Inflammatory myofibroblastic tumor as adverse outcome of eosinophilic esophagitis

An emergency esophagogastroduodenoscopy performed in a dyspeptic 19-year-old girl with hematemesis, abdominal pain, and anemia (Fig. 1) revealed a polypoid esophageal mass with distal erosion. A subsequent CT scan confirmed the bulky lesion (14 × 4 × 3.5 cm) extending from the carina to the cardia level. Both extraesophageal extension and nodal and extranodal metastases were excluded. The extent and location of the mass ruled out any endoscopic resection and a total esophagectomy with gastric pull-up was performed. Both histology and immunophenotyping (positive for vimentin, MNF116, and smooth muscle actin; negative for S100 and CD117) were consistent with the “classical variant” of inflammatory myofibroblastic tumor (IMT). A prominent eosinophilic infiltrate (Fig. 2 a–c) was also present, spreading from the lesional to the extralesional area, where the squamous epithelium featured rich eosinophilic infiltrate, fully consistent with eosinophilic esophagitis (Fig. 2 d).

Within the upper gastrointestinal tract, IMTs are rare [1], polymorphic clinicopathological entities that have been associated with gastric ulcers [2], ischemic disease, and gastroesophageal reflux [3]. In accordance with these clinical settings, the hypothesis of IMT as an “exaggerated reparative response” has been established. In eosinophilic esophagitis, the promoting role for eosinophils in the onset of subepithelial fibrosis is supported by the high transcript levels of both interleukin-5 and TGF-β detected in tissue samples from lesions [3–5]. Most recently, this hypothesis has been validated in murine models of eosinophilic esophagitis, where eosinophilia leads to collagen accumulation, finally resulting in esophageal wall remodeling [5]. A causative role for eosinophilic esophagitis is further sustained by the clinical finding that more than 50% of cases of eosinophilic esophagitis coexist with fibrotic esophageal strictures (e.g., Schatzki rings and esophageal webs), which might be seen as part of the same “tumorigenic field” in which IMT may develop.

The clinical observation of a young patient with concomitant eosinophilic esophagitis and IMT, supported by recent experimental and molecular evidence, suggests that IMTs can definitely be considered among the unfavorable outcomes of (long-standing) eosinophilic esophagitis.

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