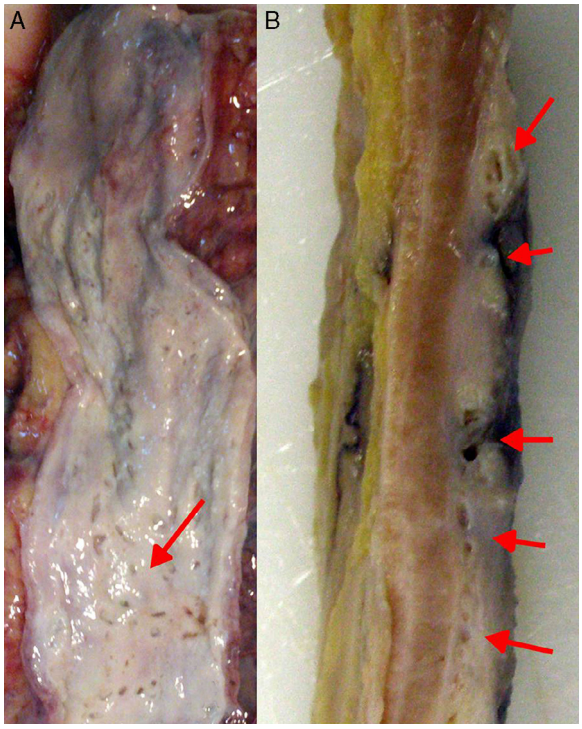

Figure 1 Macroscopic appearance of the esophagus in esophageal intramural pseudodiverticulosis. (A) Multiple openings along the entire length the mucosa. One arrow points to a group of openings. (B) Grossly visible dilated pseudodiverticula confined to the esophageal wall (some of these structures are indicated by arrows).

Discussion

We report this case in order to make a detailed description of the histopathology of EIPD and to highlight the importance of a thorough investigation during autopsy.

The prevalence of EIPD is not well known. In two large radiological studies it represented 0.1% of all patients evaluated for different processes.[7] It affects both genders, although it has a slight male predominance.[8] The average age at presentation in adults is between the sixties and seventies.[2,7] The most common symptom observed in over 80% of patients is acute, intermittent, constant, or progressive dysphagia predominantly for solid foods.[2,9,10] However, the process can be asymptomatic.