CLINICAL CASE

Mixed adenocarcinoma and squamous cell carcinoma arising in a gastric duplication cyst

Duplication gastrique de l’adulte, dégénérescence mixte en adénocarcinome et carcinome épidermoïde

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Summary

Malignant transformation of duplication cyst is a rare condition. The authors report the original case of a degenerated gastric duplication cyst in a 67-year-old patient. The histologic examination revealed a gastric duplication cyst infiltrated with both adenocarcinoma and squamous cell carcinoma. Local carcinomatosis was found at laparotomy. The patient died six months after complete macroscopic resection of the lesion, with metastatic disease.

Résumé

La dégénérescence maligne d’une duplication est un évènement rare. Les auteurs rapportent le cas d’une dégénérescence d’une duplication gastrique chez une patiente de 67 ans. L’étude histologique a révélé l’existence d’une duplication gastrique infiltrée d’un contingent adénocarcinomateux et de carcinome épidermoïde. Lors de l’intervention, une carcinose a été constatée. Malgré une chirurgie extensive, la patiente est décédée d’une évolution métastatique de sa maladie à six mois.

Introduction

Alimentary tract duplications are rare congenital malformations mostly diagnosed during childhood. Malignant lesions arising within gastric duplications in adults are exceptional and have been often reported in the literature.

Case report

A 67-year-old woman was referred at our institution for an abdominal mass that she had detected herself three weeks previously. Her medical and family history was unremar-
Mixed adenocarcinoma and gastric duplication cyst

Her only other complaint was a weight loss of five kilograms over the previous two months.

A physical examination revealed an epigastric mass. Laboratory test results were normal except for an elevated Ca 19-9 level of 280 UI/ml (Normal < 37 UI/ml), CEA level was normal. Blood γGT levels was also elevated (twice the normal value). An abdominal ultrasonography revealed an abdominal cyst of heterogeneous density seeming to belong to the pancreatic head. Her left bile duct was enlarged and some ascites was present. A computed tomography scan also detected the cyst at the head of the pancreas as well as the expansion of the left bile duct and additionally revealed two more cysts within the pancreas. Mesenteric and aortic lymphadenopathies were also noticed (Fig. 1a). Upon laparotomy, a gastric mass was discovered, which traversed the gallbladder and compressed the left bile duct (Fig. 1b). Peritoneal carcinomatosis was also revealed. The patient underwent radical distal gastrectomy with a Billroth 2 gastrojejunal anastomosis and cholecystectomy by en bloc resection. Histological examination (Fig. 1c,d) revealed a cystic mass developed in the wall of the stomach, corresponding to a gastric duplication. The cystic mass was infiltrated by an adenocarcinoma with a squamous component in the invaded gallbladder. The patient died within six months after operation with a generalised metastatic evolution (liver) under adjuvant chemotherapy consisting of five fluorouracil, folinic acid and cisplatin.

Discussion

Duplication is an unusual congenital anomaly that can involve any part of the gastro-intestinal tract. Their pathogenesis remains controversial. Duplications are usually defined as being intimately attached to and contiguous with the bowel wall, lined by mucous membrane and surrounded by at least one well-developed smooth muscle layer [1]. Two morphological types of duplications can be distinguished: the most frequent cystic variety (75%) that does not usually communicate with the bowel lumen and the tubular variety (25%), which communicates with the lumen in almost 75% of cases [2,3]. The ileum is the most common site of isolated gastro-intestinal duplication, followed by the oesophagus and colon. The stomach is the site of only 4% of all duplications. The latter are located on the distal greater curvature in 75% of cases, often cystic and non-communicating. Gastric duplication is associated with other anomalies in one

Figure 1  Abdominal CT-scan during the arterial phase after injection showing the paragastric cyst overlapping the gallbladder, perihepatic ascites and dilatation of the left bile ducts (panel a). Complete resection of the duplication cyst with the distal gastrectomy (panel b). Histologic features (Hématoxylin–phloxine–safran × 100), adenocarcinomatous component (panel c), squamous cell carcinoma component involving the gallblader (panel d).

Scanner abdominal (temps artériels) montrant le kyste para-gastrique au contact de la vésicule biliaire, une ascite péri-hépatique et une dilatation des voies biliaires intra-hépatiques gauches (figure 1a). Pièce opératoire emportant complètement la duplication kystique, associée à une gastrectomie distale (figure 1b). Aspect histologique : composante adénocarcinomateuse (coloration Hématoxyline-Phloxyne-Safran ; grossissement × 100) (figure 1c). Composante carcinomateuse à cellules squameuses envahissant la vésicule biliaire (figure 1d).
Table 1

Characteristics of the degenerated gastric duplications reported in the literature.

<table>
<thead>
<tr>
<th>Author, year of publication</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Histology</th>
<th>Size (cm)</th>
<th>Extension</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mayo, 1955 [9]</td>
<td>64</td>
<td>F</td>
<td>Asthenia</td>
<td>ADK</td>
<td>6</td>
<td>Gastric wall</td>
<td>Alive, 12 months</td>
</tr>
<tr>
<td>Tregier, 1969 [10]</td>
<td>50</td>
<td>M</td>
<td>Upper gastroduodenal obstruction</td>
<td>Epithelial carcinoma</td>
<td>17</td>
<td>Unknown</td>
<td>Unknown</td>
</tr>
<tr>
<td>Coit, 1992 [4]</td>
<td>72</td>
<td>F</td>
<td>Abdominal pain</td>
<td>ADK</td>
<td>4</td>
<td>Gastric wall</td>
<td>Alive, 12 months</td>
</tr>
<tr>
<td>Mamiya, 1996 [12]</td>
<td>71</td>
<td>M</td>
<td>Back pain</td>
<td>ADK</td>
<td>8</td>
<td>Wall of the cyst</td>
<td>Alive, 1 month</td>
</tr>
<tr>
<td>Kuraoka, 2005 [5]</td>
<td>40</td>
<td>M</td>
<td>Abdominal mass</td>
<td>Mixed ADK and squamous cell carcinoma</td>
<td>18</td>
<td>Gastric wall and peritoneal nodules</td>
<td>Local recurrence, liver metastasis, 7 months</td>
</tr>
<tr>
<td>Barussaud, 2006 present case</td>
<td>67</td>
<td>F</td>
<td>Abdominal mass</td>
<td>Mixed ADK and squamous cell carcinoma</td>
<td>18</td>
<td>Gastric wall</td>
<td>And peritoneal nodules</td>
</tr>
</tbody>
</table>

ADK: adenocarcinoma; M: male; F: female.
From Kuraoka et al., updated data.

Third of cases: oesophageal atresia or diverticula’s duplication, spina bifida or vertebral abnormalities [1–3]. They are usually diagnosed during childhood, before the age of 12, because they tend to produce symptoms early on, the most common being bowel obstruction. Sixty-seven percent of gastric duplications are discovered during the first year of life [1,4]. The associated presenting symptoms are non-specific, such as vomiting, nausea, epigastric pain or mass and weight loss. Complications such as bleeding, infection, obstruction, ulcer or pancreatitis rarely occur [5]. Malignant changes of the gastric duplication are exceptional events but are more frequently reported in the literature than malignancy in rectal duplication cyst [6] or duodenal duplication cyst [7]. We could not find any influence of the duplication variety (tubular or cystic) on the malignant transformation in the literature [8]. Kuraoka et al. [5] reported six cases of malignant transformation of the gastric duplication previously published and add one more with liver metastasis. Two of these cases involved an abdominal mass and all had lost weight, as in our case (Table 1) [4,5,9–12]. Adenocarcinoma arising in the cyst is the most frequent presentation of degenerated lesions (five of the six cases published in the literature). We report the first case of a degenerated lesion with both adenocarcinoma and squamous cell carcinoma arising in the duplication cyst. In fact, we can speculate that the mucosae lining the cyst is covered by a poorly differentiated epithelium that can lead to different histological features such as adenocarcinoma or squamous cell carcinoma.

Echoendoscopy currently seems to be the best modality for demonstrating gastric duplications [2]. This technique can show both layers of the cyst wall, contiguous with the gastric wall. CT-scan can also be useful for its diagnosis, demonstrating small duplications that may be missed with a conventional barium or ultrasound study or for assessing the size of the cyst and its relationship with the neighbouring organs. This examination also provides the possibility, as in our case, of detecting signs of malignancy: lymphadenopathies, ascites or liver metastasis [1]. Although carbohydrate antigen 19-9 might be markedly elevated, it is not clearly associated with malignancy [13]. Our case highlights the fact that once a duplication cyst has been diagnosed, even an asymptomatic one, it should be removed because of the risk of malignant changes and a complete resection of the duplication should be the procedure of choice. Some authors suggested other treatment options for the non-communicating cyst that can be converted into a communicating one by endoscopic or surgical procedure but symptoms are often recurrent and the cyst is finally not removed [2]. When a cancer is suspected, as in our case, a subtotal or total gastrectomy with reconstruction is unfortunately the only solution with a poor prognosis [1,2,14].

Conclusion

Malignant transformations in gastric duplication cysts are extremely rare and their preoperative diagnosis remains difficult. Our case is the seventh to be reported and certainly original with both adenocarcinoma and squamous cell carcinoma arising in the cyst. This emphasizes the fact
that malignant degeneration of duplication cyst can lead to aggressive treatment option and poor prognosis. Elective complete resection of the lesion should be the rule when the diagnosis of duplication cyst is suspected.

References