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Case Report

Acromegalic features and hypoglycemia: Doege-Potter syndrome

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Abstract

Background: Doege-Potter syndrome (DPS) is a condition characterized by severe, symptomatic, and sustained hypoglycemia, refractory to medical treatment, due to non-insular tumor cells.

Case Presentation: We describe the case of a 73-year-old male with acromegalic features and two months of hypoglycemic episodes, in whom a giant mass was identified in the right hemithorax via radiography. Surgical resection of the tumor resolved hypoglycemia and improved the acromegalic traits.

Solitary fibrous tumors (SFTs) are rare neoplasms that predominantly affect individuals aged 50 to 60 years. Doege-Potter syndrome is a cause of hypoglycemia, present in 2-4% of SFT cases. Its manifestations arise from abnormal synthesis and secretion of insulin-like growth factor (IGF)-2 by the SFT. In rare instances, patients with DPS may exhibit signs of acromegaly, attributed to the stimulation of the IGF-1 receptor by IGF-2. The initial diagnostic approach to DPS includes imaging and hormonal studies, while the definitive diagnosis is made through histopathology.

Conclusions: We emphasize the importance of considering DPS in patients with thoracic tumors and persistent hypoglycemia, especially when accompanied by acromegalic features.

Keywords: Acromegaly, Hypoglycemia, IGF Type 1 receptor, Solitary Fibrous tumors, Case reports.

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Doege-Potter syndrome (DPS) is a clinical condition characterized by severe, symptomatic, and sustained hypoglycemia, refractory to medical treatment, in patients with solitary fibrous tumor (SFT), due to excessive production of Insulin-like Growth Factor (IGF) Type 2 (1, 2). SFT is a rare mesenchymal neoplasm, primarily located in the pleural cavity. Fewer than 5% of patients with this tumor develop DPS (1). In 1930, Karl Doege and Roy Potter described the first patient with a fibrous tumor located in the mediastinum who presented with episodes of hypoglycemia, leading to the eponym of the syndrome (2).

The case presented herein offers practical insights and emphasizes the consideration of DPS in the differential diagnosis of unexplained hypoglycemia, especially when incidental thoracic findings and distinctive phenotypic features, such as acromegalic traits, are present. Moreover, it underscores how early diagnosis, and timely surgical management can prevent severe complications and optimize clinical outcomes. We present the case of an elderly male patient with an acromegalic phenotype who developed DPS, highlighting the association between acromegalic features and hypoglycemia.

Clinical data for this case were obtained through a review of the patient's medical records, imaging studies, and laboratory results. A direct interview was also conducted with the patient to clarify clinical history and symptoms.

Case Presentation

A 73-year-old male with a history of treated, pansensitive pulmonary tuberculosis 35 years ago and no significant family history, was evaluated preoperatively for an inguinal hernia repair at his local hospital. All identifying information was removed to ensure patient confidentiality. This study was conducted in accordance with the Declaration of Helsinki (1964) and its subsequent amendments. Written informed consent was obtained from the patient for participation in the study, as well as for the publication of this case report and any accompanying images or text.

The patient was given the opportunity to review the manuscript. The study received ethical approval from the Institutional Ethics Committee for Research of the Faculty of Medicine, Universidad Nacional de Trujillo, Peru (Approval Number: Of. N° 894-2024-UNT-FM-C.E.). During this evaluation, his blood glucose level was found to be 55 mg/dL, and he presented with diaphoresis, headache, and tremors. An incidental pulmonary mass was identified on the chest x-ray (figure 1), prompting referral to the thoracic surgery service of our national hospital for further management.

The patient reported a two-year history of progressive cough and dyspnea, as well as episodes of weakness and sweating occurring during the day or at night, that improved with food intake. These symptoms had been present for the past two months. On physical examination, the following findings were noted: weight of 81 kg, height of 1.70 m, and a body mass index (BMI) of 28 kg/m². Phenotypic characteristics such as prognathism, frontal prominence, rhinophyma, prominent nasolabial folds, frontal protrusion (figure 2), pachyderma, and increased volume of the hands and feet were observed. No other significant abnormalities were noted.

During hospitalization in the thoracic surgery department, the patient experienced multiple episodes of blood glucose levels below 50 mg/dL, requiring continuous infusion of 10% dextrose at a rate of 60 mL/hour. To determine the etiology of the hypoglycemia, hormonal studies were conducted during hypoglycemic episodes, revealing baseline low levels of insulin and C-peptide. Renal and hepatic functions were preserved, thyroid and adrenal axes were intact, and IGF-1 levels were normal (table 1).

Measurement of IGF-2 was not possible due to unavailability of the assay in our facility. Based on these findings, the primary diagnostic suspicion was a non-islet cell tumor producing IGF-2. A contrast-enhanced chest computed tomography (CT) scan was performed, revealing a heterogeneous solid mass occupying two-thirds of the right hemithorax (figure 3).

Table 1. Biochemical and hormonal tests requested during hospitalization.

Blood test	Result	Normal range
Glucose (mg/dL)	50	74 - 106
HbA1c (%)	4.9	< 5.7
Hemoglobin (g/dL)	14.2	14 - 16.5
Albumin (g/dL)	4.22	3.2 - 4.8
Creatinine (mg/dL)	0.46	0.6 - 1.1
ACTH (pg/mL)	19.2	0 - 46
Cortisol (ug/dL)	7.3	3.7 - 19.4
DHEAS (ug/dL)	49.2	228 - 283
IGF-1 (ng/mL)	52.1	24 - 200
TSH (uUI/mL)	1.6	0.35 - 4.94
Free T4 (ng/dL)	0.93	0.7 - 1.48
Basal insulin (uU/mL)	<1.6	2 - 28
C peptide (ng/mL)	0.57	0.78 - 5.19

ACTH: adrenocorticotropic hormone. DHEAS: dehydroepiandrosterone sulfate. HbA1c: glycated hemoglobin. IGF-1: insulin-like growth factor 1. Free T4: free thyroxine. TSH: thyroid stimulating hormone.

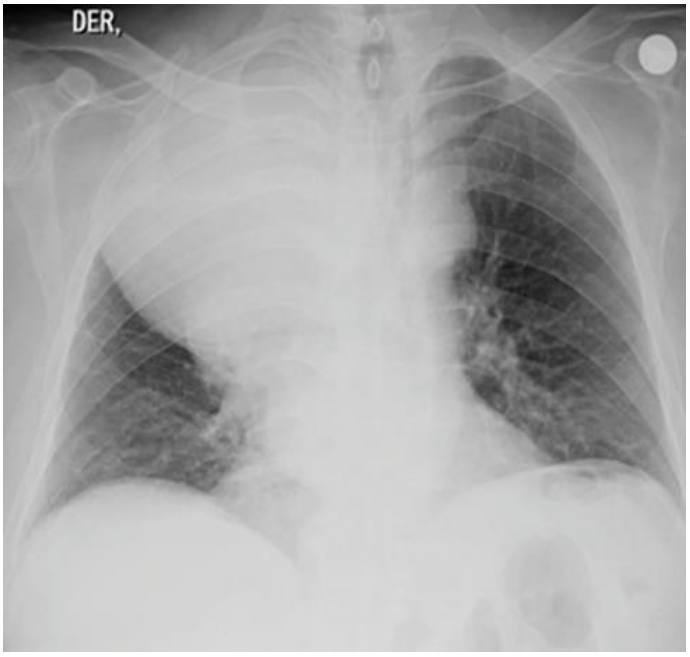

Figure 1. Chest x-ray in anteroposterior projection showing opacity in the upper half of the right hemithorax. With smooth borders. Displacing the contralateral mediastinum

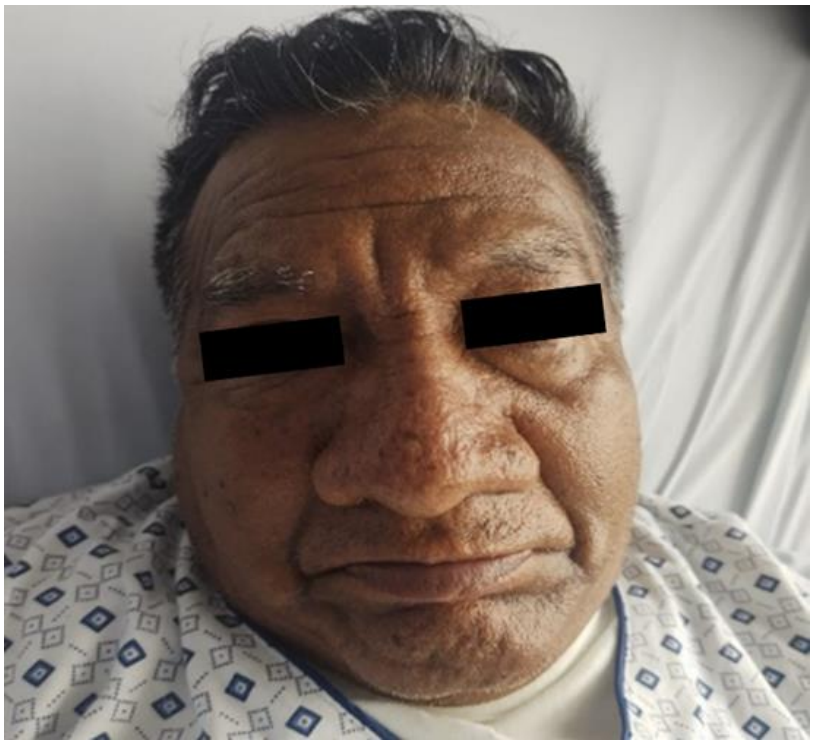

Figure 2. Acromegalic features of the patient

The patient underwent resection of a posterior mediastinal tumor via right posterolateral thoracotomy. Weighing 1500 g (figure 4). Pathological examination confirmed the diagnosis of a solitary fibrous tumor (SFT) with high metastatic potential. Immunohistochemical staining showed positivity for CD34 and a Ki67 index of 15%. With clear surgical margins (figure 4). The final diagnosis was Doege-Potter syndrome. Postoperatively. The patient did not experience any further hypoglycemic episodes and was discharged. He is currently under follow-up by the oncology and endocrinology departments. With no evidence of hypoglycemia.

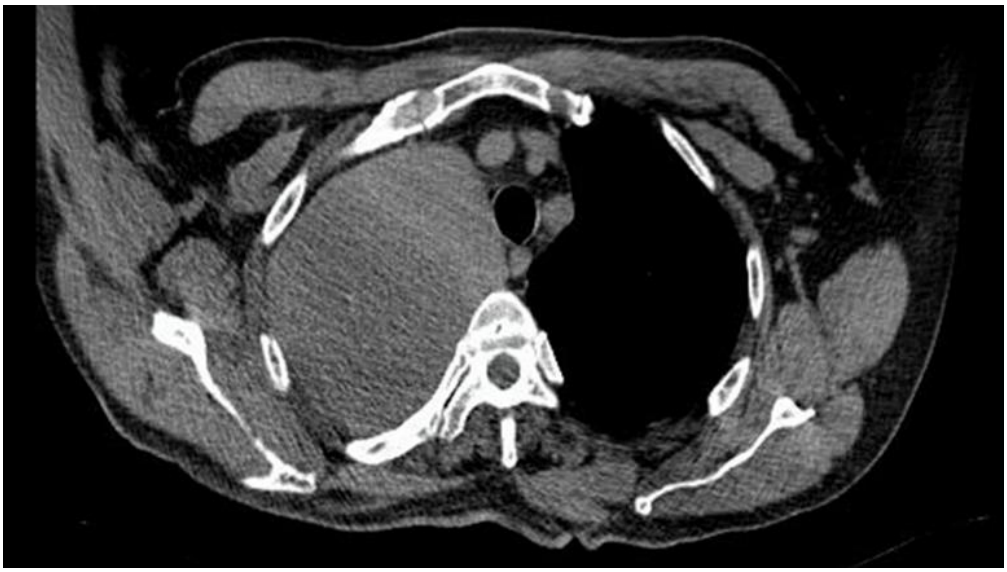

Figure 3. Contrast-enhanced chest CT (axial slice) showing a heterogeneous solid mass occupying the upper two-thirds of the right hemithorax. Causing atelectasis of the right upper lobe and the apical segment of the right lower lobe. The remainder of the examination is unremarkable. With no mediastinal lymphadenopathy.

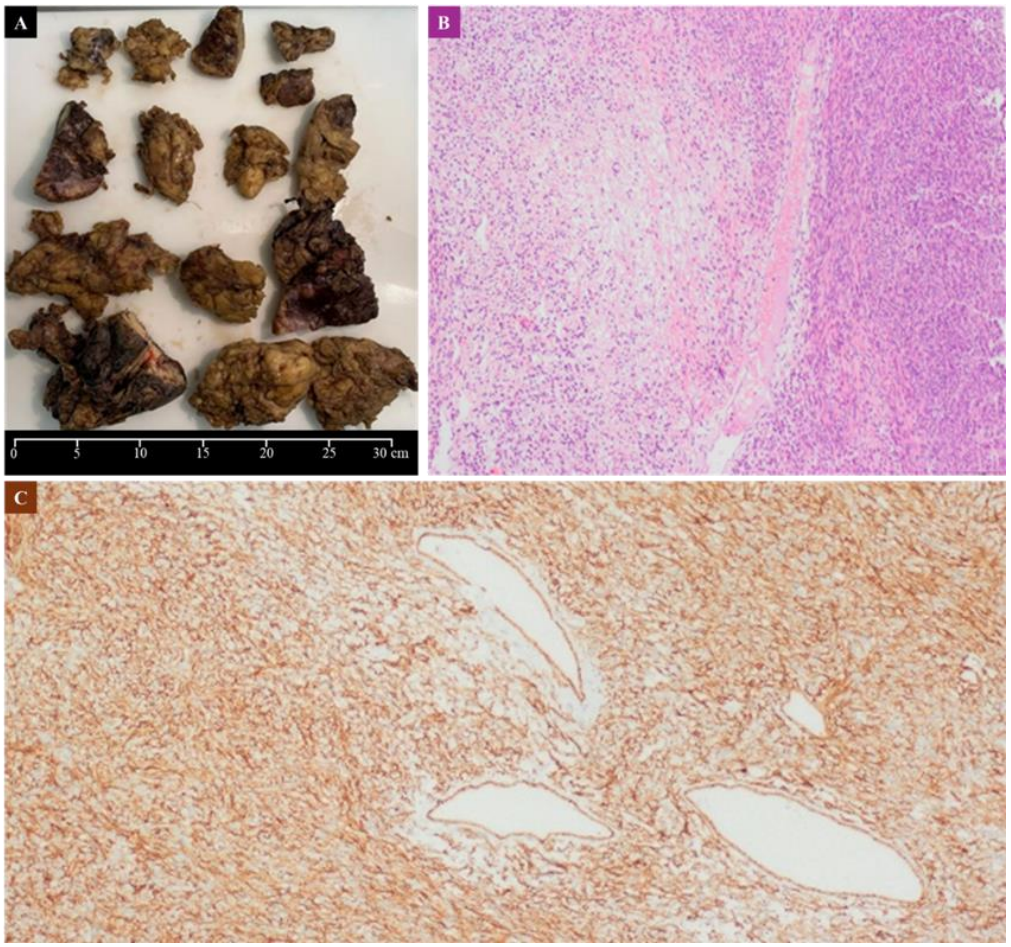

Figure 4. Histopathological Findings of the Solitary Fibrous Tumor. (A) Macroscopic examination reveals multiple fragments. The largest measuring 14 x 10 cm and the smallest 6 x 3 cm. with a grayish coloration and elastic consistency. Containing adipose and fibrous tissue. (B) Microscopic examination at 10x magnification shows a fibroblastic neoplasm with a storiform cellular proliferation. Demonstrating variable cellularity with areas of both hypocellularity and hypercellularity. (C) Immunohistochemical staining at 40x magnification shows strong and homogeneous positivity for CD34, supporting the diagnosis of a solitary fibrous tumor.

Discussion

The solitary fibrous tumor (SFT) is a rare neoplasm that originates from mesenchymal cells and represents less than 2% of soft tissue sarcomas (3).). It predominantly affects adults, typically between the ages of 20 and 70, with a median age range of 50 to 60 years. Its distribution is widespread, with common locations including the abdominal cavity (31%), extremities (29%), pleura (22%), trunk (11%), and other sites (7%) (4). Most of these tumors are benign; however, approximately 10% are aggressive, with local or metastatic recurrences (5).

Patients with SFT are often asymptomatic; however, symptoms can arise from compression of adjacent structures, depending on the tumor's location and size (3). Hypoglycemia in these patients, known as Doege-Potter syndrome (DPS), is present in 2 to 4% of cases (6). Less than 10% of patients present with hypertrophic osteoarthropathy, characterized by clubbing of the fingers, hypertrophic skin changes, and increased periosteal activity on radiographs (4), which was not observed in the case we report.

In DPS, facial findings are consistent with acromegalic features and are thought to result from IGF-2-mediated stimulation of receptors related to IGF-1 and insulin receptor subtypes (7). In a review of 71 cases of DPS by Han et al., two patients were described with acromegalic features (8). Additionally, cutaneous lesions such as seborrheic keratosis, acrochordons, and rhinophyma have been documented in patients with IGF-2-producing tumors (9). The latter lesion was present in our patient. This unusual combination of acromegalic features and hypoglycemia is not exclusive to IGF-2-producing tumors, as it can also be seen in rare cases of multiple endocrine neoplasia type 1 (MEN1), which includes both insulin-secreting tumors and growth hormone-secreting tumors. IGF-2 has multiple actions that may contribute to the development of hypoglycemia (9). The primary mechanism involves inhibition of hepatic glucose production. Like insulin, IGF-2 can inhibit gluconeogenesis, glycogenolysis, and ketogenesis via activation of the insulin receptor (10). The initial diagnostic approach to DPS includes imaging and hormonal studies, while definitive diagnosis requires histopathological confirmation (11). As it results from hypoglycemia induced by non-islet tumor cells, DPS presents with low levels of IGF-1, insulin, C-peptide, proinsulin, and beta-hydroxybutyrate. A ratio of IGF-2 to IGF-1 >10 confirms the diagnosis of hypoglycemia due to non-islet tumor cells (10). In our patient, the baseline low levels of C-peptide and insulin were due to the fact that the hypoglycemia was not the result of endogenous hyperinsulinism. As an adaptive response to the already present hypoglycemia, the body may reduce insulin and C-peptide secretion. The IGF-1 level was within the normal range, and IGF-2 was not measured due to logistical limitations in our setting. Regarding pleural SFT, its discovery is often incidental on chest radiography or computed tomography (CT) (12). CT findings are generally nonspecific, showing hypo- or hyperdensity compared to muscle and may or may not present heterogeneous enhancement (12). The definitive study is typically the immunohistochemical analysis of the resected tumor, where positive expression of CD34 (13) and STAT6 (14) confirms the diagnosis. In patients with DPS, surgical resection of the SFT is the most effective curative treatment and prevents recurrence of hypoglycemia (15). If surgery is not curative, other options such as chemotherapy or tumor chemoembolization can be considered, along with symptomatic treatment including increased caloric intake, dextrose administration, use of somatostatin analogs, recombinant growth hormone, diazoxide, and corticosteroids (11); the latter suppress IGF-2 production and stimulate gluconeogenesis (16).

Although SFTs typically exhibit a benign clinical course, with improvement of acromegalic features (7), resolution of hypoglycemia, and a survival rate greater than 90% after complete surgical resection (8), a subset of these tumors can exhibit malignant behavior, leading to local recurrence and distant metastasis (17), potentially affecting the liver, lungs, and central nervous system (18). There are histopathological criteria for poor prognosis associated with a high risk of recurrence, such as high mitotic activity, necrosis or hemorrhage, large tumor size, cellular pleomorphism, or vascular invasion (5). A systematic review found that the main factors associated with tumor recurrence are a high Ki67 index and the presence of tumor necrosis (19). In conclusion, the combination of persistent hypoglycemia, acromegalic features, and thoracic tumors should be considered a red flag for the possibility of Doege-Potter syndrome, even in contexts where specific diagnostic tests such as IGF-2 measurement are unavailable. Timely identification of DPS is crucial to improving prognosis.

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