Painless jaundice with pancreatic mass in 79-year-old man receiving bortezomib for relapsed multiple myeloma

INTRODUCTION

A 79-year-old man presented with signs of biliary obstruction (bilirubin 82 $\mu\text{mol/L}$, alanine aminotransferase 358 IU/L and alkaline phosphatase 250 IU/L) and normocytic anaemia (haemoglobin 120 g/L). He had a 4-month history of relapsed multiple myeloma and had been treated with five cycles of bortezomib (a proteasome inhibitor). Transabdominal ultrasound demonstrated an ill-defined mass in the pancreatic head associated with upstream biliary dilatation. CT demonstrated multiple uniformly enhancing soft tissue masses throughout the pancreas with enlargement of the pancreatic head, in particular, and associated biliary obstruction. There was a direct invasion into the main portal vein, suggesting a solid malignancy. There was no evidence of distal metastases, ascites or peritoneal carcinomatosis (figure 1). Immunoglobulin (Ig) G4 levels were normal but IgG2 levels were elevated (23.4 g/L). Cancer antigen 19-9 was mildly elevated (32 kU/L). Endoscopic retrograde cholangiopancreatography and biliary stent placement were performed

Figure 1 CT—single slice from a CT through the upper abdomen demonstrating enhancing soft tissue masses within the pancreas (asterisk), invasion of the main portal vein (arrow) and a dilated common bile duct (CBD) (arrowhead).

Figure 2 ERCP—single fluoroscopic image from an ERCP demonstrates a dilated CBD with a long, smooth stricture in the mid duct (arrows). ERCP, endoscopic retrograde cholangiopancreatography.

(figure 2), with a fall in bilirubin to 39 $\mu\text{mol/L}$. Biliary brush cytology was non-diagnostic.

Endoscopic ultrasound demonstrated a globally abnormal and expanded gland, which was predominantly hypoechoic but with several foci of mixed echogenicity and evidence of ascites (figure 3). A 22 g fine needle biopsy (Acquire; Boston Scientific, Nantucket, Massachusetts, USA) was performed with three passes into the body of pancreas.

QUESTION

What is the most likely diagnosis and what differentials should be considered?

What supplementary investigations should be considered on the tissue sample?

ANSWER

Histopathology of biopsy cores demonstrated diffuse infiltration by atypical plasma cells (figure 4). Immunohistochemistry showed an abnormal plasma cell phenotype consistent with infiltration of the pancreas by extramedullary myeloma. Without

Figure 3 EUS—EUS image showing replacement of the pancreatic head by a large, irregular, hypoechoic mass (marked by callipers). EUS, endoscopic ultrasound.

Figure 4 Histopathology—H&E stained section (×20 magnification) showing atypical plasma cells centrally with pleomorphic and hyperchromatic nuclei. These are surrounded by normal red blood cells.

clinical context, a differential is an extraosseous plasmacytoma.

Multiple myeloma, a neoplastic plasma cell disorder, accounts for 13% of haematological malignancies.¹ The median age of diagnosis is 69 years and two-thirds of patients are male.² Positron emission tomography (not performed in our patient) is sensitive to extramedullary myeloma, a systematic review of nine studies found rates of ranging from 2%–27%.³ Prevalence increases with duration of disease, age and number of previous therapies. The skin and subcutaneous soft tissue are the most common extramedullary foci, accounting for 69%,⁴ followed by liver, kidneys and central nervous system.⁵ Extramedullary disease is associated with multiple poor prognostic factors, and overall survival for an extramedullary relapse is <6 months.⁶

Extramedullary involvement of the pancreas is a rare entity, seen almost exclusively in the context of established multiple myeloma.⁷ This case is unique in demonstrating macroscopic venous invasion and this feature should not exclude haematological malignancy from the diagnosis.

Mathew James Keegan ¹, **Joshua Bell**,² **Alice Westwood**,³ **Alison Cairns**,³ **Raneem Albazaz**,² **Matthew T Huggett**¹

¹Department of Gastroenterology, Leeds Teaching Hospitals NHS Trust, Leeds, UK

²Department of Radiology, Leeds Teaching Hospitals NHS Trust, Leeds, UK

³Department of Histopathology, Leeds Teaching Hospitals NHS Trust, Leeds, UK

Correspondence to Dr Mathew James Keegan, Department of Gastroenterology, Leeds Teaching Hospitals NHS Trust, Leeds LS9 7TF, UK; mathewjkeegan@gmail.com

Contributors MJK and JB were involved in the literature review and drafted the manuscript layout, text content and sourcing images. MJK, JB, RA and MTH were involved in revisions of the manuscript layout and content. AW and AC advised on histopathology input and provided the demonstrative histopathology image.

Funding This research has received no specific grant from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

© Author(s) (or their employer(s)) 2020. No commercial re-use. See rights and permissions. Published by BMJ.

To cite Keegan MJ, Bell J, Westwood A, et al. *Frontline Gastroenterology* 2020;**11**:335–336.

Received 6 December 2018

Revised 2 May 2019

Accepted 3 May 2019

Published Online First 5 July 2019

Frontline Gastroenterology 2020;**11**:335–336. doi:10.1136/flgastro-2018-101163

ORCID iD

Mathew James Keegan <http://orcid.org/0000-0002-5026-6267>

REFERENCES

- 1 Palumbo A, Anderson K, Myeloma M. Multiple myeloma. *N Engl J Med* 2011;364:1046–60.
- 2 Röllig C, Knop S, Bornhäuser M. Multiple myeloma. *The Lancet* 2015;385:2197–208.
- 3 van Lammeren-Venema D, Regelink JC, Riphagen II, et al. ¹⁸F-fluoro-deoxyglucose positron emission tomography in assessment of myeloma-related bone disease: a systematic review. *Cancer* 2012;118:1971–81.
- 4 Pour L, Sevcikova S, Greslikova H, et al. Soft-tissue extramedullary multiple myeloma prognosis is significantly worse in comparison to bone-related extramedullary relapse. *Haematologica* 2014;99:360–4.
- 5 Bladé J, de Larrea CF, Rosiñol L. Extramedullary involvement in multiple myeloma. *Haematologica* 2012;97:1618–9.
- 6 Touzeau C, Moreau P. How I treat extramedullary myeloma. *Blood* 2016;127:971–6.
- 7 Short KD, Rajkumar SV, Larson D, et al. Incidence of extramedullary disease in patients with multiple myeloma in the era of novel therapy, and the activity of pomalidomide on extramedullary myeloma. *Leukemia* 2011;25:906–8.