GIANT PLEURAL TUMOR AND SEVERE HYPOGLYCEMIA: DOEGE-POTTER SYNDROME IN A PREVIOUSLY HEALTHY FEMALE

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Abstract

Introduction: *Doege–Potter's syndrome is a rare paraneoplastic syndrome, consisting in hypoglycemia and solitary fibrous tumor of the pleura. These tumors represent <5% of all pleural tumours and can only be cured by surgery. In this article, we report a case of a patient presenting with severe hypoglycemia, as the only symptom, and a mass occupying the entire left hemithorax.*

Case presentation: *A54 year old female with severe hypoglycemia, a chest radiography with almost total opacification of the left hemithorax and a computed tomography scan with a mass in the left hemithorax. Surgery was performed and a mass with 30cm × 18cm × 11cm weighing 3195g was resected. The postoperative course was uneventful with immediate resolution of the hypoglycemia. The immunohistochemistry diagnosis was solitary fibrous tumor of the pleura.*

Conclusions: *Solitary fibrous tumor of the pleura are very rare. Less than 5% are associated with hypoglycemia, taking the form of Doege-Potter Syndrome. Radiation therapy and chemotherapy have shown low response rate and complete surgical resection is the only procedure that offers cure.*

This case reports describes a rare giant solitary fibrous tumor of the pleura with severe hypoglycemia, successfully treated by surgery. Long-term follow-up of the patient after the surgery is necessary for detection of any possible recurrence.

INTRODUCTION

Doege–Potter's syndrome is a rare paraneoplastic syndrome that consists of the association of symptomatic hypoglycemia, as the result of excessive production of insulin growth factor (IGF) by the tumor cells, with a solitary fibrous tumor of the pleura (SFTP).[1]

SFTP are rare mesenchymal tumors representing <5% of all pleural tumours. These lesions occur predominantly in middle-aged adults with equal gender distribution.[2] Generally, there is no genetic predisposition or relationship to the exposure to asbestos, tobacco or any other environmental agents.[3]

SFTP can usually be distinguished from malignant mesothelioma by their radiographic features, gross appearance often pedunculated, immunohistochemistry characteristics and ultrastructural characteristics.[4]

Most tumors present as well-defined, slow-growing masses, which can only be cured by surgery.[5] Several case series have demonstrated complete resection to be associated with low rates of local recurrence and progression to metastatic disease.[6]

In this article, we report a case of a patient presenting with severe hypoglycemia and a SFTP occupying the entire left hemithorax.

CASE REPORT

We report the case of 54-year-old female patient that presented to the Hospital for persistent dizziness. Routine blood tests revealed glucose levels of 32 mg/dL, with no other abnormalities on the remaining parameters. Chest radiograph showed total opacification of the left hemithorax (Fig. 1a).

The patient underwent a thoracic and abdomen computed tomography (CT) scan that revealed a 28×10cm non-calcified mass in the left hemithorax, in contact with the lateral thoracic wall, well defined against the lung parenchyma, which it almost totally compressed and with

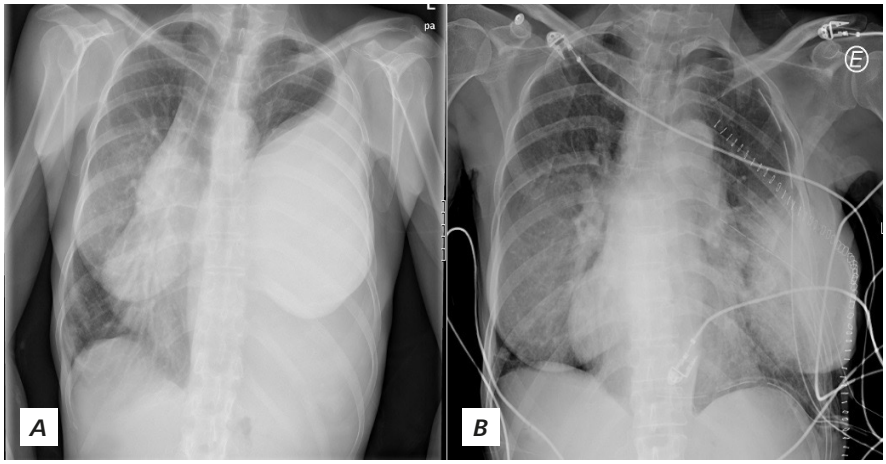

Figure 1 *(A) - Chest radiograph showed an opacification of the left hemithorax. (B) – Chest radiograph in the pos-operative, with total expansion of the left lung.*

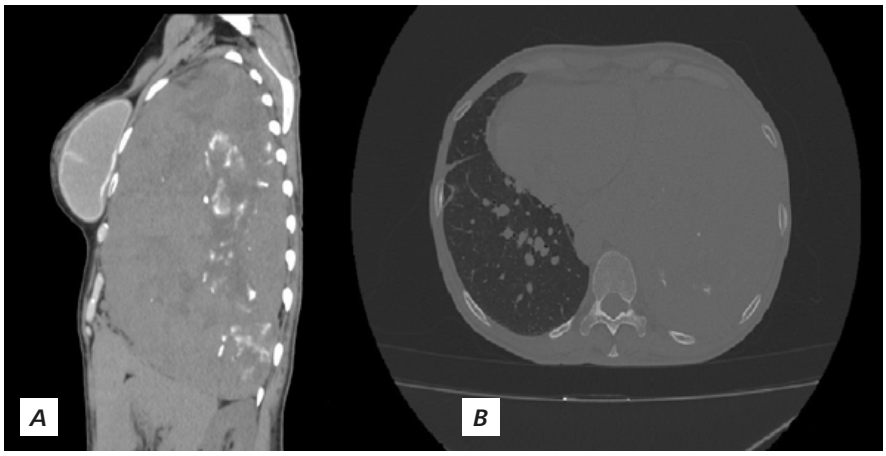

Figure 2 *Thoracic computed tomography before surgery. (A) - Mass occupying almost all the left hemithorax (sagittal view). (B) – Mass with contralateral displacement of the mediastinal structure (axial view).*

contralateral displacement of the mediastinal structures but without evidence of mediastinal or hilar lymphadenopathy (Fig. 2).

Surgery was performed under balanced anesthesia with the use of a double lumen endotracheal tube and one-lung ventilation. An antero-lateral thoracotomy through the sixth left intercostal space was performed. Resection of 2 ribs was required due to the size of the tumor (Fig. 3). Upon entering the pleura, we visualized a large encapsulated mass (Fig. 4). The tumor was attached to the superior lobe of the left lung and a wedge resection of this lobe was necessary. The main vascular pedicle of the tumor with origin in the mediastinal vessels was ligated with non-absorbable suture (Fig. 5).

The well-circumscribed, encapsulated resected mass was measured to be 30cm × 18cm × 11cm and weighed 3195 g in the fresh state (Fig. 6).

The operation took 117 min and blood loss was 250 ml. The patient remained hemodynamically stable throughout the procedure. After resection of the mass, the lung was recruited with positive pressure ventilation, achieving a good expansion of the remaining lung and no significant air leak. A thoracic epidural catheter was sited at the end of surgery and was used for post-operative analgesia. The postoperative course was uneventful, with total expansion of the lung (fig. 1b) and no respiratory complications were noted. Immediate resolution of the hypoglycemia was observed. The patient was discharged on the 6th postoperative day.

According to the morphology and cellular immunophenotype the diagnosis of malign giant pleural SFT was signed out.

The patient was observed 5 weeks after surgical resection. Follow-up chest radiography showed complete expansion of the left lung and routine blood tests were normal, with euglycemia.

DISCUSSION AND CONCLUSION

SFTP is a rare tumor with less than 800 cases reported in the literature. They can be benign (about 80%) or malign, unique or appear in multiple localizations.[7] These tumors can present with various clinical signs and symptoms, such as dyspnea, chest pain or hemoptysis.

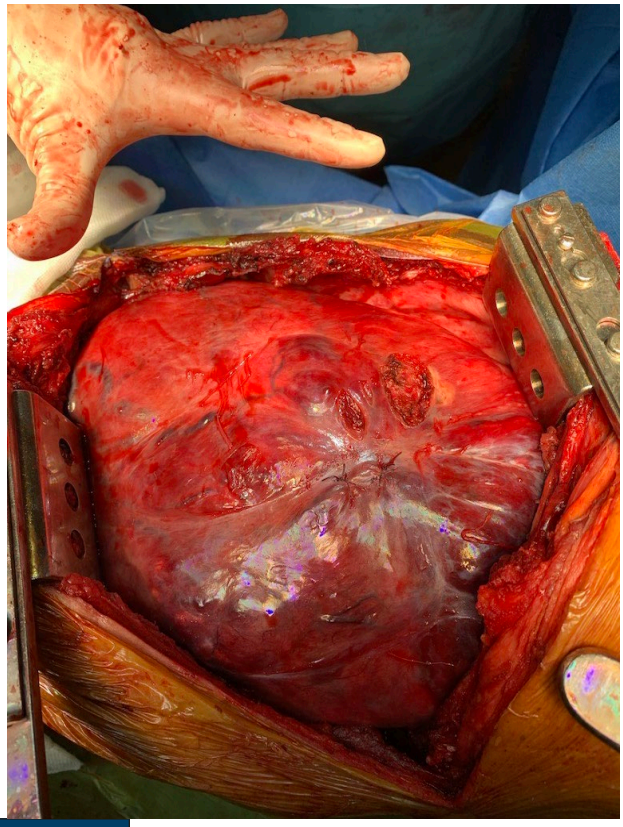

Figure 3 *Surgical approach for mass resection.*

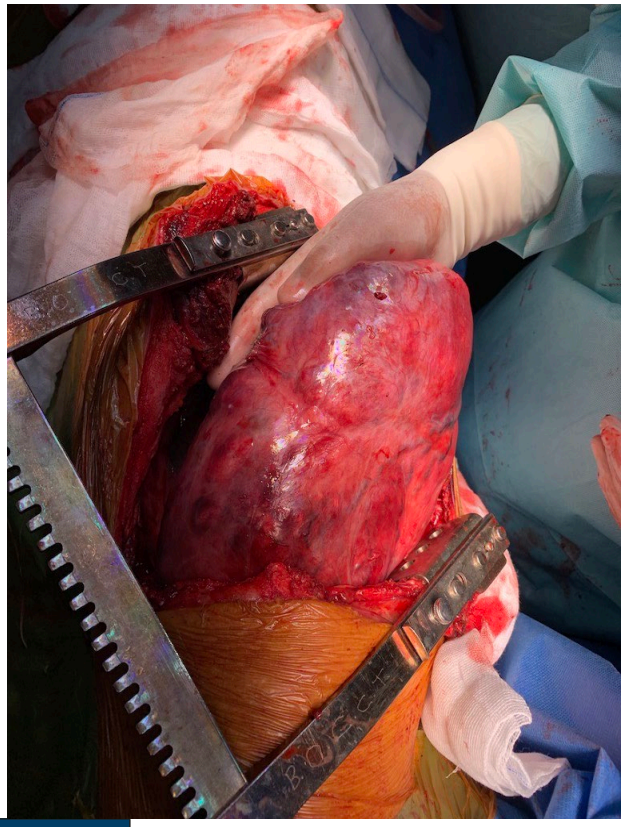

Figure 4 *SMass totally encapsulated.*

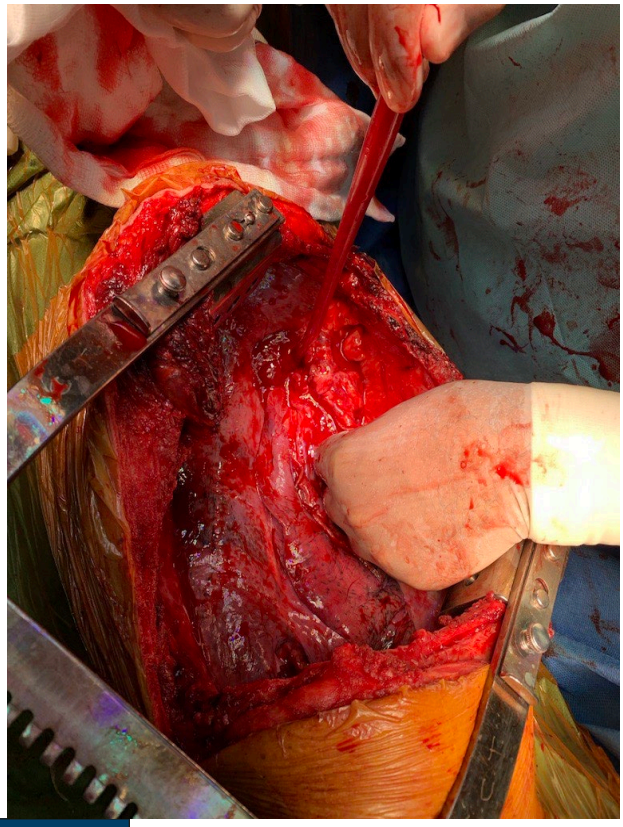

Figure 5 *Left hemithorax after mass resection.*

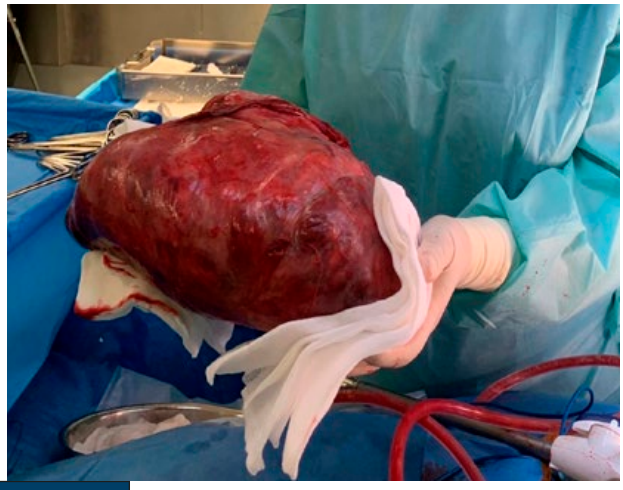

Figure 6 *A macroscopic image of the resected tumor, which weighed 3.195g.*

Less than 5% are associated with hypoglycemia, taking the form of Doege-Potter's Syndrome, a very rare entity.[8] This happens due to the ectopic secretion of IGF by the tumourcells and always resolve following resection of the tumor.[1]

A CT scan is a useful diagnostic method that can identify the localization and size of the lesion and helps to plan the surgery.

Usually SFTP are a firm and well-circumscribed mass attached by a pedicle, as in the current case.

Complete surgical resection is the procedure of choice for all the SFTP and the only procedure that offers the cure. Given the relatively indolent nature of the tumor, radiation therapy is not currently recommended after resection and chemotherapy have shown low or questionable response rate.[9] Long-term *follow-up* is necessary, even for localized benign SFTP, because of the possibility of recurrence.

In conclusion, this case describes a rare giant SFTP, detected in the context of persistent hypoglycemia that was successfully treated by surgical resection. As expected, an immediate resolution of the hypoglycemia occurred. Given the nature of the tumor a long-term *follow-up* of the patient is necessary for detection of any possible recurrence, though the post-operative period was uneventful and at 1-month follow-up the patient was clinically recovered.

REFERENCES

1. Fengwei T, Yalong W, Shugeng G, Qi X, Juwei M, Yousheng M, et al. Solitary fibrous tumors of the pleura: A single center. Thoracic Cancer 9 (2018) 1763–1769.
2- Furukawa N, Hansky B, Niedermayer J, Gummert J, Renner A. A silent gigantic fibrous tumor of the pleura: case report. J CardiothoracSurg 2011;6:122–6.
3- Walid AA. Solitary fibrous tumors of the pleura. Eur J CardiothoracSurg 2012;41:587–97.
4- Perna V, Rivas F, Morera R, Saumench J, Ramos R, Macia I, et al. Localized (solitary) fibrous tumors of the pleura: an analysis of 15 patients, Int. J. Surg. 6 (2008) 298–301.
5- Raafat E, Karunasiri D, Kamangar N. Solitary fibrous tumour of the pleura presenting as a giant intrathoracic mass, Case report. BMJ Case Rep 2017. doi:10.1136/bcr-2017-220695.
6- Davanzo B, Emerson R, Lisy M, Koniaris L, Kays J. Solitary fibrous tumor. TranslGastroenterolHepatol 2018;3:94.
7- Perrot M, Fischer S, Bründler M, Sekine Y, Keshavjee S. Solitary fibrous tumors of the pleura, Ann. Thorac. Surg. 74 (2002) 285–293.
8- Perrot M, Fischer S, BruÃàndler A, Sekine Y, Keshavjee S. Solitary fibrous tumors of the pleura, Ann. Thorac. Surg. 74 (2002) 285-293.
9- Groot JW, Rikhof B, Doorn J, Bilo HJ, Alleman MA, Honkoop AH, Graaf WT. Non-islet cell tumor-induced hypoglycaemia: a review of the literature including two new cases. EndocrRelat Cancer, 2007, 14(4):979–993.