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CLINICAL CASE

Hypoglycemia secondary to solitary fibrous pleural tumor: A doege-potter syndrome case report

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Abstract

Solitary fibrous pleural tumor is rare. When associated to hypoglycemia as paraneoplastic syndrome, it is known as Doege-Potter syndrome. The first-line treatment is surgical resection, directly associated to instant hypoglycemia remission. Hereby, we present the case of a 57-year-old woman complaining of recurrent hypoglycemia episodes. Imaging studies demonstrated on the left hemithorax an heterogeneous tumor. The patient agreed to surgical intervention. Block resection was performed through posterolateral thoracotomy. Afterward, the patient's hypoglycemia episodes ceased. In occasions, hypoglycemia episodes can be potentially lethal, if not attended. Surgical treatment may resolve hypoglycemia episodes, digital acropachy, and long-term survival with no recurrence.

Keywords: *Pleural fibrous solitary tumor. Doege-Potter Syndrome. Hypoglycemia. Thoracotomy. Thoracic surgery.*

Hipoglucemia secundaria a tumor pleural solitario fibroso: reporte de un caso de síndrome de Doege-Potter

Resumen

El tumor pleural solitario fibroso es infrecuente. Se conoce como síndrome de Doege-Potter cuando se asocia a hipoglucemia como síndrome paraneoplásico. El tratamiento de elección es la resección quirúrgica. Presentamos el caso de una mujer de 57 años, quien se aborda por múltiples episodios de hipoglucemia. Los estudios de imagen revelaron una tumoración heterogénea en hemitórax izquierdo. Se realizó resección en bloque mediante toracotomía posterolateral y posteriormente los episodios de hipoglucemia remitieron. La hipoglucemia puede ser potencialmente letal en caso de no abordarse. El tratamiento quirúrgico puede resolver los episodios de hipoglucemia, acropaquia y una supervivencia a largo plazo sin recurrencia.

Palabras clave: *Tumor pleural fibroso solitario. Síndrome de Doege-Potter. Hipoglucemia. Toracotomía. Cirugía torácica.*

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Introduction

Fibrous solitary tumor represents 5% of thoracic benign neoplastic masses and 10% pleural masses. So far, it has not been associated to any etiologic factor[1]. Its course is usually benign and very often incidental diagnosis when performing a chest radiograph[2]. It may present cough, dyspnea, thoracic pain, pleural effusion, arthralgia, acropachy, and hypoglycemia episodes. These tumors cause extra thoracic symptoms such as osteoarthrosis and refractory hypoglycemia, due to a growth hormone-like substance produced, as well as insulin-like growth factor 2 (IGF-2), respectively[3].

In this manuscript, we describe a rare case of a woman attended in the emergency room due to recurrent hypoglycemia cases. Along the diagnosis approach, we found different clinical remarks on examination which guided us to focus on thoracic imaging studies, where a round-large-sized tumor on left hemithorax was revealed.

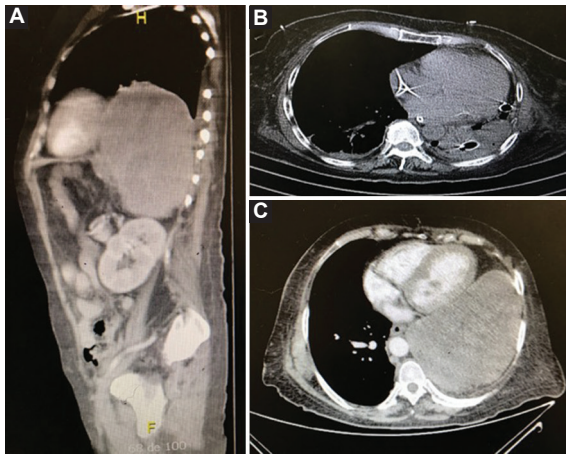

Figure 1. Tomographic imaging of thoracic mass. **A:** saggital thoracic-abdominal CT scan revealing a solid isodense tumor in between both cavities, posterior to the heart, above the spleen. **B:** Axial CT scan on T4, post-surgical image, where the rotation of the mediastinum toward the let hemithorax is observed. **C:** Axial CT scan on T6, thoracic mass posterior to the heart, misplacing it toward median line.

Case presentation

Informed consent

Informed consent for the publication of this manuscript was obtained from the patient and legally responsible relative, before the drafting of this manuscript.

Patient information and timeline

A woman with past medical history of hypertension, in her late 50's arrives to the emergency room complaining about lipothymia. Eighteen months prior, the patient recalls shortness of breath symptoms, along with violaceous coloring and size growth within his hand's nails; signs and symptoms for which she didn't seek medical assistance since she didn't consider them relevant.

Clinical findings and diagnostic assessment

At her arrival to the emergency room, her vital signs where found in normal range. Her blood oxygen-saturation was of 98% with pulse-oximeter, and a capillary glycaemia of 45 mg/dL. During physical examination, abnormal findings include left hemithorax vesicular murmur abolished, acropachy on both hands. Her lab results of hematic biometry, serum electrolytes, and serum creatinine were unremarkable within normal ranges, 12-lead resting electrocardiogram shows sinus rhythm, 75 beats/min, non-ischemic, or necrosis injury data. Chest X-ray showed opaque pattern along the left hemithorax. A hypodense mass thrusting mediastinum and inferior left lung lobule are identified through thoracic CT scan (Fig. 1).

The patient was hospitalized for follow-up related to capillary glycemic level, along with 20% dextrose in case of symptomatic hypoglycemia.

Therapeutic intervention

Needle biopsy was performed, pathology results reported non-conclusive diagnosis, therefore, surgical approach was performed at the OR. A left posterolateral thoracotomy with tumor block resection was employed, where tumor was found adhered to inferior lobule of the left lung and diaphragm. Tumor-free margins were identified before surgical closure. Histopathology results confirmed fibrous pleural solitary tumor diagnosis, measurements 20 × 16 × 11 cm (Fig. 2), without vascular or lymphatic invasion, immunohistochemistry was positive for CD-34.

Follow-up and outcomes

Posteriorly to resection, the patient was closely monitored in intensive unit care during 48 h. After 24 h

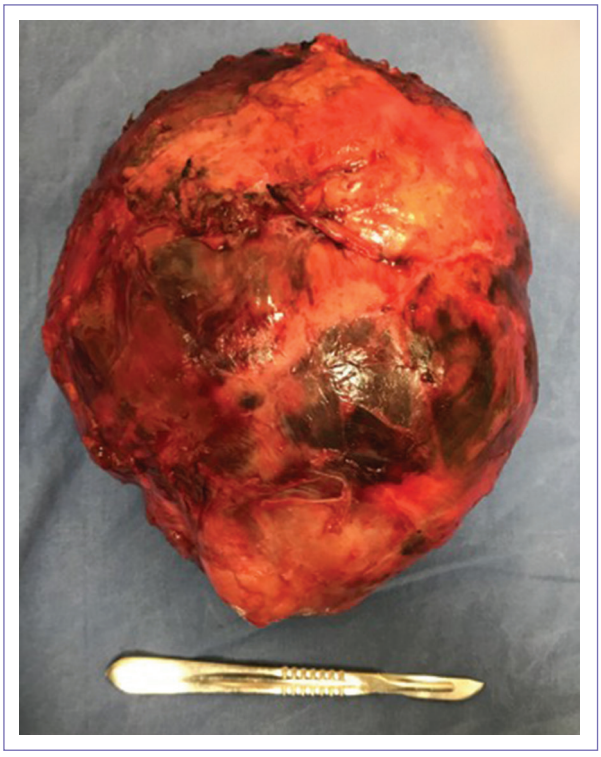

Figure 2. Macroscopic image of the resected tumor. Solitary fibrous pleural tumor removal through left posterior lateral thoracotomy with mass block resection.

post-resection, endotracheal tube was removed. Thoracostomy tube was removed 96 h post-resection, once drainage in 24 h was below 100 ml, and lung re-expansion was observed in chest X-ray. During the 120 h post-resection, there were no hypoglycemia events registered, along follow-up visits and imaging studies, no recurrence was observed, with an overall favorable outcome.

Discussion

In our patient's case, we performed a total tumor resection. For this to be possible, a block resection was required, including a diaphragm portion and left lung's inferior lobe, which explained the sudden absence of hypoglycemia episodes post-surgical intervention, along with a favorable long-term outcome.

Solitary fibrous pleural tumor is a very rare solid neoplasia. Major age incidence is 60-80-year-old population, slightly more common in male gender (58%). Up to 80% of cases turn out non-malignant. Regularly tumor's origin is visceral pleura. About 25% of cases reported to present hypoglycemia, Hippocratic fingers, or pleural effusion[1].

Hypoglycemia is present in 20% of solitary fibrous pleural tumor cases, meanwhile hypertrophic arthrosis in 55%. A solitary fibrous pleural tumor revision describes the total surgical resection accomplished in almost all subjects, resulting in a non-recurrence survival on the long-term follow-up as well as remission of hypoglycemic episodes along with acropachy[1].

Hypoglycemia in solitary fibrous pleural tumor is associated to large tumor size and high mitosis rate, yet the pathophysiology is known to be caused by the neoplastic synthesis of high-molecular-weight IGF-2 (known as HMW-IGF-2)[1]. This protein hormone shares a molecular structure similar to insulin, so forth capable of activating insulin receptors, as well as stimulating peripheric glucose capture, and inhibiting hepatic gluconeogenesis; resulting in hypoglycemia episodes difficult to resolve, with continuous recurrences[2]. Recurrences are expected, since this high weighted molecule has an increased half-life and ranges a higher circulating concentration in contrast to insulin[2].

Treatment of choice is surgical resection. Total tumor resection leads to an effective clinical resolution (hypoglycemia and Hippocratic fingers)[1,2]. Several surgical techniques have been previously described (thoracotomy, thoracoscopy, and video-assisted thoracic thoracoscopy), depending on size, location, and tumor's characteristics. Most patients' prognosis is favorable, especially when tumor is non-malignant and total surgical resection is possible. Local recurrence is expected when incomplete tumor resection is performed; incidence of recurrence in described cases is of 2-8%. Furthermore, metastasis is expected in malignant tumors with incomplete surgical excision[4]. When tumor is unresectable and metastatic, chemotherapy, long-term glucocorticoid treatment, continuous dextrose or glucagon infusion, and selective embolization of tumor arteries are described alternatives of treatment to ease the recurrent symptomatic hypoglycemia, although neither one is considered as treatment of choice. Radiotherapy can be used as adjuvant therapy after surgical resection in a malignant tumor[5]. Recurrence is more frequent in malignant tumors in comparison to benign tumors, 75% and 25%, respectively[5]. A 10-year survival prevalence of 97% is known in benign tumors and 89% in malignant tumors[4].

Although we acknowledge the limitation of a case report, we consider the low prevalence of this disease, as well as the clinical insight and surgical approach described in our manuscript which may impact future

Doege-Potter syndrome cases when attended by our colleagues.

Conclusion

Solitary fibrous pleural tumor is a subset of mesenchymal neoplasia. Two out of three of these tumors occur in visceral pleural, sparing third of these occurring in parietal pleura. It is known in rare occasions these hypoglycemia episodes can be potentially lethal, particularly if not attended. This severe and recurrent hypoglycemia episodes are triggered by HMW-IGF-2 effecting its biological and hormonal activity within insulin receptors. Surgical treatment may not only resect total tumor but also it resolves hypoglycemia episodes, digital acropachy, and long-term survival with no recurrence.

Authors' contributions

Conception and design: Erwin R. Flores-Vázquez and Rodrigo Uribe-Pacheco.

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Analysis and interpretation of data: Erwin R. Flores-Vázquez, Rodrigo Uribe-Pacheco, Guadalupe J Vázquez-Ramírez, Luis E. Suárez-Luna, and Irene Irisson-Mora.

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Approved the final version of the manuscript on behalf of all authors: Erwin R. Flores-Vázquez and Rodrigo Uribe-Pacheco.

Administrative/technical/material support: Guadalupe J Vázquez-Ramírez, Luis E. Suárez-Luna, and Irene Irisson-Mora.

Study supervision: Erwin R. Flores-Vázquez, Rodrigo Uribe-Pacheco, and Guadalupe J Vázquez-Ramírez.

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Conflicts of interest

The authors declare that they have no conflicts of interest.

Ethical disclosures

Protection of people and animals. The authors declare that no experiments were performed on humans or animals for this research.

Confidentiality of the data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the informed consent of the patients and/or subjects referred to in the article.

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