Heteropic gastric pancreas associated with type II esophageal atresia and presenting with acute gastrointestinal bleeding

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1. Abstract

Heterotopic pancreas is defined as pancreatic tissue found outside its normal localization without vascular or anatomic communication with the pancreatic gland. It is usually diagnosed incidentally, but it may also be responsible for acute clinical pictures. We discuss an interesting case of ectopic pancreas associated with type II esophageal atresia and presenting with gastrointestinal acute bleeding.

2. Introduction

Heterotopic pancreas is a rare congenital anomaly due to the development of pancreatic tissue outside its common localization. It can be associated with other congenital anomalies. Affected patients are often asymptomatic and they require a conservative follow – up. We report the case of a boy affected by type II esophageal atresia who has been diagnosed with ectopic gastric pancreas.

3. Case Report

A 20 days old male (R.G.) was admitted to our Unit, from another Hospital, with the diagnosis of esophageal atresia of 3rd type. The baby was born at 40th weeks of gestational age with cesarean section. He weighed 3,1 kg and he experienced respiratory distress. In order to avoid further complications he underwent to right posterior-lateral toracotomy with a section of
the trachea-esophageal fistulae 24 hours after birth but it was impossible to create a direct anastomosis. R.G. was later transferred to our Hospital since we have a specialized Pediatric Surgery Unit supported by a Pediatric Intensive Care Unit.

In our hospital, we performed a gastrostomy and we evaluated the esophageal stumps. When the baby was 35 days old, an esophagus-esophageal anastomosis was created. No complications occurred during the surgical procedure and there were not any short term complications so the baby was discharged with ranitidine therapy 30 days after surgery.

Figure 1. Endoscopic aspect of the gastric tumor compatible with heterotopic pancreas

Two months later the child was re-admitted with the diagnosis of pneumonia due to gastro-esophageal reflux. Signs of severe esophagitis were identified during the EGDS together with the presence of a pre-pyloric broad-based polypoid tumor with a central crater, diagnostic for ectopic pancreas (Fig. 1). Multiple biopsies were performed during the procedure. The histological examination displayed normal gastric mucosae, it confirmed the presence of heterotopic pancreas and the research of H. Pylori was negative. Therefore a therapy with omeprazole, ranitidine and Gaviscon® was set up. Complete macroscopic resolution of the esophagitis was achieved after 15 days of therapy.

Since the baby did not present other symptoms we started our follow-up protocols for patients with esophageal atresia that provides for the performance of endoscopic evaluations every six months in the first year, then only in presence of disphagia or other symptoms.

At the next control parents reported that R.G. had experienced one episode of caffean vomit and hematemesis two weeks earlier. We immediately performed an EGDS: the esophagus was patent and lesion-free without any sign of previous bleeding. An endoscopic examination of the upper respiratory tract was performed too. Also in this case the origin of the bleeding was not identified. Apparently the gastric mucosae was not damaged too, and the tumor was still present. We decided to keep a conservative approach based on the use of an antireflux therapy and on the endoscopic evaluation every 12 months unless the appearance of clinical signs or symptoms.

4. Discussion and Conclusions

Heterotopic pancreas is defined as pancreatic tissue found outside its normal localization without any connection with the pancreatic gland [(1, 2). Theories about its formation (such as pancreas metaplasia or pancreatic split during foregut rotation) have been proposed but the exact mechanism is unknown.

Ectopic pancreas is usually found within the abdominal digestive tract, in particular in the stomach antrum, duodenum or jejunum. The ileum is the most common location. Other ectopic pancreas sites have been described (lungs, liver, spleen, gallbladder, esophagus and Meckel diverticulum). According to Heinrich’s classification, there are 3 types of heterotopic pancreas: type 1 expresses all components of the normal pancreas (ducts, acini and endocrine’s islets), type 2 has no islets and type 3 is called adenomyomas (ducts with few acins).
Heteropic gastric pancreas associated with type II esophageal atresia

It is a rare entity in children (girls are affected more than males [3] and it is often discovered incidentally. The lack of symptoms leads to underestimate the real incidence. This is confirmed by the fact that heterotopic pancreas is found in 1/500 surgery of the upper abdomen and in 0.6-5.6% of autopsies (1,3,4). Clinical manifestations depend on the location of the aberrant pancreatic tissue. The symptoms are due to the release of irritant hormones and to the activation of the inflammatory cascade. Pain and bleeding can occur when the lesion is in the stomach wall. In cave organs the excess tissue can obstruct the outflow or be the stimulus for intussusceptions (13.2%) (5). In our patient we think that the ectopic pancreatic secretions may have caused gastric irritation and occult bleeding.

Recent works have stated that there is a connection between some congenital anomalies, such as esophageal atresia, and the presence of pancreatic rest. (2,6) According to Moreau et al. esophageal atresia is associated with gastric heterotopic pancreas in 18.7% of cases irrespective of other malformations (2,7). J. Park reported the case of a female infant affected by esophageal atresia associated with pancreatic and gastric ectopic pancreas. The heterotropic esophageal pancreas was found incidentally during the creation of an esophagoesophagostomy and the gastric heterotopy remained undiagnosed until it led to gastric perforation. Both lesions were excised completely and the diagnosis was confirmed by histological examination (pancreatic acini and ducts in the muscular layer). In our case we identified the lesion during routine endoscopy and we decided to avoid surgery. Biopsies performed during the EGDS were negative probably because we did not reach the deep ectopic tissue. Nevertheless, we identified the 3 main macroscopic features of gastric heterotopic pancreas (we described an antral tumor with a central umbilication and intact mucous membrane) (1). Fine needle aspiration biopsy can be useful for diagnosis while imaging techniques (contrast x-ray, CT and US scan) lack specificity.

Heterotopic pancreas tends to reduce its size until it disappears. The possibility of malignant transformation is described in less than 30 cases in the literature (89 but there is not a common consensus on the existence of this correlation. In our case we opted for a conservative approach since the tumor did not involve the gastric mucosa and it was not big enough to create an obstruction. The lesion was below 1.5 cm of diameter that is thought to be the limit for clinical relevance (9). In addition a possible surgical complication could have been the damage of the pylorus because it was really close to the lesion. However, surgical or endoscopic removal should be considered in rapidly growing tumors or in lesions found during laparotomy or in children with relevant symptoms.

5. References


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