



Doege-Potter Syndrome and How a Benign Tumour Spreads: Case Report

Caldeira V^{1*}, Sá T¹, Silva S¹, Murinello N¹, Reis JE^{2,4}, Bravio I³, Martelo F⁴ and Cardoso J¹

¹Department of Pneumology, Hospital Santa Marta, Brazil

²Department of Cardiothoracic Surgery, Hospital Santa Marta, Brazil

³Department of Thoracic Surgery, Instituto Português Oncologia, Portugal

⁴Department of Thoracic Surgery, Hospital da Luz, Portugal

Abstract

Solitary Fibrous Tumors of the Pleura (SFTP) are rare tumors, usually benign. Doege-Potter Syndrome (DPS) occurs in less than 5% of TFSP and consists of refractory hypoglycemia associated with these tumors. We report the case of a 65-year-old woman with a confusional syndrome of sudden onset associated with hypoglycaemia. Chest Computed Tomography (CT) revealed a massive mass occupying almost the entire left hemithorax, which CT guided biopsy revealed a SFTP. The patient underwent surgery with complete excision of the tumor and resolution of the hypoglycemia. The pathological anatomy of the specimen identified a benign SFTP; however, 2 years later she developed a lung metastasis. SFTPs are slow-growing tumors usually benign; however a recent review reported an increased malignancy in the DPS form. The clinical course of these tumors is unpredictable and long-term surveillance is essential because of the risk of late recurrence or metastatic disease.

As peculiarities of this case, we highlight the rarity of the form of presentation (with DPS), the fact of being a giant tumor of the pleura, and although histologically benign, showing a malignant evolution with metastatic disease.

Introduction

Solitary Fibrous Tumors of the Pleura (SFTP) are rare primary tumors of mesenchymal origin, accounting for less than 5% of pleural tumors. Mostly benign, about 5% to 14% have malignant behavior, specially the larger ones, with risk of local recurrence after excision and metastatic disease [1-3].

Hypoglycemia accompanying SFTP was independently described by Karl Doege and Roy Potter in 1930 and is referred as Doege-Potter Syndrome (DPS), occurring in 5% to 10.4% of all patients [2]. It is caused by hyper secretion of an abnormal molecule of IGF-II (insulin-like growth factor-II) by these tumors [4]. According to a recent review, there are 76 published cases between 1989 and 2016, 60.6% of which were malignant, a rate significantly higher than previously reported [2]. Usually without sex predominance, most recent data suggest a slightly male predominance and a peak of incidence in the 6th to 8th decade of life [2]. The effective treatment of these tumors is complete surgical resection, with favorable prognosis in the majority of cases and allowing the resolution of hypoglycemia associated with this syndrome [2]. The risk of recurrence or metastasis seems to increase with the tumour size and if a malignant component is detected in histologic analysis [5]. Although uncommon, distant spread and aggressive growth could occur in some benign tumours [6-8].

Case Presentation

A 65-year-old female patient, non-smoker with a history of rheumatoid arthritis and hypothyroidism on regular therapy and a presumptive diagnosis of pulmonary tuberculosis treated with classic antibacterial regimen. She developed acute confusional syndrome and lost consciousness in the context of severe hypoglycemia (45 mg/dl). Clinical examination revealed abolition of the vesicular murmur throughout the left hemithorax and hypoxemia (pO₂ 61.4 mmHg). Computed Tomography (CT) scan confirmed a solid and heterogeneous mass, very vascularized and occupying almost the whole of the left hemithorax, with passive pulmonary atelectasis and contralateral deviation of the mediastinum. During hospitalization, the patient experienced several episodes of difficult-to-control hypoglycemia. CT-guided transthoracic aspiration biopsy revealed a SFTP, but with no

OPEN ACCESS

*Correspondence:

Vânia Caldeira, Department of
Pneumology, Santa Marta Hospital,
Brazil,
E-mail: vania.medicina@gmail.com

Received Date: 16 Jan 2019

Accepted Date: 09 Feb 2019

Published Date: 13 Feb 2019

Citation:

Caldeira V, Sá T, Silva S, Murinello N,
Reis JE, Bravio I, et al. Doege-Potter
Syndrome and How a Benign Tumour
Spreads: Case Report. *Clin Oncol.*
2019; 4: 1572.

Copyright © 2019 Caldeira V. This is
an open access article distributed under
the Creative Commons Attribution
License, which permits unrestricted
use, distribution, and reproduction in
any medium, provided the original work
is properly cited.

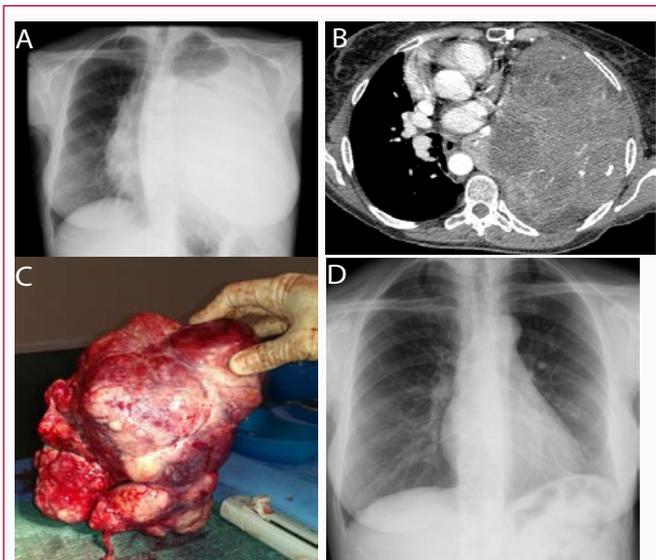


Figure 1: A - Chest X-ray with the left hypo transparency. B - Computed tomography of the chest showing a solid and heterogeneous mass, very vascularized and occupying almost the whole of the left hemithorax and with deviation of the mediastinum. C - The excised tumour. D - Follow-up chest X-ray 1 week after the surgery.

possibility of additional histological characterization. The patient underwent left antero-latero-posterior thoracotomy with complete excision of bulky capsulated, vascularized and pediculated mass, and subsequent pulmonary expansion was observed. The postoperative period was unremarkable and the patient was discharged on the 10th day. The anatomopathological study identified a mass with 30 cm × 18 cm × 12 cm and 2645 g, showing tumor formation without atypia, with slight hemorrhage, but without areas of necrosis, with about 2 mitoses per 20 fields of enlargement and low proliferative index (5%). Immunohistochemical analysis was positive for CD34 and bcl2. These results confirmed that the SFTP had benign histological characteristics (D). Two months later, the patient was asymptomatic, with a chest X-ray without alterations, and normalization of glycemia and insulinemia levels. However, in the follow-up period, 2 years after the surgery it was found a single nodule in the contralateral lung. The patient underwent nodule resection and the histology was again compatible with a benign SFTP although the malignant behavior with a lung metastasis. The patient maintained surveillance and another 2 years later appeared with a new nodule, now in the left lung. Because of the history of previous surgery, after discussion in multidisciplinary reunion, it was proposed for radiotherapy. However, the patient was not willing to undergo further treatment and she abandoned the follow-up.

Discussion

SFTP are rare and slow-growing tumors remaining asymptomatic for long periods of time [1], as happened in this clinical case, in which after reviewing the patient's history it was possible to establish the existence of a lesion compatible with the current location of the tumor, with indolence of about 2 decades. As in other cases, hypoglycemia with neurological symptoms was the presentation of the disease, known as DPS [2]. The laboratory determination of IGF-II is not always available, however, the resolution of hypoglycemia with surgical excision of the tumor, as happened in our case, confirms the diagnosis [1,9]. In addition to the rare clinical presentation, in

this report, the size of the tumor is also highlighted. Giant pleura tumors, greater than 10cm or heavier than 2 kg, are extremely rare and more likely to be malignant [10]. Complete surgical excision is the treatment of choice for all SFTP allowing, as in this case, the resolution of hypoglycemia [1]. Surgery besides curative is diagnostic of SFTP in many cases, although in this patient, it was possible to perform the histopathological diagnosis preoperatively [1]. According to the criteria of England, due to the low cellularity and low mitotic index, absence of nuclear atypia and areas of necrosis in the operative part, our case was a SFTP with benign characteristics and, therefore, with expectable favorable prognosis [11]. Recently, large tumor size (>15 cm), age older than 55 years and mitotic activity >4 per 10 high power fields were considered to be worst prognosis factors [12]. It is also known that these tumors have a very unpredictable behavior, with some cases of late recurrence despite the histology of benignity, so long-term surveillance is fundamental in these patients [1,8]. Our patient had two important risk factors for aggressive tumor behavior - her age and the large size of the tumor [12]. In fact, despite the diagnosis of benignity, she had a rare evolution with metastatic lung disease. There is a previous case report of a patient with a benign SFTP with small foci of malignant disease [6] and we could suppose if the same happened with our patient. As described in the literature, recurrence is almost always ipsilateral [13] and metastatic disease usually affects lung, liver and bone [8]. However, our patient had a contralateral lung metastasis. Radiotherapy and chemotherapy have been described as an alternative treatment but its efficacy and prognostic impact is not clear [4,5]. In this case, although multidisciplinary discussion suggested radiotherapy, according to patient wills, no additional treatment was performed.

Finally, we conclude that in cases of hypoglycemia without apparent cause, the DPS should be considered in the differential diagnosis. Although rare, this paraneoplastic syndrome is an important and reversible cause of hypoglycemia, which is resolved by complete surgical excision of the tumor. However, the clinical course even of the benign tumors is unpredictable and long-term surveillance is recommended for the detection of eventual late recurrences.

References

- Meng W, Zhu HH, Li H, Wang G, Wei D, Feng X. Solitary fibrous tumors of the pleura with Doege-Potter syndrome: a case report and three-decade review of the literature. *BMC Research Notes*. 2014;7:515.
- Han G, Zhang Z, Shen X, Wang K, Zhao Y, He J, et al. Doege-Potter syndrome. A review of the literature including a new case report. *Medicine*. 2017;96(27): e7417.
- Balduyck B, Lauwers P, Govaert K, Hendriks J, De Maeseeneer M, Van Schil P. Solitary fibrous tumor of the pleura with associated hypoglycemia: Doege-Potter syndrome: A Case Report. *J Thorac Oncol*. 2006;1(6):588-90.
- Zafar H, Takimoto CH, Weiss G. Doege-potter syndrome: hypoglycemia associated with malignant solitary fibrous tumor. *Med Oncol*. 2003;20(4):403-8.
- Ahluwalia N, Attia R, Green A, Cane P, Routledge T. Doege-Potter Syndrome. *Ann R Coll Surg Engl*. 2015;97(7):e105-7.
- Scrimgeour LA, Grada Z, Aswad BI, Ng T. Lessons learned from an untreated "benign" thoracic tumour. *Ann Thorac Surg*. 2017;103(2):e135-7.
- de Perrot M, Kurt AM, Robert JH, Borisch B, Spiliopoulos A. Clinical behavior of solitary fibrous tumors of the pleura. *Ann Thorac Surg*. 1999;67(5):1456-9.

8. Ali JM, Ali A, Van Leuven M, Bartosik WR. Giant solitary fibrous tumor of the pleura an unpredictable entity: case series and literature review. *Ann R Coll Surg Engl.* 2017;99(6):e165-e171.
9. Strâmbu IR, Leonte DG, Bolca CN. Large pleural tumor revealed by severe hypoglycemia: Doege-Potter syndrome. *Rom J Morphol Embryol.* 2017;58(1):287-91.
10. Furukawa N, Hansky B, Niedermeyer J, Gummert J, Renner A. A silent gigantic solitary fibrous tumor of the pleura: case report. *J Cardiothorac Sug.* 2011;6:122.
11. England DM, Hochholzer L, McCarthy MJ. Localized benign and malignant fibrous tumours of the pleura. A clinicopathologic review of 223 cases. *Am J Surg Pathol.* 1989;13(8):640-58.
12. Demicco EG, Park MS, Araujo DM, Fox PS, Bassett RL, Pollock RE, et al. Solitary fibrous tumor: a clinicopathological study of 110 cases and proposed risk assessment model. *Mod Pathol.* 2012;25(9):1289-306.
13. Santos RS, Haddad R, Lima CE, Liu YL, Misztal M, Ferreira T. Patterns of recurrence and long-term survival after curative resection of localized fibrous tumors of the pleura. *Clinical Lung Cancer.* 2005;7(3):197-201.