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Case Report

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Doege Potter Syndrome, A Very Rare Cause of Hypoglycemia

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Abstract

Doege-Potter syndrome is a rare paraneoplastic syndrome that presents as hypoinsulinemic hypoglycemia from ectopic secretion of an insulin-like growth factor II prohormone (IGF-II) from a solitary fibrous tumor. Surgical resection is curative in most cases [1]. We presented a 32-year-old male patient with Doege-Potter syndrome after pulmonary resection after investigations due to hypoglycemia attacks, in the light of literature.

Introduction

Doege-Potter syndrome is a rare paraneoplastic syndrome diagnosed incidentally during questioning of hypoglycemia of unknown etiology. It is characterized by hypoglycemia from a solitary fibrous tumor (SFT), a non-islet cell tumor secondary to overproduction of the hormone IGF-II [2]. Generally, these tumors are intrathoracic, benign, and asymptomatic. Occasionally it can occurs as a paraneoplastic syndrome; Hypertrophic osteoarthropathy in Pierre-Marie-Bamberger syndrome and hypoglycemia in Doege-Potter syndrome. NAB2-STAT6 gene fusion is the hallmark of SFT [3]. In this article, we present a case of a 32-year-old non-diabetic male with recurrent episodes of hypoglycemia diagnosed with Doege-Potter syndrome secondary to PFT pleura. The tumor was positive for the NAB2-STAT6 gene fusion on RT-PCR. The giant tumor became asymptomatic within 24 hours following resection of the mass and remained asymptomatic at 12-month follow-up.

Case

A 32-year-old male patient was admitted to our clinic because of the intermittent episodes of hypoglycemia for 6 months. After episodes of hypoglycemia, it improves after receiving an intravenous infusion of 10% dextrose and intermittent intravenous bolus administration of 50% dextrose. On clinical examination, there is a weight loss of approximately 10 kg without cough, shortness of breath or chest pain. The patient had no history of diabetes mellitus and was not taking any glucose-lowering medication. He had a 10 pack-year smoking history. On physical examination, without any enlargement of the superficial lymph nodes; decreased breath sounds, dullness to percussion, and tactile fremitus were absent in the right middle and upper lung areas. Cardiovascular and abdominal examinations were unremarkable. Routine clinical laboratories, urine and peripheral blood studies, and tumor markers (ie, carcinoembryonic antigen [CEA], alpha-fetoprotein Carbohydrate antigen 125, Carbohydrate antigen 199, neuron-specific enolase, and Cytokeratin 19 fragments) were all within normal limits. Serum growth hormone (GH) level was normal. Despite marked hypoglycemia (1.4 mmol/L), the serum insulin level was less than 0.2 μIU/mL (normal range: 2.6–24.9) and the C-peptide level was 0.41 nmol/L (normal range: 1.1–4.0 nmol/L). Serum IGF-II level was 1038.71 μg/dL and IGF-I level was less than 25 μg/L.

Computed tomographic (CT) scan revealed a large heterogeneous mass 5.4 × 6.3 × cm in size, covering the upper region of the right hemithorax, with rough edges and poor borders (Figure 1).

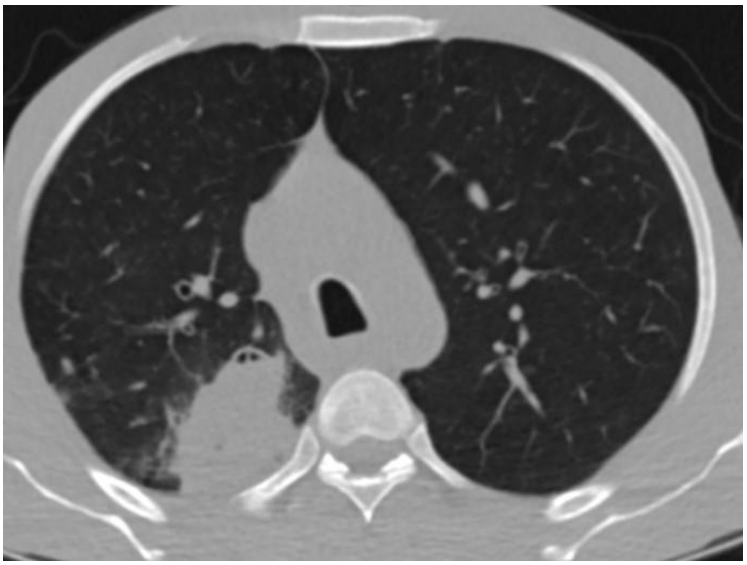

Figure 1: On computed tomography (CT), a large heterogeneous mass of 5.4 ×6.3 × cm, covering the upper region of the right hemithorax, with rough edges and poor borders.

Bone emission of liver and pancreas was unremarkable on CT and MR scan. A 60 mm lesion was observed in the right upper lobe on positron emission tomography (PET-CT) (SUVmax 4.6). No endobronchial lesion was detected in fiberoptic bronchoscopy. The diagnosis could not be made by bronchoscopy and transthoracic cutting needle biopsy. Pulmonary function tests were within normal limits. With video-assisted thoracoscopic surgery, the mass originated from the anterior mediastinum and was completely resected with invasion of the right lung upper lobe, pericardial and phrenic nerves. Right upper lobe of lung, part of pericardium and phrenic nerve were resected. The tumor was a grayish-white solid measuring 20×16×12 cm and partially encapsulated by local necrosis at the surface. Histological examination revealed nested fascicles of spindle tumor cells regular with minimal nuclear pleomorphism and negligible mitotic activity. Immunohistochemical stains showed strong positivity for CD34 expression and positive expression for IGF-II (figure 2). Pathology and clinical status confirm the diagnosis of malignant PFT.

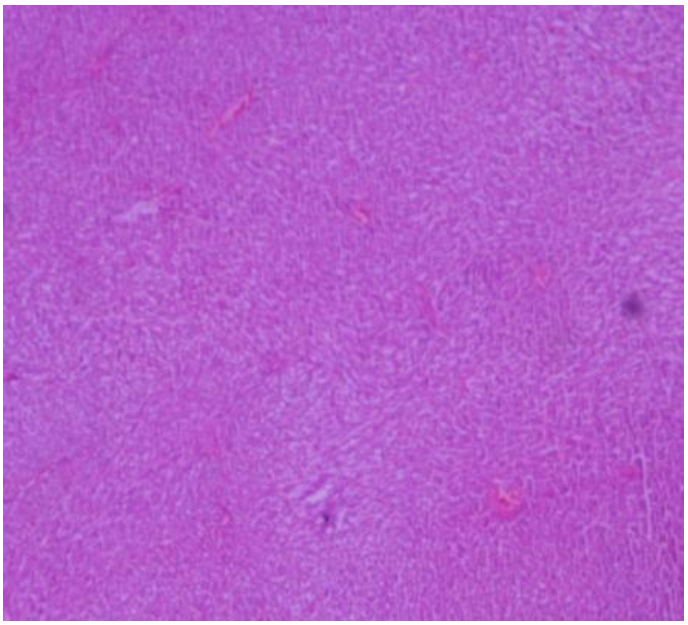

Figure 2: Histopathology imaging (100% magnification) reveals nested fascicles of regular spindle-shaped tumor cells with minimal nuclear pleomorphism and negligible mitotic activity.

Postoperatively, serum glucose levels returned to normal and episodes of hypoglycemia resolved. Blood sugar and insulin levels return to normal. In addition, serum IGF-II levels decreased from 1029.51 to 604.66 μg/dL. During the 6-month follow-up period, no cases of hypoglycemia occurred. No pathology was observed in the postoperative period. Patient consent was obtained for our study.

Discussion

Solitary fibrous tumors (SFT) are a rare group of neoplasms originating from mesenchymal cells. Doege-Potter syndrome is a rare manifestation of SFTs, occurring in less than 5% of patients with SFTs. It is a paraneoplastic syndrome with recurrent non-islet cell tumor hypoglycemia (NICTH) due to over-secretion of "large" insulin-like growth factor II (IGF-II) from the PFT [2,3]. This syndrome was first described in 1930 [4] and has only been reported in case reports since then. To date, there are no systematic studies on this rare clinical entity. These are usually slow-growing (80-85%) neoplasms with low malignant potential. Han et al. [5] reported that SFTP is the most common type of SFT associated with Doege-Potter syndrome. The fusion of NAB2-STAT6 (NGFI-A binding protein 2 [NAB2] gene and signal transducer and activator of transcription 6 [STAT6] gene) results in a chimeric nuclear protein that causes constitutive activation of transcription and abnormal proliferation of cells leading to tumorigenesis [4,5].

The underlying tumor may be a benign or malignant pleural tumor but may be present in extrapleural sites. For the diagnosis of Doege-Potter syndrome, hypoglycemia and symptoms attributable to low blood glucose levels must be present with secretion of the prohormone IGF-II [6-8]. Our patient had hypoglycemia attacks. While investigating the cause of hypoglycemia attacks, PFT was detected. After the surgical resection, there were no episodes of hypoglycemia.

Definitive management of Doege-Potter syndrome includes treating the underlying tumor, including complete resection of the tumor mass, palliative debulking, chemotherapy, radiofrequency ablation, cryoablation, or chemoembolization. Until definitive treatment, treatment of hypoglycemia includes increased caloric supplementation, dextrose infusion, glucocorticoids, glucagon, or recombinant human growth hormone (rhGH). Glucocorticoids act by increasing serum large IGF-II clearance and impairing endogenous insulin activity [6]. Few case reports documenting success with oral prednisone 40-60 mg once daily; however, in large tumors and extensive disease, oral dexamethasone was better than prednisone at reducing the degree of hypoglycemia. A continuous infusion of glucagon can be used for resistant hypoglycemia, but is often unsustainable. The use of rhGH has been successful in some cases, but carries a theoretical risk of tumor progression due to increased IGF-1 levels.[7] In our case, the symptoms did not impro ve with m edical treatment. After surgical resection, a complete response to the treatment was obtained in our case.

Treatments and outcomes depend on the histological grade and stage of the tumor. Benign lesions are best treated by simple resection, and complete resection prevents further hypoglycemic episodes. Malignant lesions require complex en bloc resection of the tumor and adherent tissues. Radiotherapy and chemotherapy have been described for treatment and palliation in histological analysis following resection or when surgical treatment is not possible for the patient.

Resources

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