Huge pulmonary artery aneurysm

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Pulmonary artery aneurysms (PAAs) are uncommon entities. PAAs are caused mostly by trauma (often iatrogenic), infections and Behcet's disease (BD). Less common causes are pulmonary hypertension, congenital heart disease and neoplasm. BD is a multisystem disorder presenting with recurrent oral and genital ulcerations, as well as ocular involvement, and PAA is one of its rare complications. A case of huge PAA, in which the usual criteria for the clinical diagnosis of BD were present, is described. Transcatheter embolization resulted in clinical improvement.

Key Words: *Behcet's disease; Pulmonary artery aneurysms*

Pulmonary artery aneurysms (PAAs) are uncommon conditions that are most commonly caused by trauma (often iatrogenic), infections or Behcet's disease (BD). Less common causes are pulmonary hypertension, congenital heart disease and neoplasm (1). BD is a multisystem disorder presenting with recurrent oral and genital ulcerations, skin lesions and ocular involvement. It was first described by the Turkish dermatologist Hulusi Behcet in 1937 (2-4). The mean age at which BD occurs is 20 to 30 years. BD is most prevalent in the Mediterranean region, Middle East and Far East. Men are affected two to five times more often than women (2,5). BD involving the chest can manifest as a wide spectrum of abnormalities that include abnormalities of the vessel lumen and wall, lung parenchyma, pleura and mediastinal structure. PAA is a rare but serious complication of BD (5,6). We present one such interesting patient with BD who developed huge PAA and was treated successfully by using transcatheter embolization that resulted in clinical improvement.

CASE PRESENTATION

A 41-year-old Sudanese man presented to the emergency room of Assir Central Hospital, Abha, Saudi Arabia, with a three-month history of fever and pleuritic chest pain, and a 20 kg weight loss during the previous six months. There was no history of hemoptysis, productive cough, trauma or recent surgery. His medical history documented previous visits to another hospital during the past three years for recurrent painful mouth and genital ulcers that were associated with a decrease in vision of the right eye at least three times per year. He was told that the most likely cause of his symptoms was BD. At the time, he was not started on treatment for BD. A posteroanterior chest x-ray taken on the patient's initial presentation showed a right lower zone oval density with superior and inferior poles extended beyond the interposed rib (Figure 1). His family and

Volumineux anévrisme de l'artère pulmonaire

Les anévrismes de l'artère pulmonaire (AAP) sont peu fréquents et sont le plus souvent causés par un traumatisme (souvent iatrogène), par des infections et par la maladie de Behcet. Ses causes moins fréquentes sont l'hypertension pulmonaire, la maladie cardiaque congénitale et le cancer. La maladie de Behcet est un trouble plurisystémique qui se manifeste par des ulcérations orales et génitales récurrentes, de même que par une atteinte oculaire; et l'AAP est l'une de ses complications rares. Les auteurs décrivent ici un cas d'AAP volumineux en présence des critères diagnostiques cliniques habituellement associés à la maladie de Behcet. Une embolisation transcathéter a donné lieu à une amélioration clinique.

Figure 1) Posteroanterior chest x-ray showing a right lower zone oval density (arrow)

social history was not significant. Physical examination showed a gaunt man who was not in respiratory distress. Vital signs were within the normal range. Examination findings were unremarkable. Laboratory studies revealed a leukocyte count of $140 \times 10^9/L$, hemoglobin count of 156 g/L; urea and electrolytes within the normal range. His erythrocyte sedimentation rate was 24 mm/h. Immunoglobulin (Ig) assays revealed an IgG concentration of 9.89 g/L, IgM concentration of 2.418 g/L and an IgA concentration of 2.975 g/L. His electrocardiogram was normal. A posteroanterior chest x-ray (Figure 2A) and lateral

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Figure 2) (A) Posteroanterior and (B) lateral chest x-rays showing a homogenous, rounded and well-defined mass at the posterior of the right lower lobe (arrows)

Figure 3) Computed tomography scanning with contrast enhancement shows an aneurysm approximately 6.4 cm x 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 (arrow)

chest x-ray (Figure 2B) revealed a homogenous, rounded and well-defined mass within the right lower lobe. Computed tomography scanning with contrast enhancement showed an aneurysm of approximately 6.4 cm x 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 (Figure 3). Pulmonary artery angiography showed an aneurysm that was fed from the posterior basal branch of right pulmonary artery and apparent distal arterial beaded tortuosity (Figure 4). His clinical picture was consistent with PAA in BD. Consequently, the patient was started on steroids. To ensure safety for the patient against PAA complications, transcatheter embolization of the right lower pulmonary artery was performed using Guglielmi detachable coils (Boston Scientific, USA) with no complication noted (Figure 5). Three weeks later, follow-up computed tomography scanning with contrast was performed and showed a marked reduction in the size of the aneurysm (Figure 6). The patient showed clinical improvement and was discharged. After a two-year follow-up period, the patient returned to our clinic asymptomatic. A follow-up chest x-ray

Figure 4) Pulmonary artery angiography showing an aneurysm that is feeding from the posterior basal branch of the right pulmonary artery (arrow) and an apparent arterial beaded tortuosity lateral to the aneurysm (curved arrow)

Figure 5) Transcatheter embolization using Guglielmi detachable coils (arrow) (Boston Scientific, USA)

revealed a remnant of metallic coils at the base of the right lower lobe (Figure 7).

DISCUSSION

The diagnosis of BD is essentially a clinical one, based on the presence of multiple physical signs. Four major criteria have been proposed: recurrent mouth ulcerations, genital ulcerations, ocular inflammation and skin lesions (2,3). Vasculitis caused by BD can occur in three forms: venous occlusion and varix formation, arterial occlusion and/or pulseless disease, and arterial aneurysm formation (7). Venous system involvement is more common than arterial system involvement (8). PAAs secondary to BD are a rare and serious complication.

Figure 6) Computed tomography scanning after embolization showing a marked reduction in the size of the aneurysm (arrow)

PAA's are almost always associated with hemoptysis, which can be fatal. PAA's may be single or multiple, unilateral or bilateral, and true or false (5,6). PAA in BD follows a lethal sequence and must be managed urgently, especially once associated with hemoptysis. PAA's are located most frequently in the right lobar arteries, followed by the right and the left main pulmonary arteries (9). Chest x-rays are commonly used for the initial assessment of thoracic manifestations of BD (5). Computed tomography is most helpful in the diagnosis and follow-up of PAA in BD, and will give detailed information concerning the vessel lumen and wall, the lung parenchyma, pleura and mediastinal structures (5,9-11). Only a few cases have been treated successfully by transcatheter embolization. Embolization is the first line of treatment for massive hemoptysis in patients with BD (12). PAA is a poor prognostic factor in BD. The majority of patients with PAA associated with BD die within one year after onset of hemoptysis. Massive hemoptysis secondary to aneurysmal rupture is the usual cause of death in these cases (8,13).

CONCLUSION

BD is the most common cause of PAA secondary to vasculitis (5). Imaging such as computed tomography with contrast is useful in making a diagnosis and for follow-up. PAA in BD is a serious complication and should be managed urgently to prevent complications of PAA. Embolization is a new modality and a promising treatment option, and in our opinion, it should be the first invasive procedure performed when dealing with this potentially devastating abnormality.

Figure 7) Posteroanterior chest x-ray shows a remnant of metallic coils at the base of the right lower lobe (arrow)

REFERENCES

1. Neuyen ET, Silva CI, Seely JM, Chong S, Lee KS, Muller NL. Pulmonary artery aneurysms and pseudoaneurysms in adults: Findings at CT and radiography. *AJR Am J Roentgenol* 2007;188:W126-34.
2. Kasper D, Fauci A, Longo D, et al. *Harrison's Principles of Internal Medicine*, 16th edn. New York: McGraw-Hill, 2005:2014-15.
3. International Study Group for Behcet's Disease. Criteria for diagnosis of Behcet's disease. *Lancet* 1990;335:1078-80.
4. Behcet H, Uber R. Aphthos durch ein virus verursachte Geschwure am Mund, am Auge und an den Genitalien. *Dermatol Wochenschr* 1937;105:1152-7.
5. Hiller N, Lieberman S, Chajek-Shaul T, Bar-Ziv J, Shaham D. Thoracic manifestations of Behcet's disease at CT. *Radiographics* 2004;24:801-8.
6. Uzun O, Akpolat T, Erkan L. Pulmonary vasculitis in Behcet's disease: A cumulative analysis. *Chest* 2005;127:2243-53.
7. Numan F, Islak C, Berkmen T, et al. Behcet's disease: Pulmonary arterial involvement in 15 cases. *Radiology* 1994;192:465-8.
8. 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.
9. Tunaci M, Ozkorkmaz B, Tunaci A, Gul A, Engin G, Acunas B. CT findings of pulmonary artery aneurysms during treatment for behcet's disease. *AJR Am J Roentgenol* 1999;172:729-33.
10. Emad Y, Abdel-Razek N, Gheita T, el-Wakd M, el-Gohary T, Samadoni A. Multislice CT pulmonary findings in Behcet's disease (report of 16 cases). *Clin Rheumatol* 2007;26:879-84.
11. Zidi A, Ben Miled Mrad K, Hantous S, Nouira K, Mestiri I, Mrad S. Thoracic involvement in Behcet's vasculitis. *J Radiol* 2005;87:285-9.
12. Bozkurt A. K. Embolisation in Behcet's disease. *Thorax* 2002;57:469-70.
13. Hamuryudan V, Yurdacul S, Moral F. Pulmonary artery aneurysm in Behcet's disease: A report of 24 cases. *Br J Rheumatology* 1999;33:48-51.