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Doege-Potter Syndrome: A Rare Case of Metastatic Hemangiopericytoma with Persistent Hypoglycemia in a 27-year-old Male

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Zarra VLD, Demegillo KJN, Naranjo M
Southern Philippines Medical Center

INTRODUCTION

Doege-Potter syndrome (DPS) is a rare paraneoplastic condition characterized by hypo insulinemic hypoglycemia secondary to a solitary fibrous tumor. The underlying mechanism is secretion of pro-insulin-like growth factor (IGF) II by the tumor which causes decreased release of glucose into the circulation resulting to hypoglycemia. Only forty-five (45) cases of DPS have been reported since 1979 worldwide.

CASE

This is a case of a 27-year-old Filipino male who had multiple admissions due to the recurrence of a right temporo-zygomatic mass. He underwent excision five times and eventually was subjected to radiotherapy. The patient was asymptomatic with no evidence of disease for two years until a recent consultation was sought due to hypoglycemia presenting as seizure and decrease in sensorium. Metastatic work-up revealed multiple metastases in the liver, the lungs and the right adrenal gland; hence, a suspicion of hypoglycemia as a paraneoplastic event was entertained. This was confirmed by a 72-hour fast protocol which ruled out Insulinoma and pointed towards a non-islet cell tumor as the culprit of hypoglycemia.

The patient was initially managed with continuous dextrose infusion, increase in caloric intake and steroids which ameliorated hypoglycemic episodes. He was eventually started on a weekly doxorubicin as a palliative treatment with an intent of lessening the tumor burden.

CONCLUSION

Due to its complexity, the management of DPS from a metastatic hemangiopericytoma was challenging and required a multidisciplinary approach; hence, an early screening for metastases was emphasized to prevent the undesirable sequelae of this disease process.

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Ectopic Parathyroid Adenoma, a Diagnostic Challenge

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Yen Nee L, Masni M, Azraai Bahari N, Nurain MN, Zanariah H
Division of Endocrinology, Department of Internal Medicine, Hospital Putrajaya, Malaysia

INTRODUCTION

Primary hyperparathyroidism is the most common cause of hypercalcemia. Approximately 85% of primary hyperparathyroidism is caused by solitary parathyroid adenomas and 5-10% of cases derived from ectopic adenomas. Ectopic parathyroid glands remain a diagnostic and operative challenge in terms of localizing the culprit gland.

CASE

We reported a case of primary hyperparathyroidism due to ectopic parathyroid adenoma in the thymus. This 31-year-old gentleman presented with renal calculi, confirmed primary hyperparathyroidism with hypercalcemia.

However initial ultrasound and CT neck were unable to localize the parathyroid adenoma. Technetium – 99 (Tc-99m) sestamibi scan repetitively showed focus in the anterior mediastinum. CT Thorax showed a nodule in the mediastinum, which coincides with the focus of increased uptake in the sestamibi scan. He subsequently underwent median sternotomy and thymectomy. Intra-operatively found left thymus with a nodule size less than 1 cm within. Histopathology confirmed an ectopic parathyroid adenoma within the left thymus. Post operatively his calcium level remains stable with minimal calcium supplement.

CONCLUSION

This case illustrates the challenges in localizing ectopic parathyroid adenoma, which lead to delay in the surgery.