Short reports

Adenocarcinoma in cystic fibrosis

TME DAVIS, EH SAWICKA

From the Brompton Hospital, London

The occurrence of malignancy in a patient with cystic fibrosis is a rare event. Most such tumours are those found at a young age in the normal population.1 A report of cholangiocarcinoma in a 27 year old woman with cystic fibrosis2 has, however, raised the possibility that cystic fibrosis predisposes to certain malignancies. We present the second recorded case of carcinoma in a young woman with cystic fibrosis.

Case report

A 23 year old woman presented with abdominal pain. Cystic fibrosis had been diagnosed at 12 months of age, when a sweat test was performed (sweat sodium 93 mmol (mEq)/l) because of symptoms of pancreatic insufficiency. She was treated with a low fat diet and, when she was 13 years old, pancreatic supplements were added. Subsequently her bowel action was regular with well formed stools. At presentation she was 95% of the mean predicted body weight for her sex and height.

Her respiratory symptoms were never severe. At 10 years of age she developed cough with sputum production and required intermittent antibiotic treatment. Pseudomonas aeruginosa was first isolated from the sputum two years later. From the age of 16 years her lung function was 60% and 80% of that predicted for age, sex and height, and she noted little sputum production.

At presentation she complained of vague, right sided upper abdominal pain, which had been present for 2 weeks. It was dull, poorly localised, unrelated to meals, and not associated with nausea or vomiting. It was not controlled by simple analgesics.

On auscultation there were scattered inspiratory crackles over both lung fields. A 5-7 cm diameter mass was palpable below the right costal margin and was tender on inspiration. A blood count was normal and the erythrocyte sedimentation rate was 25 mm in one hour. Random plasma glucose concentrations were always below 10 mmol/l. A serum amylase level and results of liver function tests (including serum bilirubin) were normal apart from a raised γ glutamyl transferase concentration of 52 IU/l (normal range 3-14). A midstream specimen of urine was sterile on culture. A plain abdominal radiograph showed appearances consistent with constipation.

Upper abdominal ultrasound examination showed a large gall bladder, consistent with the physical findings. No gall stones were seen. The pancreas appeared atrophic, but its head contained a bilobular mass. Abdominal computed tomography confirmed these findings. An endoscopic retrograde cholangiopancreatogram showed a narrowed pancreatic duct consistent with extrinsic compression by tumour.

Needle aspiration of the pancreatic mass was performed under ultrasound control. An adjacent mass, thought to be an enlarged lymph node, was also identified. Histological examination of seven tissue aspirates showed groups of adenocarcinoma cells which might have arisen in glandular or ductal tissue.

Surgical treatment was considered inappropriate because of the size of the tumour and the presence of an enlarged lymph node in a patient with a chronic debilitating disease. Severe back pain developed, but was controlled with a coeliac plexus nerve block and oral analgesia. She subsequently deteriorated rapidly with anorexia, malaise, and weight loss of 12 kg over a two month period, and became deeply jaundiced.

She died within six months of presentation. At necropsy a 6 cm diameter tumour was found replacing the head of the pancreas, and histological examination confirmed the diagnosis of adenocarcinoma.

Discussion

In a young woman carcinoma of the pancreas is rare3 and its chance association with cystic fibrosis, although possible, seems unlikely. This is the second reported case of carcinoma occurring in an organ directly affected by cystic fibrosis, which raises the possibility that the disease predisposes to certain forms of malignancy. Such a predisposition might become more obvious as the life expectancy of patients with cystic fibrosis increases.

Early detection of pancreatic carcinoma is difficult. Many patients have a raised serum alkaline phosphatase level at presentation,4 but this would be difficult to interpret in cystic fibrosis and it was not a feature here. Raised serum concentrations of the tumour marker carcinoembryonic antigen (CEA) are suggestive of intra-abdominal malignancy.5 In this case the serum CEA concentration at the time of histological diagnosis was 13 μg/l, at the upper limit of the range of results obtained from eight other patients with cystic fibrosis of similar age (normal range <10). CEA concentrations may not, however, be useful in identifying patients with intra-abdominal malignancy unless a level several times the upper limit of normal is found.4 Thus the single CEA estimation at the time of presentation would not have contributed to earlier diagnosis in this case.

Upper abdominal pain is common in patients with cystic fibrosis.
fibrosis and is usually due to constipation. Gall stones, pancreatitis, duodenal ulceration, narrowing of the second part of the duodenum, and meconium ileus equivalent are less frequent but well recognised complications. In the investigation of persistent pain, however, the possibility of these rare tumours should be borne in mind. Unusual ultrasound or computed tomography appearance of the pancreas and hepatobiliary system should not be dismissed as distortion of structure due to cystic fibrosis.

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References

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