A Case of Possible Autoimmune Pancreatitis

Dr S Buxton, Dr P Bellis, Dr M Nayar, Dr B McLain, Dr A Mukhapadhyay, Dr RS Parmar, Dr J Thomas

The authors report no conflicts of interest

Introduction
Autoimmune pancreatitis (AIP) is a rare paediatric condition with management driven by adult guidelines. However, evidence suggests that paediatric disease is a distinct entity which may follow a different clinical course. Management and long term outcomes are unclear with limited evidence to suggest any management strategy is superior to another.

Case
A previously well 12-year-old Caucasian male presented with a week of pruritus and bruising with a 48-hour history of diarrhoea and jaundice. There was no relevant family history. Examination revealed petechiae and ecchymosis but no positive abdominal findings. Initial imaging (ultrasound) had suggested a mass in the pancreatic head obstructing the common bile duct (CBD). He subsequently underwent further imaging and had a ERCP where a pancreatic biopsy was performed along with a CBD stent being inserted.

Blood results
- Bilirubin 277μmol/L (201μmol/L conjugated)
- Alanine transaminase 88unit/L
- Alkaline phosphatase 802unit/L
- Gamma glutamyltransferase 79unit/L
- International normalised ratio 0.9
- Amylase and lipase were normal throughout.
- Viral hepatitis screen negative
- Liver autoimmune screen negative
- Lipid profile normal
- Alpha 1 Antitrypsin normal
- IgG4 0.6 g/L (within normal range)
- Faecal elastase 155μg/g
- Glucose 6.4 mmol/L
- Chronic pancreatitis genetic panel awaited

MRCP
Marked intra and extrabiliary duct dilatation to mid CBD (at insertion into pancreas). Pancreatic duct is not objectively dilated, but visible in the tail and neck of the gland and obliterated/not seen in the head. This infers a 3cm mass in the pancreatic head (see image to right).

Histology
- Mild chronic inflammation with minor acinar atrophy, intralobal oedema and some fibrosis.
- No overt plasma cells or eosinophils
- No storiform nature to fibrosis
- Obliterative phlebitis lesions not seen
- Immunochemistry for IgG4 did not reveal positive plasma cells
- No evidence of neoplasm

EUS
- Diffusely enlarged pancreatic parenchyma with a prominent head of pancreas with distal biliary dilatation
- Pancreatic duct not seen

ERCP
- Smooth single stenosis (max length 20mm) in the distal and mid common bile duct.
- Pre stenotic dilatation present
- Straight plastic stent inserted into the CBD

Bloods normalised by 4 weeks post stent insertion as per diagram (red line denotes stent insertion date)

Repeat MRCP 3 months post stent insertion showed no biliary duct dilatation, some irregularity of the distal pancreatic duct but no dilatation or strictureting and an otherwise normal pancreas.

The stent is planned for removal electively with no maintenance treatment and close monitoring.

Summary
This case does not meet the diagnostic criteria for type 1 or 2 AIP using adult criteria. However this child’s presentation (and radiological findings) were suggestive of AIP. Often children receive steroid therapy as a treatment modality but thus far this has not been required in this case.

Conclusion
Autoimmune pancreatitis is a rare condition in children and management is often extrapolated from adult guidelines. Collaborative working is required for the creation of paediatric guidelines to aid diagnosis and management of this rare condition.

Diagnostic Limitations in Children using Adult Criteria
- IgG4 is rarely positive in children therefore Type 1 AIP is excluded
- Type 2 AIP requires histological confirmation
- ERCP is not always readily available in the paediatric setting
- Paediatric pancreatic specimens are more likely to be obtained by fine needle aspiration rather than tru-cut biopsy therefore reducing the diagnostic yield from specimens

Management
No medical management was required.

References