

Correspondence

Comment on: Insulinoma in tuberous sclerosis: an entity not to be missed

To the Editor

I read, with interest, the article by Al Qahtani et al,¹ in which they presented an extremely rare association between tuberous sclerosis which is an autosomal dominant disease with multi-system disorders, and insulinoma which is a very rare but functioning tumor of the pancreas. The case was well-presented with a sufficient review of the relevant literature. Cases with multi-system involvements usually imply integrative management between the different medical and surgical specialties. So, the current case represents a successful and practical model of the multidisciplinary approach to disease management.

Also, it is a very obvious example of the incidental discovery of a mass during follow-up that allowed timely intervention. This was based on employing sensitive imaging technologies including combined scintigraphic body scanning using computed tomography and single photon emission computed tomography (SPECT/CT) techniques. This may confirm the important role of these new techniques in localization and characterization of the lesion and its proper management with a specific role of the intraoperative ultrasonography.^{1,2} Previously, however, the sensitivity of imaging techniques was doubtful and the outcomes were disappointing.³

Besides the very rare disease associations, this case was unique in that the patient had an interesting surgical history including laparoscopic total proctocolectomy for familial adenomatous polyposis, renal angioembolization for angiomyolipoma, and other endoscopic interventions.¹ No doubt that these interventions added more risks to the original disease burden and complexed its course and management. The sequences of management of this patient provided an obvious example for the fact that early diagnosis enhances the proper and timely diagnostic and therapeutic interventions that could maximize the disease outcomes and patient's survival.

Classically, the clinical manifestations of hypoglycemia and laboratory workups including blood glucose levels are fundamental to the diagnosis of insulinomas.^{2,3} However, the laboratory profile of the

current patient was not mentioned and it would be better to present it relative to the chronological events of diagnosis and interventions. This issue may provide a reason that the hypoglycemic status was not diagnosed until the lesion was incidentally seen by imaging. Also, it would be better if the non-surgical and minimally-invasive approaches of management of insulinoma were addressed more comprehensively with a specific reference to its applicability to the current case which had a considerable history of surgical interventions. This may help in revolutionizing the surgical role with the advancement of surgical techniques in the management of these lesions.²

To overcome unfavorable practical scenarios in the management of these very rare cases, the underlying pathogenesis and disease behavior or natural course should be understood comprehensively. From a practical perspective, this rare association between tuberous sclerosis and insulinoma should be considered in those patients, especially when they present with unexplained manifestations such as hypoglycemia or deteriorating neurocognitive symptoms.^{1,4}

Rabea A. Gadelkareem

Assiut Urology and Nephrology Hospital

Faculty of Medicine

Assiut University, Egypt

Saudi Med J 2021; Vol. 42 (5): 578

doi: 10.15537/smj.2021.42.5.578

Reply from the Author

No reply was received from the Author.

References

1. Al Qahtani MS, Bojal SA, Alqarzaie AA, Alqahtani AA. Insulinoma in tuberous sclerosis: an entity not to be missed. *Saudi Med J* 2021; 42: 332-337.
2. Zaver HB, Alnahhal KI, Ghaz H, Paz-Fumagalli R, Raimondo M, Lukens FJ. Insulinoma: a recurrent pancreatic tumor amenable to computed tomography-guided ethanol-lipiodol injection. *ACG Case Rep J* 2021; 8: e00539.
3. Shin JJ, Gorden P, Libutti SK. Insulinoma: pathophysiology, localization and management. *Future Oncol* 2010; 6: 229-237.
4. Kang MY, Yeoh J, Pondicherry A, Rahman H, Dissanayake A. Insulinoma and tuberous sclerosis: a possible mechanistic target of rapamycin (mTOR) pathway abnormality? *J Endocr Soc* 2017; 1: 1120-1123.