A Not-So-Daughter Tumour: An Atypical Presentation of a SPN Tumour

Dr James Mooney, Mr Gareth Kirk
Belfast City Hospital, Belfast Health and Social Care Trust

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Introduction

A Solid Pseudopapillary Neoplasm (SPN) tumour is a pancreatic malignancy that represents up to 2.7% of all exocrine pancreatic tumours. These tumours show a strong 10:1 female predilection with median age of diagnosis of 24.5 years (1); hence the coined term "Daughter tumour". SPN tumours have an excellent prognosis and complete resection is considered to be curative. These tumours can be asymptomatic or alternatively present with an epigastric mass, epigastric tenderness, nausea and rarely will these tumours present with metastatic disease. Their histogenesis is theorised to be of pancreatic pluripotent cell origin (2).

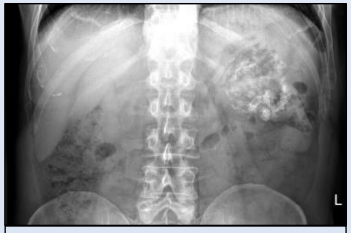

Image 1: Abdominal X-ray film demonstrating a right upper quadrant calcified mass

Description

A 58-year male and presents with an incidental finding of a calcified mass on DEXA scanning. This patient is asymptomatic and an otherwise fit and well competitive cyclist. The DEXA scan was prompted after a low impact wrist fracture. The calcificied mass was confirmed on AXR (Image 1). Tumour markers (Including CA19-9) alongside other routine bloods tests were normal.

CT imaging proposed a 9x8x8cm pancreatic malignancy associated with gastric invasion; extensive lymphadenopathy alongside concerning pulmonary nodules and hepatic cysts (Images 2&3. Prudent radiological reporting stated an SPN tumour as a possible differential diagnosis.

EUS biopsy supported the SPN diagnosis. Surgery involved an open distal pancreatectomy and splenectomy. Resectional histopathology confirmed the SPN diagnosis alongside a new diagnosis of Chronic Lymphocytic Leukaemia (CLL). 6-month follow up CT imaging demonstrated no evidence of SPN recurrence. The patient remains under active surveillance for CLL Progression.

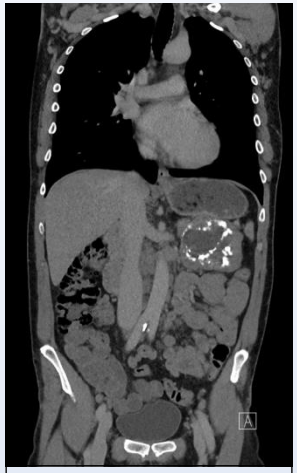

Image 2: Coronal CT demonstrating a intrapancreatic right upper quadrant calcified mass

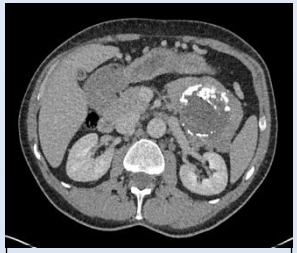

Image 3: Transverse CT demonstrating a heterogenous intra pancreatic calcified mass

Discussion

This patient was incidentally diagnosed with CLL and a rare SPN tumour in an atypical demographic. In any atypical presentation of a disease with a dual pathology we must consider the possibility of a causal link.

This case also emphasises the importance of histopathological analysis for diagnosis. This patient's index CT scan could have been interpreted as a T3N3M1 pancreatic malignancy (Stage IV) with a poor prognosis and few surgical options. With a confirmed SPN diagnosis the option of a potentially curative surgical resection became a possibility. By confirming the SPN diagnosis the patients expected 5-year survival increased from 6.5% to 98%. (3)(4)

When the clinical presentation does not correlate with the radiological findings; we must reconsider our initial diagnosis and explore the possibility of an alternative diagnosis or dual pathology to explain our observations.

Important points

- The radiological awareness, identification and delineation of SPN tumours is improving.
- When the clinical picture does not fit the radiological or biochemical findings; we should re-examine our initial diagnosis. Is there an alternative diagnosis that better fits the picture?
- In an atypical patient with 2 rare pathologies consider that there may be a causal link.
- This is a case where the Occam's razor approach does not prove useful. Alternatively, Hickam's Dictum appears to be a more appropriate adage to adopt:

  "A man can have as many diseases as he damn well pleases." - John Hickam, MD

References

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