Diffuse myelomatous infiltration of the pancreas

Evan Kwong, David Sanders, Stephen Kwong

DESCRIPTION
A man in his early 60s with a 3-year history of multiple myeloma (MM) presented to his oncologist during his outpatient appointment with increasing kappa free light chain (102 mg/L, normal range 3.3–19.4 mg/L), increasing M protein (21.6 g/L, normal range <30 g/L) and painless jaundice. He was first diagnosed with MM after he presented with hip pain and was found to have lytic bone lesions. His initial 6 months of treatment included six cycles of cyclophosphamide, bortezomib and dexamethasone chemotherapy followed by autologous stem cell transplant (ASCT) and lenalidomide maintenance therapy. Unfortunately, the patient relapsed 1 month following his ASCT and was placed on daratumumab, bortezomib, cyclophosphamide and dexamethasone. Bortezomib and cyclophosphamide were discontinued after eight cycles of treatment, although cyclophosphamide was reintroduced roughly 2 years after initial diagnosis due to increases in the patient’s M protein levels.

He presented with painless jaundice 1 year later. A CT scan showed marked pancreatic enlargement with no significant peripancreatic oedema or focal mass (figure 1). The marked and diffuse increase in pancreas size was more consistent with an infiltrative pathology than an inflammatory process. Interestingly, he did not exhibit pancreatic insufficiency. His jaundice was relieved by an endoscopic retrograde cholangiopancreatography and stenting. His cholangiogram and pancreatogram showed parallel strictures in the head of the pancreas. Given that his brushings were non-diagnostic, an endoscopic ultrasound guided fine needle aspiration biopsy of both the head and body of the pancreas revealed diffuse infiltration of clonal plasma cells consistent with extramedullary spread of MM (figure 3) in both samples.

Extramedullary disease (EMD) is present in 6%–20% of relapsed MM patients, with prevalence increasing over threefold between 2005 and 2014. EMD may be more common following ASCT. MM patients with EMD face a poor prognosis. EMD affecting soft tissues is often highly aggressive and associated with poor outcomes, even when compared with skeletal EMD. One study found the mean survival time of MM patients presenting with nonskeletal EMD on relapse to be 38 months.

Pancreatic EMD is rare, with one review finding 63 examples of pancreatic plasmacytomas between 1950 and June 2016. Of these cases, only two involved the tail/body of the pancreas. Diffuse enlargement of the entire pancreas in the context of MM had been reported previously due to amyloidosis, but to our knowledge the diffuse myelomatous infiltration of the pancreas seen in this case had not been previously reported.

Curative radioablation of the cancer was not possible, as the volume of affected tissue would require dosage exceeding renal and spinal...
Images in...

Figure 3  Fine needle aspiration biopsy of the head and body of the pancreas confirmed diffuse myelomatous infiltration of the organ. Images shown were taken from the pancreatic head, but cytology of the pancreatic body did not significantly differ. (A) Haematoxylin and eosin stain. (B) Kappa light chain immunohistochemistry (IHC) stain, consistent with kappa light chain restriction. (C) Positive CD138 IHC stain, consistent with myeloma but non-specific. (D) Negative CD20 IHC stain, more consistent with multiple myeloma than B-cell lymphoma. (E) Negative cytokeratin AE1/AE3 IHC stain, inconsistent with pancreatic carcinoma. (F) Negative lambda light chain IHC stain, consistent with kappa light chain restriction. (G) Congo red stain, demonstrating amyloid deposition. Green birefringence was seen in this sample under polarised light (not shown). (H) Negative CD3 IHC stain, inconsistent with T-cell involvement.

Learning points

► Extramedullary multiple myeloma (MM) is increasingly common and may present with diffuse infiltration of solid organs. This case demonstrated diffuse infiltration of the pancreas with painless jaundice.
► If cross-sectional imaging of an MM patient detects concern for organ involvement, imaging-guided biopsy is advised.
► For gastrointestinal involvement, endoscopic ultrasound with fine needle aspiration can be used to confirm diagnosis.

Contributors  The following authors were responsible for drafting of the text, sourcing and editing of clinical images, investigation results, drawing original diagrams and algorithms, and critical revision for important intellectual content: EK, DS, SK. The following authors gave final approval of the manuscript: EK, DS, SK.

Funding  The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests  None declared.

Patient consent for publication  Consent obtained from next of kin.

Provenance and peer review  Not commissioned; externally peer reviewed.

Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

REFERENCES