Wegener's granulomatosis presenting as a pleural effusion

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Notes
Lesson of the week

Wegener's granulomatosis presenting as a pleural effusion

Adrian G Blundell, Simon Roe

Wegener’s granulomatosis is one of the pauci-immune small vessel vasculitides. It classically presents with the triad of upper and lower respiratory tract granulomas and necrotising focal segmental glomerulonephritis. It is associated with the presence in the serum of autoantibodies against components of neutrophil cytoplasm—antineutrophil cytoplasmic autoantibodies (ANCA). The illness can develop at any age but is more common in their 50s and 60s and in men. The incidence of vasculitis is increasing with about 10–20 people per million affected. We present a case that in retrospect had many clues at the initial time of admission, but it took five months and six different hospital teams to make the diagnosis.

Case report

A 64 year old woman, who had had breast carcinoma that had been treated with wide local excision and radiotherapy six years previously, was admitted to her local hospital at the end of March 2001. She presented with a two week history of an influenza-like illness, including a blocked nose and right ear, dry cough, and intermittent sweats. She was feverish and had a left pleural effusion, which was confirmed radiologically. Her inflammatory markers were raised (total white cell count 13.1x10^9/L, C reactive protein 322 mg/l (normal range < 5 mg/l)). Treated was started with intravenous antibiotics for a possible empyema. Despite three different antibiotics, her symptoms failed to improve over the next two weeks. Ultrasound scanning of the chest confirmed a fluid collection, but several attempts at aspiration and drainage were unsuccessful. She was transferred to a teaching hospital under the care of the cardiothoracic surgeons, but she became increasingly breathless and developed atrial tachyarrhythmias and presumed acute pulmonary oedema. At this time there was evidence of renal impairment (serum creatinine concentration 130 μmol/l) and she was deemed unfit for surgery, so she was transferred back to the referring centre.

A computed tomogram of the thorax showed bilateral pleural effusions, and transthoracic echocardiography showed a pericardial effusion. Owing to persisting fever and raised inflammatory markers, her antibiotic regimen was again altered and she was transferred to a different tertiary centre for a respiratory opinion. Soon after admission she developed respiratory failure and needed intubation and ventilation. She was found to have no empyema. She recovery slowly and was transferred back to her original team at the beginning of June without a uniform diagnosis. Repeat echocardiography at this time showed resolution of the pericardial effusion and her creatinine concentration was 124 μmol/l.

At the end of June she was transferred to a community hospital for rehabilitation. Over the next month she had recurrent episodes of syncope and bradycardia. Her serum potassium concentration was persistently raised and her renal function deteriorated markedly (creatinine concentration 618 μmol/l). She had a cardiac arrest, from which she was successfully resuscitated. She was subsequently transferred to the intensive care unit of our hospital, where she needed ventilation support and continuous venovenous haemofiltration for acute renal failure. She was found to be strongly seropositive for cytoplasmic ANCA (C3 or C4) (titre > 2560 units) for antibodies to proteinase 3 with enzyme linked immunosorbent assay (ELISA), and was treated with pulsed intravenous methylprednisolone, followed by oral prednisolone and cyclophosphamide. Two weeks later she developed pulmonary haemorrhage and needed reintubation and treatment with plasma exchange for two weeks. She improved slowly over the next few weeks and was discharged at the end of October on a combination of prednisolone and azathioprine; her creatinine concentration at this time was 200 μmol/l.

Discussion

Vasculitis can be categorised by the size of vessel affected (small, medium, or large). Wegener’s granulomatosis is a small vessel vasculitis classically involving the upper and lower respiratory tracts and kidneys. A limited form

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The greater man, the greater courtesy

It was another busy admission day. I was working as a surgical senior house officer in Calcutta Medical College and was part of an eight member team, coping with the usual 30 or so admissions. It was the middle of the night when I saw a 9 year old boy who had just been brought to us by his father. The boy seemed to have developed generalised peritonitis and was not looking good.

"Why did you bring him so late?" I asked rather abruptly.

The boy's father, a softly spoken man, tried to explain his difficult social circumstances, lack of transport at this time, etc. Then he suddenly paused and said, "I understand his condition is not good. Please do whatever you can." I was taken aback by the polite resoluteness of the man.

We started the treatment. The child needed a laparotomy for what turned out to be a perforated small bowel due to typhoid. By the time I left next morning, his condition was slightly better but still far from good.

The next day I went away on holiday for 10 days. On my return to Calcutta, I faced a tremendous downpour of rain as I left Howrah railway station. And, guess what, I had just missed the 44 bus that would take me close to my college. Howrah railway station. And, guess what, I had just missed the 44 to Calcutta, I faced a tremendous downpour of rain as I left still far from good.

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As I was cursing my luck, something strange happened. The bus reversed, and a friendly voice addressed me: "Doctor-Sahib, please get up quickly." I thanked the driver for his kindness, but it was only after I had left the bus that I realised that the driver was the father of the boy whom I met in the hospital.

The next day, I proudly boasted to my colleagues: "Has a bus ever reversed just for you? It happened to me yesterday!"

After sharing my story, I asked about the boy. "He died after a few days," came the reply.

I was humbled twice in two weeks. Firstly, by the determination of a man who kept his head cool at a time when his son was most unwell. Secondly, when he unservicedly showed his appreciation by picking me up in bad weather, perhaps even risking his job.

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