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Scientific Letters

Pyopneumothorax Due to Wegener's Disease

Pioneumotórax por enfermedad de Wegener

Wegener's granulomatosis is an autoimmune disease that may present as necrotising granulomatosis, vasculitis of the upper and lower respiratory tract, necrotising glomerulonephritis and diffuse vasculitis, mainly affecting the lungs, airway, and kidney.[1] When the lung is affected, the clinical manifestations are nonspecific and include coughing, dyspnoea, and haemoptysis. Plain chest radiology usually shows areas of consolidation or single or multiple nodules, which are asymptomatic in a third of the affected patients. Cavitation of these nodules is common because of central necrosis. When this phenomenon affects the visceral pleura, a bronchopleural fistula occurs and pneumothorax or pyopneumothorax can appear. This complication is extremely rare and only 9 cases have been reported in literature worldwide.[2,3] We present the case of a patient with pyopneumothorax due to Wegener's granulomatosis and its surgical resolution, treated in the Thoracic Surgery Department of our hospital.

We report the case of a 35-year-old male patient who was admitted to the Internal Medicine Department for symptoms of fever associated with coughing and purulent sputum. A chest X-ray showed a right basal opacity and the initial diagnosis was community-acquired pneumonia. Forty eight hours after admission, the patient presented clinical deterioration with poor ventilatory mechanics, and required transfer to the intensive care unit and mechanical ventilation. A twenty point fall in haematocrit levels and associated kidney failure were found. C-ANCA and proteinase 3 were requested, which were positive, and the presumptive diagnosis of Wegener's granulomatosis was made. Treatment was started with 500 mg of cyclophosphamide and plasmapheresis was carried out. The cough and mucopurulent sputum persisted, and chest axial CT scan was performed, which displayed predominantly right bilateral basilar infiltrates (Fig. 1a). Antibiotic treatment with vancomycin, meropenem, and colistin was started. Plasmapheresis was recommenced and due to a favourable response the patient was transferred to the general medical ward.

The fever persisted, and therefore treatment was rotated to colistin and imipenem for 14 days due to the isolation of *Klebsiella pneumoniae* and *Pseudomonas aeruginosa* in the bronchoalveolar lavage. The clinical course was favourable and the patient was discharged with 60 mg/day of prednisone. At home, he started feeling a penetrative rib pain associated with dyspnoea, and went to the Emergency Department. In the chest X-ray, a right grade II pneumothorax was observed and pleural drainage was performed. A subcutaneous emphysema appeared and a persistent bronchopleural fistula with air-leak and purulent sputum. Axial computed tomography was requested, and showed pyopneumothorax and a lung mass abscess and cavity in the right upper lobe (Fig. 1b). Given the persistence of purulent sputum and lung expansion failure, surgical treatment was performed. The approach was a posterolateral thoracotomy and an abscessed and fistulised pleural granuloma was identified, of 11 cm in diameter, located in the right upper lobe (Fig. 2a). A complete resection was performed with mechanical suture (Fig. 2b), completing the surgery with pulmonary decortication (Fig. 2c). Following the operation, he was immediately transferred to the intermediate care unit for 48 h, and was discharged 6 days after surgery with a favourable postoperative course. The histopathology report of the surgical specimen (Fig. 2d) confirmed the diagnosis of Wegener's granulomatosis.

Pneumothorax or pyopneumothorax is an extremely rare manifestation of Wegener's granulomatosis (WG), with only 9 cases reported in literature worldwide.[2] There was predominance in males, as in our case, and an average age of 48.9 years was observed. On admission, pneumothorax was present in 6 of the 9 patients, pyopneumothorax in 3, and associated empyema in 1.[3,4]

Lung nodules represent over 70% of lung lesions in patients with Wegener's granulomatosis.[3] These are usually subpleural and can cavitate in nearly 50% of cases. Pleural involvement, however, is infrequent, being less than 10%. The lungs are the most common location of nodules in this disease and the contact between these nodules and the pleura is frequent,[3] which coincides with our case.

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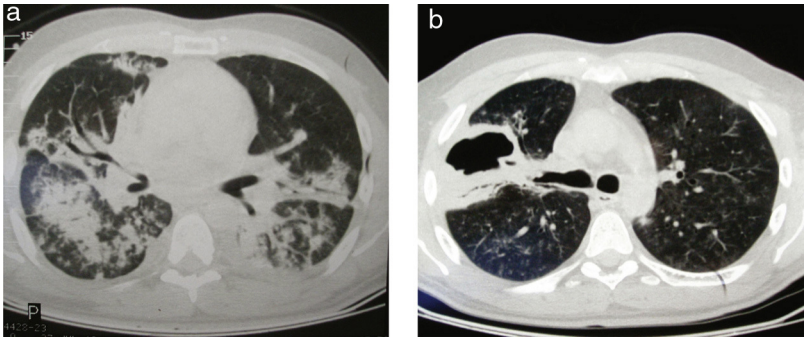

Fig. 1 – (a) Multiple poorly defined areas of bilateral consolidation with a right predominance, associated with pseudo-nodular images and congestion of both lung hilum. (b) Bilateral, subpleural micronodules and nodule cavitated with hydro-aerial level in the right upper lobe.

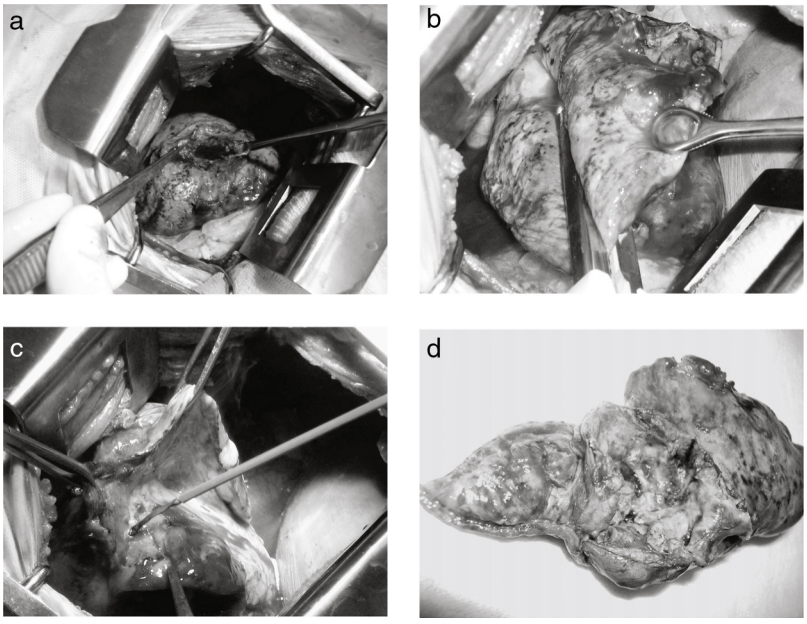

Fig. 2 – (a) Cavitated granuloma in the right upper lobe. (b) Sublobar resection of the right upper lobe with mechanical suture. (c) Decortication. (d) Surgical specimen.

On admission, our patient had small subpleural nodules and a partially cavitated solid lesion in the right lung; this is the most common radiographic image of Wegener's granulomatosis.[3]

Bronchopleural fistula and necrotic granulomatous lesions with expansion to the pleura have been previously reported.[2] Lung expansion failure, which did not improve with continuous aspiration, has also been reported and is associated with infection and pyopneumothorax,[1] *which was the case for our patient.*

Pleural drainage was performed in 5 of the 9 cases and lobectomy in one. It was decided not to conduct surgery in 2 of the 9 cases (22.2%). In our case, the initial surgery was pleural drainage for pyopneumothorax. Its failure led us to perform a posterolateral thoracotomy on the patient and resect the lesion.

Postoperative mortality associated with patients admitted with pyopneumothorax was observed in 66.6% (2 out of 3 patients).[3]

Immunosuppressive therapy, particularly with cyclophosphamide, has undoubtedly improved survival rates.[1]

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Gallstone Ileus in a Patient With Crohn's Disease

Íleo biliar en paciente con enfermedad de Crohn

The link between Crohn's disease (CD) and gallstone ileus has been described previously in the literature.[1–5] In these cases a biliary-enteric fistula is usually observed, and instead of the calculus impacting in the ileocecal valve, it does so in the segment of small intestine that is stenosed due to CD.

Ulcerative colitis (UC) and CD are 2 distinct illnesses, although both are considered chronic inflammatory bowel disease. One of the main differences between them is that CD can affect any part of the gastrointestinal tract. Inflammation of the terminal ileum is the most common location, in comparison to UC, where only the large bowel is affected.[6] We present the case of an elderly patient with a prior diagnosis of ulcerative colitis who was urgently operated on for gallstone ileus. The diagnosis was confirmed preoperatively by an abdominal CT scan.

An 84-year-old male with a personal history of type II diabetes mellitus, chronic bronchitis and a 60 year diagnosis of UC treated with aminosalycilates, presented to the Emergency Department complaining of abdominal pain, vomiting, and abdominal distension of 48 h duration. In the abdominal CT scan, pneumobilia was observed (Fig. 1), and distension of the small bowel up to the ileum where a calculus had impacted (Fig. 2) in a stenotic segment of terminal ileum with wall thickening that extended to the ileocecal valve. With the diagnosis of a gallstone ileus and terminal ileitis, an emergency laparotomy was performed corroborating the existence of an ileitis with a calculus of 3 cm impacted in the thickened bowel and distension of the rest of the small bowel. The segment of the affected distal ileum measured around 40 cm and had the typical macroscopic appearance of CD with thickening of the mesentery and its growth toward the antimesenteric margin. No signs of active inflammation were observed in the large intestine. An enterotomy was performed on the healthy proximal bowel and the calculus was extracted. The patient did not refer any symptoms of recent bowel obstruction and therefore, given the long period without any prior resections, it was decided to preserve the affected ileum and no action was performed on the gallbladder.

At present, it is very uncommon for a patient with CD to be wrongly diagnosed with UC, because radiology techniques, histological studies, and endoscopic procedures have advanced considerably. However, 5% of patients present characteristics of both CD and UC and cannot be classified as either type. In these cases the term non-classified inflammatory bowel disease is used.[6]

At the time of surgery, our patient displayed terminal ileitis with microscopic characteristics typical of CD, in spite of his previous diagnosis of UC. The treatment he followed with oral aminosalycilates could be effective both for CD and UC and did not help to discriminate between the diseases. In the abdominal CT scan, which was performed as an emergency procedure to confirm the diagnosis of small bowel obstruction, the existence of CD with gallstone ileus was considered for the first time.

The manifestation of a gallstone ileus in patients with CD[1–5] is exceptional. Cases of small bowel obstruction caused by foreign bodies impacting on stenosed bowel segments in patients with CD have been described.[7–9] They usually occur in elderly individuals who have had CD for a long time and in some

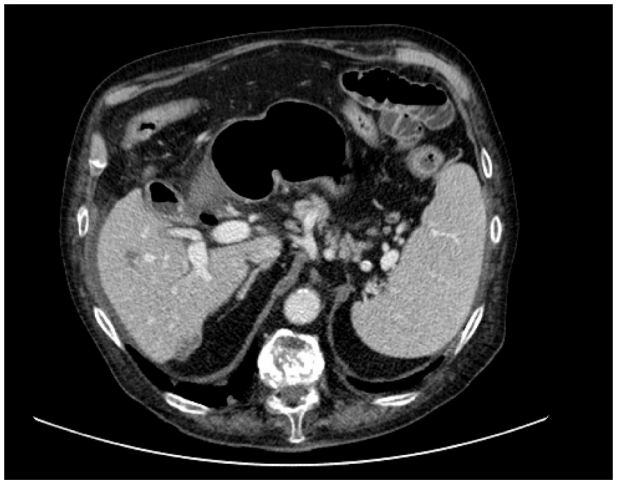

Fig. 1 – Pneumobilia can be observed in the image with air inside the gall bladder.

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