Pulmonary aneurysms in Behcet disease completely resolved after medical therapy

Behcet s disease (BD) is a systemic vaculitis of unknown origin, involving arteries and more frequently the venous system [1]. Pulmonary artery (PA), is the second most affected vessel after the aorta [2]. The reported prevalence of pulmonary manifestations ranges from 1 to 7.7%. The prognosis of Behcet s patients with pulmonary artery aneurysm (PAA) is serious, since fatal rupture of PAA is still the major cause of death in this disease. PAA requires urgent management to prevent complications. Several treatments have been proposed including steroids, immunosuppressive drugs, embolisation and surgery [3].

Herein, we report a patient with Behcet s disease with multiple pulmonary artery aneurysms, which completely resolved after a combined treatment with corticosteroids and azathioprine.

Case report

A 36-years old man, with a personal history of BD since 1992, was admitted to the department of internal medicine for chest pain and hemoptysis.

BD was diagnosed on the basis of: recurrent orogenital ulcers, folliculitis, deep venous thrombosis of the lower right leg and positive pathergy test. His previous treatment included a long-term colchicin, oral corticosteroids 10mg/day and temporary anticoagulation.

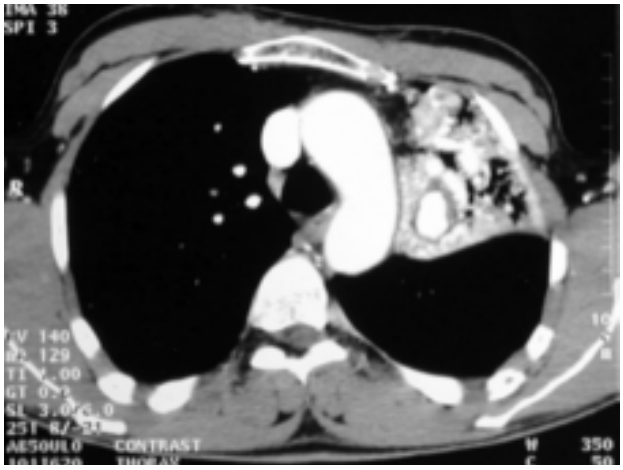
On January 2003, he complained about fatigue, dry irritating cough, chest pain, shortness of breath, slight but recurrent hemoptysis and concomitant oral ulcer. He denied any fever, chills, pain or leg swelling, neurological symptoms and visual troubles. He did not discontinue his treatment.

On physical examination he was tense, alert and oriented to time, place and person. His blood pressure was about 120/80 mm Hg with tachycardia 120bpm. The cardiac auscultation didn't reveal any murmur, rub or gallop. No signs of heart failure were present. He had tachypnea without nasal flaring, intercostals, sternal retractions or cyanosis. Crackles were audible at the bases of the right lung field. Neurological and vascular examinations were within normal.

The laboratory investigations indicated: a recent normocytic normochromic anemia: hemoglobin (Hb) level about 11g/dl (the last RBC showed 16g/dl of Hb). The differential count was normal. The erythrocyte sedimentation rate ESR and C-reactive protein CRP were: 100mm and 35mg/l receptively. Creatinine level and liver function tests were within normal range. The arterial blood gas showed hypoxia: 75 mmHg, hypocapnia: 30 mmHg and respiratory alkalosis. The ECG revealed sinus tachycardia, right axis deviation, right bundle-branch block and PQ segment depression in DI, DII, DIII and aVF leads. No ECG microvoltage or ST segment, T waves changes were noticed.

The venous Doppler ultrasound didn't reveal any signs of phlebitis in the legs. The chest X-Ray showed a left hilar enlargement. The echocardiography data were consistent with pericarditis without cardiac thrombosis. Computed tomography scanning with contrast enhancement showed an aneurysm of approximately 6.4 cm × 6.7 cm in size at the left upper lobe, with enhancement of the patent lumen and a circumferential thrombus. Anterior to the aneurysm a parenchymal consolidation was noted (Figure 1).

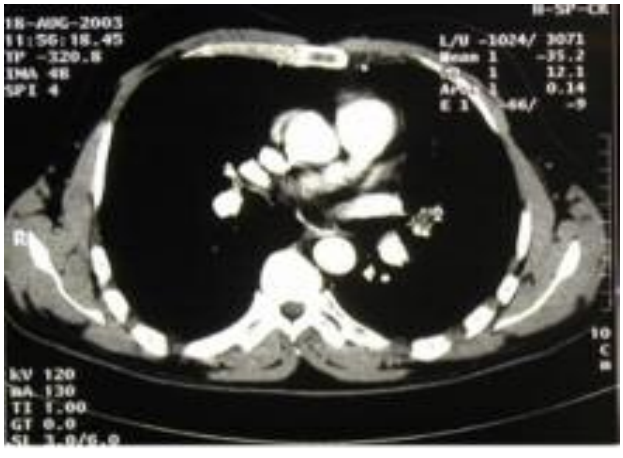
Figure 1 : Computed tomography scanning with contrast enhancement shows an aneurysm approximately 6.4 cm × 6.7 cm in size at the posterior basal segment of the right lower lobe, with enhancement of the patent lumen and a circumferential thrombus



Additionally there were two aneurysms located in the right middle lobe and right lower lobe pulmonary arteries that were measured at 1.5 x 1.5 cm and 2 x 2 cm in size, respectively. The diagnosis of PAA complicating BD was established. Ophthalmic examination was normal. Consistently, the patient received three 1g methylprednisolone boli, monthly intravenous cyclophosphamid, followed by oral prednisolone 1mg/kg/day. Colchicin therapy (1 mg/day) was continued.

A repeat CT scan performed 18 days later revealed a complete thrombosis of the aneurysm located in the left upper lobe (figure 2). Transcatheter embolization of the other PAA was proposed without any success.

Figure 2 : Computed tomography scanning after embolization showing a marked reduction in the size of the aneurysm



Six months later, follow-up computed tomography scanning with contrast was performed and showed persistent small aneurysms. Azathioprine was switched to cyclophosphamide. After a two-year follow-up period, the patient returned to our clinic asymptomatic and serial control scans show no aneurysm formation.

Conclusion

Pulmonary artery aneurysms (PAA) are reported to indicate poor prognosis and high mortality. CT of the chest and angiography are the most common diagnostic procedures used in the diagnosis or evaluation of PAA. Immunosuppression is the main therapy for the treatment of PAA in BD and should be associated to embolisation. Anti-TNF factors are indicated in case of unresponsiveness to immunosuppressive drugs.

References

1. Celenk C, Celenk P, Akan H, Basoglu A. Pulmonary artery aneurysms due to Behcet's disease: MR imaging and digital subtraction angiography findings. AJR Am J Roentgenol. 1999;172:844–5.
2. Sraieb T, Ben Romdhane N, Longo S, Manna J, Louzir B, Othmani S. Behcet disease and arterial aneurysms: 3 cases report. 1999; 20:517-21.
3. Harmurydan Y, Yurdakul S, Moral F, Numann F, Tuzum H. Pulmonary arterial aneurysms in Behcet syndrome: a report of 24 cases. B J Rheumatol; 1994; 33:48-51.

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Urinary peritonitis caused by gangrenous cystitis

Gangrenous cystitis (GC) is an uncommon but it is life threatening condition that ranges in severity from necrosis of the mucosa and submucosa to necrosis of the entire bladder wall (1). With the widespread use of antibiotics, only sporadic cases have been reported in the recent literature (2). The aetiology of this condition is probably multifactorial and it is difficult to identify a unique cause (3,4). Presenting symptoms are usually non-specific, and accurate diagnosis may be extremely difficult (2,3,5,6). Upon suspicion, CTscan is very helpful in the establishment of correct diagnosis (2,5). As soon as the diagnosis is made, treatment should be early and aggressive. The removal of all necrotic tissues is mandatory (4,6).

Herein, we report on a rare case of urinary peritonitis secondary to GC, and support it with a brief survey of the literature on this topic.

Case report

A 36-year-old man was admitted to the emergency intensive care unit with a diagnosis of acute peritonitis. His past medical records did not indicate any major illness except chronic alcoholism.

During 3 days, the patient complained of severe abdominal pain, abdominal distension, nausea, vomiting, total hematuria and many episodes of urinary retention.

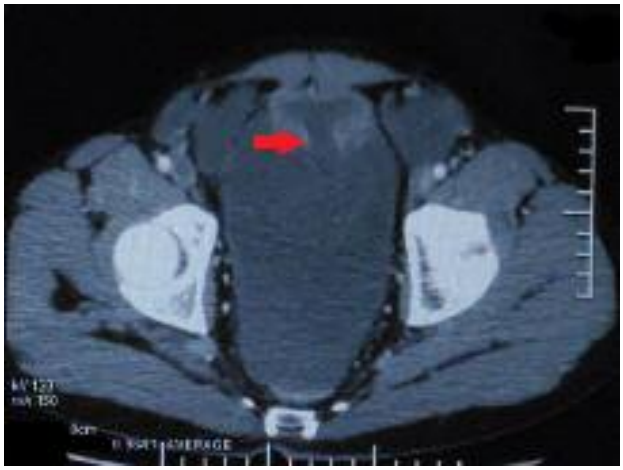
Physical examination revealed board-like stiffness and rigidity of the abdominal wall, with absence of bowel sounds. He was hypothermic (36,6°C). In addition, he has tachycardia (104 beats per min) and tachypnea (26 breaths per minute). An urethral catheter was inserted into the bladder. It drained residual bloody urine mixed with debris.

Blood analysis indicated high leukocytes count (18,000 wbc) with normal Hb (11.3g/dl) and platelets (467000/mm^3) levels. Hepatic and pancreatic blood analysis revealed high values of (AST/ ALT: 81/36 UI/L, amylasemia: 166 U/L, lipasemia > 10 x nl, CRP: 176.6 mg/l, prothrombin ratio: 79%, Quick's time: 35.2 s and INR: 1.16). Blood urea nitrogen was 2.67 g/l, creatinine was 103 mg/l and electrolytes values were also abnormal: Na/K: 123/6.4 meq/l. necessitating two hemodialysis sessions.

Urinary analysis showed pyuria, and culture was positive for *Escherichia coli*.

The abdominal X-ray showed multiple hydroaeric shadows in the small intestine, without free gas below the diaphragm. The computer tomography scan showed dilated small intestinal loops, without apparent obstruction of the passage of the bowel contents. Besides, bladder overdistension was confirmed with bladder rupture of about 12 mm in length (Figure 1).

Figure 1 : CT Scan: Bladder overdistension with bladder rupture (arrow)



Diagnosis of urinary peritonitis due bladder perforation secondary to bladder over distention then was suspected. The explorative laparotomy revealed generalized acute peritonitis, with 1100 ml serous exudate in the abdominal cavity, and dilated intestinal loops covered by multiple fibrous coatings. The detailed revision established marked, dark greenish, necrosis and perforation of the bladder wall, which was the main cause for the peritonitis. There was no extensive necrosis in the area of Retzius or in the retroperitoneal space. As only 4 cm of the bladder dome looked necrotic, an extensive