A Case of Anomalous Pancreaticobiliary Junction (APBJ) and Primary Gallbladder Carcinoma

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Case Report:

A 41-year-old female with no significant past medical history presented with a 2-week history of abdominal pain. The pain was located in the epigastric and right upper quadrant area and associated with nausea. Her past medical history was significant only for scoliosis. She smoked less than a pack of cigarettes a day for 25 years. Family history revealed that a cousin and 2 of her aunts had breast cancer. The patient’s physical was unremarkable except for right upper abdominal tenderness to palpation, but no palpable mass. An abdominal CT scan showed what appeared to be advanced metastatic disease to the liver and surrounding lymph nodes with a possible gallbladder mass. (Figures 1A, B) EUS was performed to assess and stage the gallbladder mass and examine for evidence of potential obstruction of the intestine or biliary tree. Informed consent was obtained for the procedure. At the time of the EUS (Video 1), the tumor appeared to arise primarily from the gallbladder and infiltrated into the liver. Liver metastasis and abnormal adenopathy were found in the extragastric and extraduodenal spaces. No pathologic features of metastasis were noted above the diaphragm. Both radial array and linear EUS examination of the pancreaticobiliary tree identified a long common channel communicating between the distal common bile duct and the pancreatic duct. The
common bile duct was not dilated. These features suggested an anomalous pancreaticobiliary junction without choledochocele. Fine needle aspiration of the celiac node (also shown in figure 1B) revealed small cell carcinoma that was TTF-1 (thyroid tissue factor) negative, suggesting an extrapulmonary origin. A dedicated chest CT showed no evidence of pulmonary disease.

Discussion

Anomalous pancreaticobiliary junction (APBJ) is an unusual congenital abnormality in which there is a distal common channel for the bile duct and pancreatic duct, generally of 1.5 cm or greater length. About 2/3 of the cases of APBJ are associated with a type I choledochocele, defined as bile duct cross section of 1 cm or greater\(^1,2\). Endoscopic ultrasound has been reported as a good method for the detection of APBJ\(^3,4\).

APBJ is thought to predispose to biliary duct reflux and is strongly associated with gallbladder adenocarcinoma\(^1,2,5\). At first, the patient in this report seemed to fit that description as imaging demonstrated a grossly abnormal gallbladder, along with imaging showing suspected nodal and liver metastasis. The biopsy result in this case, however, was quite surprising with histochemistry and imaging suggesting that this was an extrapulmonary small cell carcinoma, likely of gallbladder origin.

Extrapulmonary small cell carcinoma is a relatively unusual neoplasm, and is estimated to occur in less than 5% of small cell cancer cases\(^6,7\). About half of the cases of extrapulmonary small cell carcinoma seem to have a gastrointestinal origin, but even in this group, gallbladder primary cancers are distinctly abnormal\(^8,9\). The cases reported to have primary gallbladder cancer had a higher incidence of cholelithiasis, female gender, and a significant proportion had mixed histology suggesting that these tumors can be part of a histological progression in the spectrum of more routine gallbladder adenocarcinoma\(^6-11\). The association with tobacco use is present, but is not as strong as it is for small cell carcinoma of the lung\(^6\).

We are unaware of a case of ABPJ associated with an extrapulmonary (gallbladder) small cell carcinoma. Since extrapulmonary small cell carcinoma can be at the end of a spectrum of histology from a nonsmall cell origin, and since the patient had a condition which predisposed her to gallbladder cancer, it is reasonable to assume the APBJ was related to her tumor and the histology to her smoking history. The rarity of this combination of events however, makes all conjectures somewhat speculative.

In summary, this report identifies a young woman with an advanced malignancy that appears to be a gallbladder cancer. Her evaluation revealed APBJ without choledochocele. Her biopsies showed a small cell cancer.
References:


