Case report

Hypoglycaemia associated with a solitary fibrous tumour of the pleura

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Abstract

We report the interesting case of an elderly woman who presented with hypoglycaemic episodes and weight loss. She was found to have a solitary fibrous tumour weighing more than 1.7 kg arising from the diaphragmatic pleura, which had been producing insulin-like growth factor II. After surgical removal of this well-encapsulated, pedunculated tumour her hypoglycaemia resolved and she returned to normal both clinically and biochemically. © 1999 Elsevier Science B.V. All rights reserved.

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1. Introduction

Solitary fibrous tumours are rare spindle cell neoplasms with diverse clinical and pathologic features. Although generally associated with the pleura (it was previously called fibrous mesothelioma) it is seen arising from the peritoneum and in somatic soft tissues such as the breast, thigh and buttock [1]. Solitary fibrous tumours have been reported in many unusual sites throughout the body such as the orbit, nasal cavity [2], meninges [3], parotid and adrenal glands. They are often benign but can be malignant especially if they grow to a large size [1].

We report a case of a patient with a large solitary fibrous tumour of the pleura, which had begun to secrete insulin-like growth factor II (IGF II) and produce symptoms of hypoglycaemia.

2. Case study

A 76-year-old female non-smoker was referred to us for bronchoscopy and thoracotomy from a district general hospital. She had presented to their clinic 3 months earlier with night sweats, fatigue, 5 kg weight loss and episodes of unsteady gait, diplopia and dysarthria lasting a few hours. CT brain and carotid Doppler examinations had been negative.

On examination she had nail clubbing, with distended anterior chest wall veins, a dull left base and everted umbilicus.

Investigations had revealed an apparently elevated left hemidiaphragm on chest radiograph with mediastinal shift to the right, worsening anaemia (9.3 g/dl) and a high ESR. A previous bronchoscopy had been normal. On admission for investigation she was found to have profound hypoglycaemic episodes every night and a dextrose infusion was started.

A CT scan of the thorax and abdomen showed a large mixed attenuation mass in the lower left hemithorax measuring 16 × 15 × 12 cms. No other abnormality was seen. A CT guided biopsy was obtained which was reported as 'certainly malignant, possibly sarcoma'. Her biochemistry revealed serum glucose of 1.1 mol/l, non-detectable levels of insulin, IGF I levels of 10.2 nmol/l and IGF II levels of 142.8 nmol. She was referred to our cardiothoracic unit with a diagnosis of an IGF II secreting tumour.

At surgery a large smooth encapsulated tumour was found in the left hemithorax (Fig. 1). It arose from the diaphragm by a narrow stalk and in two localised area was attached to (but did not appear to invade) the adjacent lung and pericardium. It was very vascular with dilated surface veins. It was removed through a left anterolateral thor-
acotomy. There were no enlarged lymph nodes and the rest of the lung and hilum appeared normal.

The histopathology reported a smooth fibrous tumour of the pleura weighing 1715 g and measuring 20 × 18.5 × 12.5 cm. There were foci of necrosis, hypercellularity and increased mitotic activity within the tumour. Immunohistochemical staining with CD-34 (which helps to differentiate solitary fibrous tumour from mesothelioma [4]) was positive.

The patient made a good recovery and had no further hypoglycaemic episodes. Her repeat biochemistry on the third postoperative day showed serum glucose of 7 mol. Her insulin levels had risen to 31 munits/l. IGF II had fallen to 45.3 nmol/l and IGF I was 9.6 nmol/l. Ten months after her operation she was fit and well. Her clubbed fingers were returning to normal and her anterior chest wall veins were no longer evident.

3. Discussion

Although solitary fibrous tumours have been well described in the thorax, we were interested to find one of these tumours secreting IGF II and which give rise to symptomatic hypoglycaemia.

Non-islet cell tumour hypoglycaemia (NICTH) is a paraneoplastic syndrome occurring in patients harbouring large, slow growing tumours [5]. It has been described in association with sarcomas, hepatomas and other mesodermal tumours. Diagnosis is based on clinical suspicion of unexplained hypoglycaemia and serum biochemistry. IGF II is one of the insulin-like peptides commonly thought to be responsible for NICTH, although it is not always easy to measure this rise in IGF II directly. IGF II may be produced by the tumour, or it may be produced as a larger precursor (big IGF II) by the tumour. Alternatively another tumour could stimulate the production of IGF II by the liver. Insulin and IGF I can be suppressed by IGF II, so an abnormal IGF I:IGF II ratio may indicate an IGF II secreting tumour. In our patient the IGF II levels were high and the true insulin was suppressed preoperatively, and both these returned to normal levels once the tumour was removed.

Surgical removal of the IGF II secreting tumour is the treatment of choice if possible, especially as the tumours are more likely to undergo malignant change if they grow to a large size. While awaiting surgery or in inoperable cases, growth hormone administration and chemotherapy have been shown to alleviate hypoglycaemic symptoms in NICTH.

References

[1] Nielsen GP, O’Connel JX, Dickerson GR, Rosenberg AE. Solitary


