Good's syndrome with primary intrapulmonary thymoma

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Hypogammaglobulinaemia associated with thymoma is known as Good's syndrome. Whilst most thymomas present as an anterior mediastinal mass they seldom occur within the lung parenchyma.

CASE HISTORY

A woman aged 72 had for 10 years experienced recurrent chest infections and left-sided pleuritic chest pain. She was now breathless and had a temperature of 39°C. On auscultation of the chest she had coarse bilateral inspiratory crackles and diminished breath sounds at the left base. A chest radiograph showed extensive shadowing in the lower half of the left hemithorax. Sputum culture yielded a heavy growth of *Pseudomonas aeruginosa*. Her white cell count was raised at $14.1 \times 10^9/L$ and C-reactive protein was 515 mg/L. Blood urea, electrolytes and liver function tests were normal. Serum immunoglobulin concentrations were low with IgG 1.2 g/L (normal 7–16), IgA 0.47 g/L (0.7–4) and IgM <0.05 g/L (0.4–2.3). Thoracic CT revealed a 12.8 cm mass in the left lower lobe with associated pleural effusion (Figure 1a). There was no evidence of an anterior mediastinal mass. A needle biopsy of the mass showed typical mixed cortical and medullary pattern thymoma. It was felt that the immune deficiencies identified were sufficient to explain her clinical condition, so more complex tests of immune status, such as B cell response to protein (tetanus) and polysaccharide (pneumococcal polysaccharide) antigens, were not performed; nor did we assess antibody responses to test immunizations.

The patient was treated with intravenous antibiotics and started on immunoglobulin replacement therapy. Surgical resection of the thymoma was considered but her general condition and lung function were poor. She was therefore offered a course of radiotherapy and received 2000 cGy in

Figure 1 CT scans of thorax before and after radiotherapy
(a) First scan showing large lobulated mass in left lower lobe with associated pleural effusion; (b) the mass has become smaller and has changed in consistency. There is intravenous contrast in the great vessels

five daily fractions. Repeat CT eight weeks later showed that the tumour had halved in volume and was more liquid in consistency (Figure 1b).

COMMENT

Thymomas, which have an incidence of 0.15 per 100 000,¹ can be associated with various clinical syndromes including myasthenia gravis, red cell aplasia, pancytopenia, collagen vascular disease and endocrinopathies.² The parathymic syndrome of hypogammaglobulinaemia known as Good's syndrome² is characterized by susceptibility to infection by encapsulated bacteria, viruses and fungi; chronic bronchial sepsis is commonly a feature. In contrast to antibody deficiency disorders such as common variable immunodeficiency and X-linked agammaglobulinaemia Good's syndrome includes defective T-cell function—hence susceptibility to cytomegalovirus, herpes simplex, varicella zoster, HHV-8 and other virus infections. The proportion of thymomas associated with hypogammaglobulinaemia has been estimated as 6–11%.^{3,4} Because of the T-cell defect, the use of live vaccines in patients with Good's syndrome

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poses a significant risk. For example, a survey of complications associated with yellow fever immunization showed that 4 out of 23 vaccinees who developed vaccine-associated viscerotropic disease had an underlying thymoma (and, we presume, Good's syndrome).⁵ Most thymomas are slow growing with a tendency to recur locally; they seldom metastasize and can usually be cured by surgical resection.⁶ Primary intrapulmonary thymoma without an associated mediastinal component, as seen here, is exceptionally rare.⁷ These tumours arise from ectopic embryonic tissue. As far as we are aware, this is the first reported case of hypogammaglobulinaemia associated with such a tumour.

REFERENCES

- Engels EA, Pfeiffer RM. Malignant thymoma in the United States: demographic patterns in incidence and associations with subsequent malignancies. *Int J Cancer* 2003;**105**:546–51
- Kelleher P, Misbah SA. What is Good's syndrome? Immunological abnormalities in patients with thymoma. *J Clin Pathol* 2003;**56**:12–15
- Souadlian JV, Enriquez P, Silverstein MN, *et al.* The spectrum of diseases associated with thymoma. *Arch Intern Med* 1974;**134**:374
- Rosenow EC, Hurley BT. Disorders of the thymus. *Arch Intern Med* 1984;**144**:763–72
- Eidex RB, for the Yellow Fever Vaccine Safety Working Group. History of thymoma and yellow fever vaccination. *Lancet* 2004;**364**:936
- Maggi G, Casadio C, Cavallo A, *et al.* Thymoma: results for 241 operated cases. *Ann Thorac Surg* 1991;**51**:152–6
- Srivastava A, Padilla O, Alroy J, *et al.* Primary intrapulmonary spindle cell thymoma with marked granulomatous reaction: report of a case with review of the literature. *Int J Surg Pathol* 2003;**11**:353–6

Spontaneous bilateral tubal ectopic pregnancy

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Ectopic pregnancy is still an important cause of maternal mortality. Bilateral tubal ectopic pregnancy is very rare, and is usually the result of an assisted reproduction technique.

CASE HISTORY

A woman aged 26 was admitted after experiencing severe abdominal pain. She had been amenorrhoeic for six weeks

and a urine pregnancy test was positive. She and her partner had been trying for conception and this had been achieved spontaneously. There were no risk factors for ectopic pregnancy in her medical history. The pain had been present for five days and had not been associated with any vaginal discharge or bleeding. A sudden increase in pain had prompted her self-referral.

She was mildly tender in the right iliac fossa but on pelvic examination there was no cervical excitation or adnexal tenderness. An early-pregnancy ultrasound scan was requested. After 24 hours on the ward she alerted the staff to a sudden increase in her abdominal pain, and abdominal examination now revealed generalized tenderness with rebound and guarding. At diagnostic laparoscopy, extensive blood was seen within the peritoneal cavity. Neither fallopian tube was clearly visible so a ruptured ectopic pregnancy was diagnosed. Mini-laparotomy, by Pfannenstiel's incision, disclosed a large ruptured ectopic pregnancy with destruction of the left fallopian tube. There also appeared to be a small unruptured ectopic pregnancy in the right fallopian tube. Left salpingectomy and right salpingotomy were performed and histological examination confirmed synchronous bilateral ectopic pregnancy. The β -human chorionic gonadotropin, which had been raised preoperatively, became normal—confirming complete removal of the pregnancies. The patient was counselled on her high risk of ectopic pregnancy and advised to attend for an early ultrasound scan in any future pregnancies.

COMMENT

In the UK about 11 in 1000 pregnancies are ectopic.¹ The most recent Confidential Enquiry into Maternal Deaths, reports 11 deaths from this cause in the past triennium; the number has been rising since 1991–1993. Synchronous bilateral ectopic pregnancy is very rare, and in most cases results from assisted reproduction techniques. The incidence is thought to be somewhere between 1 in 125 and 1 in 1580 extrauterine pregnancies.² Of the handful of reported cases of spontaneous bilateral ectopics, one came from our own hospital.³

Comprehensive clinical guidelines for the treatment of ectopic pregnancy have been published by the Royal College of Obstetricians and Gynaecologists.⁴ Because of its rarity, synchronous ectopic pregnancy is not covered, but the principles of treatment can still be applied. Laparoscopic surgical treatment is preferred to open procedures, because the patient recovers more quickly and subsequent rates of intrauterine and ectopic pregnancy are similar.⁵ Our patient, because of her acute symptoms, was not suitable for either laparoscopic surgery or medical management with methotrexate. At the time of surgery, examination of the contralateral tube governs treatment. In the present case the left tube had been destroyed, so salpingectomy

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