Case Report

DOI: 10.5455/2349-2902.isj20150223

Occult insulinoma in a Meckel's diverticulum: a case report

Bharati Pandya*, Ravinder Narang, Ulhas N. Jajoo

Department of Surgery, MGIMS, Sevagram, Wardha-442102, M.S., India

Received: 14 December 2014 Accepted: 30 December 2014

*Correspondence: Dr. Bharati Pandya,

E-mail: Bharati_pandya@rediffmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Insulinoma is a deceptive endocrine tumour of the pancreas due to its bizarre and nonspecific symptom complex. It is also difficult to localize preoperatively, especially when present in an ectopic location. A 65 year old woman presented with recurrent episodes of hypoglycaemia. She was erroneously under treatment for psychotic disorder. A suspicion of insulinoma led us to investigate her further. Investigations revealed fasting hypoglycaemia, endogenous hyperinsulinism, and a pancreatic parenchymal lesion. On exploration, an incidental Meckel's diverticulum with a nodule was also found. Histopathology showed an insulinoma of the Meckel's diverticulum and a benign nonfunctioning adenoma in the pancreatic nodule. Surgery eventually resulted in restoration of euglycaemia and complete disappearance of patient's symptoms.

Keywords: Occult insulinoma, Heterotopic pancreas, Neuroglycopenia, Meckel's diverticulum

INTRODUCTION

Heterotopic or ectopic pancreas is pancreatic tissue outside boundaries of pancreas without anatomic or vascular connections to pancreas. It has a reported frequency of 0 to 13.7%. It is usually found in the stomach, duodenum and upper part of jejunum and less commonly in the ileum, biliary system and spleen. Its presence in Meckel's diverticulum is well documented, but rare. Neoplasms arising in Meckel's diverticulae are uncommon and those reported in the literature are mainly carcinoid tumours, gastrointestinal stromal tumours, and gastric, intestinal or rarer still pancreatic adenocarcinomas. To the best of our knowledge, pancreatic insulinoma in a Meckel's diverticulum has not been reported before. An insulinoma is a neuroendocrine tumor, deriving mainly from pancreatic islet cells. It is usually solitary, sporadic and less than 2 cm in dimension.² It secretes insulin. Only 2-3% are extrapancreatic in origin.² Insulinomas are difficult to diagnose and patients often get misdiagnosed with psychiatric illnesses or seizure disorders. Localizing these tumours is equally difficult. Occult insulinomas are biochemically proven tumours with indiscriminate anatomical site before operation. We describe a patient who was found to have such a tumour. The clinical and pathological aspect along with the related literature is reviewed.

CASE REPORT

A 65-year-old woman presented with a 15 day history of decreased oral intake and associated weakness. She was on drugs for psychotic depression for the past 12 years and had documented episodes of hypoglycaemia on her previous admissions. Physical examination including a thorough nervous system examination was unremarkable.

Investigations showed normal hemogram, kidney and liver functions. During her hospital stay, she had frequent episodes of early morning hypoglycaemia with associated hunger binges, mental disturbances and violent

behaviour. Her blood sugar levels would drop to as low as 37 mg/dl. Insulinoma was suspected and her Insulin and C-peptide levels confirmed it. Abdominal ultrasound did not show any pancreatic lesion. CECT imaging revealed a 2 cm. lesion in the head of pancreas, with a distinct tumour blush, but no evidence of metastasis or lesions elsewhere.

Patient was accordingly prepared for laparotomy. Pancreatic adenoma was palpable and was successfully enucleated. On random inspection of the bowel, an incidental Meckel's diverticulum was found. CT scan done preoperatively had failed to pick this up. It was 2 cm. in size with a narrow neck and 55cm, from the ileocaecal valve and had a 1.5 cm. nodule at the tip (Fig 2). No other nodules, lymph nodes or evidence of malignancy could be found on thorough exploration. In view of a long-standing history, small size of the tumour, absence of lymph nodes and narrow neck of the Meckel's diverticulum, wedge resection with adequate margin was performed. Both the specimens were sent for histopathology. Histopathology reported enucleated pancreatic lesion to have multiple, mucinous, cystic lesions with no evidence of neuroendocrine tumour. The typical insulinoma was seen arising denovo from ectopic pancreatic tissue present in the Meckel's diverticulum. There was no evidence of malignancy and resection margins were clear. Postoperatively insulin drip was required for a week. Her glucose levels stabilized subsequently and full oral diet was resumed. She was later discharged, euglycaemic and off anti-psychotic therapy. On follow-up the patient is doing well for over two years now.



Figure 1: Pancreatic adenoma being enucleated.



Figure 2: Meckel's diverticulum with the insulinoma.

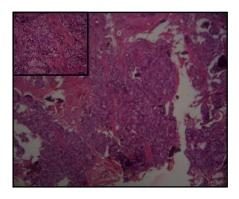


Figure 3: Histology: insulinoma (inset) in Meckel's diverticulum.

DISCUSSION

Hormone secreting pancreatic tumours with clinical repercussions are rarely seen. Their occurrence has been estimated to be 1-4 per million, per year. Insulinomas are hormone secreting pancreatic tumours which present clinically with symptoms of episodic hypoglycaemia. They are usually solitary and less than 2 cm in size.² Only 5-11% of these tumours are malignant.² They have a slight female preponderance (1.4:1 - female:male ratio) and median age at diagnosis is about 47 years.² These tumours pose a diagnostic challenge owing to their dramatic, at times, puzzling presentations. Diagnosis may delayed because symptoms are nonspecific. Neuroglycopenic symptoms are chiefly seen in these patients and they are misdiagnosed as epilepsy or neuropsychiatric disorder in as high as 20% cases. 3,6 Our patient was 65 years old and was shuffling between physicians and psychiatrists for the past 12 years. A strong index of suspicion and relevant investigations led us to the diagnosis. In patients with insulinoma, other than behavioral changes, diplopia, sweating and palpitations are seen at a rate of 85%; confusion or abnormal behavior in 80%, unconsciousness and amnesia is seen in 53% and epilepsy in 12%.6

Insulinomas were the first pancreatic neuroendocrine tumors to be identified, as reported by Whipple & Frantz. P7-98% Insulinomas are intrapancreatic and only 2-3% of these tumours are in ectopic locations. The first resection of an insulinoma was performed in 1927, when W. J. Mayo removed an insulin-secreting tumor and injected its extracts into rabbits, they subsequently developed hypoglycemia. A classic history of hyperinsulinaemic, hypoglycaemic syndrome, which responds to administration of glucose in a clinical setting, suggests the diagnosis of an insulinoma (Whipple's triad). Insulinoma is diagnosed biochemically with low blood glucose; elevated insulin, C-peptide and proinsulin levels.

Meckel's diverticulum is the most prevalent (2%) congenital abnormality of the gut, but is often difficult to diagnose. Overall tumors that arise in Meckel's

diverticulum are rare and occur in only 0.5% to 1.9% of cases. 4 Majority of them are carcinoids (33%), followed by gastrointestinal stromal tumours, benign leiomyomas and less commonly gastric, intestinal and benign or malignant pancreatic exocrine tumours (3.2%). Ectopic tissue, found in 50 percent of them, consists of gastric tissue in 60 to 85 percent and pancreatic tissue in 5 to 16 percent cases. Gastric mucosa is more often found at the base while the pancreatic tissue is more at the apex or tip of the Meckel's diverticulum. Meckel's diverticulum is typically short and wide mouthed, which makes its detection difficult. The success rates of investigations as reported by Groebli et al are, small bowel enema 36%, 99mTc scan 75%, angiography 33%, and computed tomography only 7%. 11 Overall tumors that arise in Meckel's diverticulum are rare and occur in only 0.5% to 1.9% of cases.⁴ The majority of them are carcinoids (33%), followed by gastrointestinal stromal tumours, benign leiomyomas and less commonly gastric, intestinal and benign or malignant pancreatic exocrine tumours (3.2%). In our patient even on retrospect, Meckel's diverticulum could not be detected on CT scan and the Meckel's diverticulum had a narrow neck with a 1.5 cm nodule confined to the tip of the diverticulum. There were no evident lymphnodes or metastatic lesions and wedge resection was giving adequate margins. Lopez et al. 12 observed that malignant tumours are characterized by a shorter history, age over 50 years, weight loss (as opposed to weight gain seen in benign), more severe hypoglycaemia, tumour size more than 2 cm, its multiplicity and presence of lymphnodes or liver secondaries. These characteristics aid in clinical assessment of insulinomas.

Localization of insulinomas is difficult owing to its small dimension and occasional ectopic location.^{2,12} Inability to preoperatively localize a biochemically proven tumour led clinicians to coin the term 'occult insulinoma'. Localization usually is done by computed tomography, MRI, or transabdominal and endoscopic ultrasonography. Best localization has been reported with somatostatin receptor scintigraphy,2 which helps even when the tumours are in ectopic locations. Machado et al. have observed that, notwithstanding the progress of diagnostic methods during the latest years, the identification of insulinomas remains a difficult clinical problem both for clinicians and surgeons. According to Norton et al. intraoperative ultrasound (IOUS) for the treatment of benign insulinomas is the best modality for precise operative localization. The definitive treatment is surgical removal of the tumor, which gives 90% success.⁶ Laparoscopic surgery for localized lesions is increasingly being reported. Medical treatment is reserved only for unresectable tumors, preoperative preparation or for unsuitable candidates for surgery.

CONCLUSION

A clinical setting of Whipple's triad is strongly suggestive of an insulinoma. Nonetheless, a very high

index of suspicion is required because the presentation of neuroglycopenic symptoms can be elusive. An even greater challenge is in localizing these rare pancreatic endocrine tumours especially when in heterotopic location. Here we have presented an occult insulinoma in a very rare location, the Meckel's diverticulum. In this case a lesion diagnosed on CT, erroneously led us to believe that it was a pancreatic adenoma we were dealing with. Intraoperatively we detected another suspicious lesion which later on was proved to be a benign occult insulinoma arising denovo in a Meckel's diverticulum. This experience suggests that a meticulous laparotomy despite all available diagnostic modalities, should not be skipped.

Funding: No funding sources Conflict of interest: None declared Ethical approval: Not required

REFERENCES

- Allison JW, Johnson JF III, Barr LL, Warner BW, Stevenson RJ. Induction of gastroduodenal prolapse by antral heterotopic pancreas. Pediatr Radiol. 1995;25:50-1.
- Bassam Abboud, Joe Boujaoude. Occult sporadic insulinoma: Localization and surgical strategy. J World Gastroenterol. 2008 Feb;14(5):657-65.
- 3. Service FJ, McMahom MM. Functioning insülinoma-incidence, recurrence and long term survival of patient: a 60 year study. Mayo Clin Proc. 1991;66(7):771-9.
- Justin MM. Cates, Timothy L. Williams, Arief A. Suriawinata. Intraductal papillary mucinous adenoma that arises from pancreatic heterotopia within a Meckel's diverticulum. Arch Pathol Lab Med. 2005 Mar;129(3):e67-9.
- 5. Hoey C. Koh, Blaithin Page, Catherine Black, Ian Brown, Stuart Ballantyne, David J. Galloway. Ectopic pancreatic-type malignancy presenting in a Meckel's diverticulum: a case report and review of literature. World J Surg Oncol. 2009;7:54.
- Mesut Ozkaya, Mehmet F. Yuzbasioglu, Irfan Koruk, Erman Cakal, Mustafa Sahin, Basak Cakal. Preoperative detection of insulinomas: two case reports. Cases J. 2008;1:362.
- 7. Ana Cláudia Ferreira Rosa, Márcio Martins Machado, Marcella Stival Lemes, Mariana Caetano Barreto, Rodrigo Alvarenga Nunes, Nestor de Barros, et al. Intraoperative ultrasonographic evaluation of insulinomas: an update. Radiol Bras. 2006. Sep/Oct;39(5):361-5.
- 8. Whipple AO, Franz VK. Adenoma of islet cells with hyperinsulinism. Am Surg. 1935;101:1299-335.
- 9. John P. Martin, Pamela D. Connor, Kerri Charles, JD, University of Tennessee. Meckel's diverticulum. Am Fam Physician. 2000 Feb;61(4):1037-42.

- 10. John J. Park, Bruce G. Wolff, Matthew K. Tollefson, Erin E. Walsh, Dirk R. Larson. Meckel's diverticulum. Ann Surg. 2005 Mar;241(3):529-33.
- 11. Groebli Y, Bertin D, Morel P. Meckel's diverticulum in adults: retrospective analysis of 119 cases and historical review. Eur J Surg. 2001 Jul;167(7):518-24.
- 12. López Alvarenga JC, Cáceres Agreda N, Rivera L, Gamboa A, Gómez Pérez FJ, Rull JA. Diagnosis of

malignant and benign insulinoma. Experience of the National Institute of Nutrition. Rev Invest Clin. 1999 May-Jun;51(3):167-73.

DOI: 10.5455/2349-2902.isj20150223

Cite this article as: Pandya B, Narang R, Jajoo UN. Occult insulinoma in a Meckel's diverticulum: a case

report. Int Surg J 2015;2:105-8.