International Journal of Surgical Procedures

ISSN 2517-7354

Solid Pseudopapillary Neoplasm of the Pancreas Mimicking an Adrenal Incidentaloma

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Abstract

Objective:

Solid Pseudopapillary Neoplasms (SPN) of the pancreas are a rare malignancy, accounting for less than 3% of tumours of the exocrine pancreas. They are characteristically identified in a particular demographic population, as demonstrated by this case patient.

Method and results:

A 29 year old pregnant female presented with right sided weakness. Ultrasound scans performed at this time revealed right sided hydronephrosis, in addition to, a left supra-renal mass. This was further evaluated with contrast enhanced computed tomography and magnetic resonance imaging. The pre-operative diagnosis was a non-functioning adrenal incidentaloma and laparoscopic left adrenalectomy was scheduled. A large mass arising from the tail of the pancreas was identified at surgery and a laparoscopic distal pancreatectomy and splenectomy was performed. Subsequent histopathology revealed a solid pseudopapillary tumour, a rare find, for this unusual presentation of a pancreatic incidentaloma.

Conclusion:

The slow growth and seemingly benign nature of this neoplasm leads to most cases being identified during routine diagnostic imaging- but as was depicted in this case, the pancreatic neoplasm was masked as an adrenal incidentaloma despite several imaging modalities.

Keywords

Hiatal hernia; Cardiopulmonary compression; Life threatening

Introduction

Solid pseudopapillary tumours, classically known as the Frantz tumour, are an uncommon occurrence in the world of pancreatic malignancies. This tumour has a typical inclination, as it pertains to the patients affected. Our case history depicts all of the classic features, a young, black female – within the age group of 20 – 40 years, and no evidence of metastatic spread [1–3]. However, despite several imaging modalities, the revelation of this pancreatic tumour was only noted during the intra-operative period. A Frantz tumour mimicking an adrenal mass

Case History

A twenty nine (29) year old female, with no known medical conditions, presents with right sided abdominal pain, with associated weakness of the right lower limb at thirty six (36) weeks gestation. Ultrasound at that time revealed severe right hyrdronephrosis, not uncommon in the third trimester, and also a heterogeneous mass adjacent to the left kidney. Several months post-partum, contrast enhanced computed tomography revealed a mixed density supra-renal mass measuring 8 cm x 8.2 cm x 6.2 cm (Figure 1). Magnetic resonance imaging confirmed the location of this mass, and excluded the presence of intrahepatic lesions (Figure 2).

The working diagnosis at this time was that of a left adrenal incidentaloma. Negative blood and urine laboratory investigations omitted the presence of a functional mass. Given the size of the mass, laparoscopic adrenalectomy was planned. During the procedure, a large solid mass was found arising from the tail of the pancreas (Figure 3). A distal pancreatectomy and splenectomy was performed and the specimen was removed in a retrieval bag through a Pfannenstiel Incision. An uneventful post-operative course followed and histology results of the distal pancreatic lesion demonstrated a pT2pN0 solid pseudo-papillary tumour, 8 cm x 7.5 cm x 7 cm in size (Figure 4).

Article Information

DOI: 10.31021/ijsp.20181116
Article Type: Case Report
Journal Type: Open Access
Volume: 1 **Issue:** 4
Manuscript ID: IJSP-1-116
Publisher: Boffin Access Limited

Received Date: 16 July 2018
Accepted Date: 27 August 2018
Published Date: 3 September 2018

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Citation: Bascombe N, Calderon C, Ann Bobb K, Greaves W, Dan D. Solid Pseudopapillary Neoplasm of the Pancreas Mimicking an Adrenal Incidentaloma. Int J Surg Proced.2018 Aug;1(4):116

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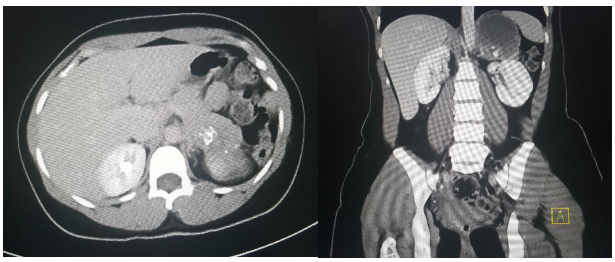

Figure 1: Axial and coronal views of CT scan images of left supra-renal mass

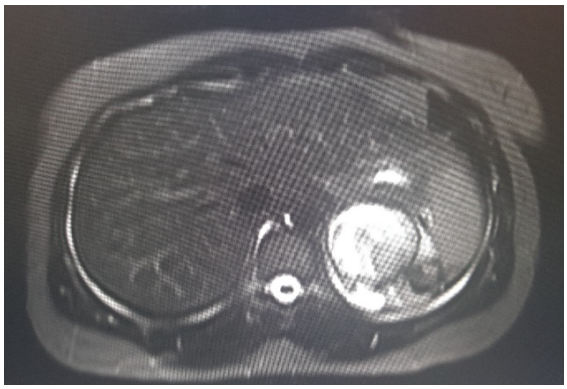

Figure 2: Axial view of MRI scan depicting left supra – renal mass and absence of intra-hepatic lesions

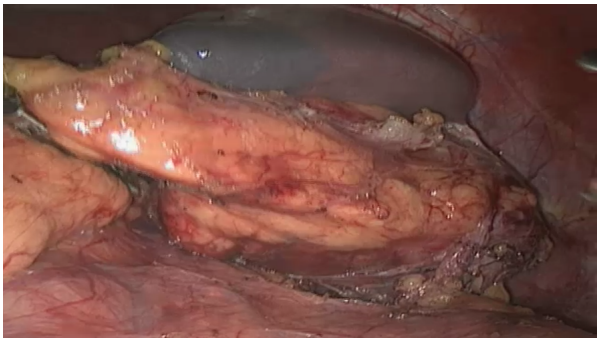

Figure 3: Laparoscopic Image of mass arising from tail of pancreas with attached spleen

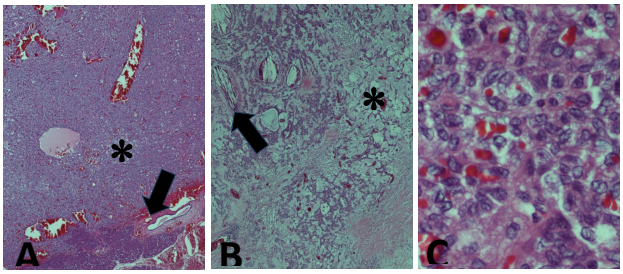

Figure 4: Solid-Pseudopapillary Neoplasm (H&E). A. Low magnification (20x) highlights the predominantly solid sheet-like growth pattern (∗) of this tumour as well as a relatively well-demarcated interface with the adjacent non-neoplastic pancreas (→). B. A typical finding is the presence of foamy macrophages with clear cytoplasm (∗) and cholesterol clefts (→). C. Higher magnification (400 x) highlights the tumour cells showing relatively uniform nuclei with nuclear grooves and eosinophilic cytoplasm.

Discussion

The Frantz tumour, eponymous with Virginia Frantz in 1959, accounts for a very small percentage of tumours affecting the exocrine pancreas (1-2%) [2,4]. There is a clear predilection in the female population, as striking as ninety percent (90%) ; and usually manifests during the second to fourth decades of life , although outliers have been highlighted [5]. Fortunately these neoplasms are rarely aggressive in nature, and usually take on an indolent course [6].

These tumours have no characteristic clinical manifestations, and in fact, most times symptoms are often obscure, non-specific and even misleading. As such, most clinicians place heaving weighting on imaging modalities in making a diagnosis [2,5,7]. In this current era of readily available ultrasounds and computed tomography imaging, together with a low threshold for utilising these tools, it may be stated that it is easier to make a verdict of this pancreatic mass. This is shown true, with the increase incidence of Frantz tumour diagnosed in the last decades [8].Our case scenario demonstrated that despite numerous imaging tools, it was not enough to guide us in the right direction.

Computed tomography of the abdomen is the ideal tool for visualisation of the solid pseudopapillary neoplasm, which usually presents in the tail of the pancreas. It reveals a mixed consistency mass, with both solid and cystic components. Notable calcifications, necrosis and areas of haemorrhage may also be identifiable [9,10]. This mass could become considerable in size and as a result, may have ability to compress and displace surrounding structures [11,12]. We believe that this case of mistaken identification was due to the displacement of the left adrenal gland by a four hundred and twenty cubic (420 cm^3) volume pancreatic tail neoplasm.

The incidental finding of a Frantz tumour via the laparoscopic approach is extremely uncommon [2]. The laparoscopic approach employed in this case, was both diagnostic and therapeutic, with the mainstay for this low grade neoplasm being operative resection. It is important to keep the equilibrium between clear margins and the preservation of disease-free pancreas, as best as possible [8,9,13]. This approach may be altered, and require a more aggressive route, in the setting of more advanced disease process being encountered, seen in approximately 15% of patients[14,15].

Along with the distinguishing age group and sex predilection, these neoplasms have a relatively predictable pattern of behaviour post operation. They depict low recurrence rates in the setting of a R0 resection and the likelihood of metachronous metastasis is uncommon. Hence, patients managed appropriately have an excellent prognosis [16,17].

Consent

Patient approval was taken.

Conflicts of interest

None

Conclusion

An unexpected laparoscopic finding of a solid pseudo-papillary neoplasm is very uncommon. Imaging modalities and an element of clinical suspicion are usually sufficient to make the definitive diagnosis pre-operatively. Fortunately, once managed operatively with clear resection margins, these patients have an excellent outcome.

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