

Churg–Strauss Syndrome Presenting with Endobronchial Masses

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Abstract

Churg–Strauss syndrome is a condition with unknown etiology and asthma, allergic rhinitis, eosinophilic infiltration of blood and tissues, and transient infiltration of the lungs. It occurs mostly in the 3rd–4th decades of life with an incidence of 2.4/1000000. Presentation frequently involves nodular lung infiltrations, infiltrations with cavity, ground-glass appearance, and alveolar opacity. However, endobronchial mass is an unexpected presentation. In the current case report, we present a 45-year-old male patient who was receiving asthma therapy for 5 years. In the last follow-up visit, we identified a mass in the right hilum on X-ray radiography and performed fiberoptic bronchoscopy. Pathologic examination of biopsy material verified the diagnosis of Churg–Strauss syndrome. Bronchial mass is an unexpected presentation of Churg–Strauss syndrome and pathologic examination is essential to distinguish it from pulmonary malignancies

Keywords: Bronchocentric granulomatosis, bronchoscopy, Churg–Strauss, endobronchial mass

INTRODUCTION

Churg–Strauss syndrome (CSS) is a vasculitis with unknown etiology and involves small and middle-sized vessels (1). It is characterized by chronic rhinosinusitis, asthma, and increased eosinophils in blood and tissues. Transient infiltrations occur in the lungs. Vasculitis is not a characteristic finding of the early periods of the disease. The involvement of lungs and skin is common. Cardiopulmonary, gastrointestinal, renal, and central nervous systems are also affected. The mortality and morbidity of the disease mostly depend on vasculitic involvement of extrapulmonary organs. Although the exact incidence is not known, it is estimated to be 2.4/1000 000.

CASE PRESENTATION

A 45-year-old male patient was admitted to hospital with complaints of cough, breathlessness, and discharge of green-colored sputum. The patient had been taking inhaled steroids twice a day and long-acting beta-mimetic drugs to treat asthma for 5 years. Family history was insignificant. He was a non-smoker and was not consuming alcohol. Physical examination revealed that his general condition was good. The respiratory rate was 17/min, heart rate was 90/min, and arterial blood pressure was 125/70 mmHg. Auscultation showed bilateral inspiratory and expiratory rhonchi. Lymphadenopathy was absent. Skin examination revealed a black spot on the right thumb lunula (Figure 1a); superficial veins on the right tibia were marked and ecchymotic (Figure 1b). There was another ecchymosis on the left deltoid posterolateral aspect. Laboratory findings showed urea: 21 mg/dL, creatinine: 0.7 mg/dL, C- reactive protein: 0.3 mg/dL, erythrocyte sedimentation rate: 15 mm/h. Liver function tests and amylase were within normal limits. The white blood cell count was 11000cells/mm³, blood eosinophils were 18% (1900cells/mm³), and total IgE was 2000 IU/mL. The plain X-rays of the chest showed non-homogenous density increase on the right hilum extending to anterior space (Figure 1c), and paranasal sinuses mucosal thickness of the left maxillae and polypoid lesions of right middle meatus (Figure 1d). There were homogenous infiltrations on the left medial peripheral regions. Chest CT showed a non-homogenous density increase of the right upper lobe anterior segment and left hilum extended (Figure 1e). A fiberoptic bronchoscopy was performed under local anesthesia. Two masses obstructing the left lower lobe medial subsegment bronchus (Figure 1f1) and right upper lobe anterior subsegment were observed (Figure 1f2). Pathologic examination of bronchoscopy material showed no malign cells. Instead, eosinophilic granulomatous infiltration was seen (Figure 1g1, 1g2). Fungal culture showed no



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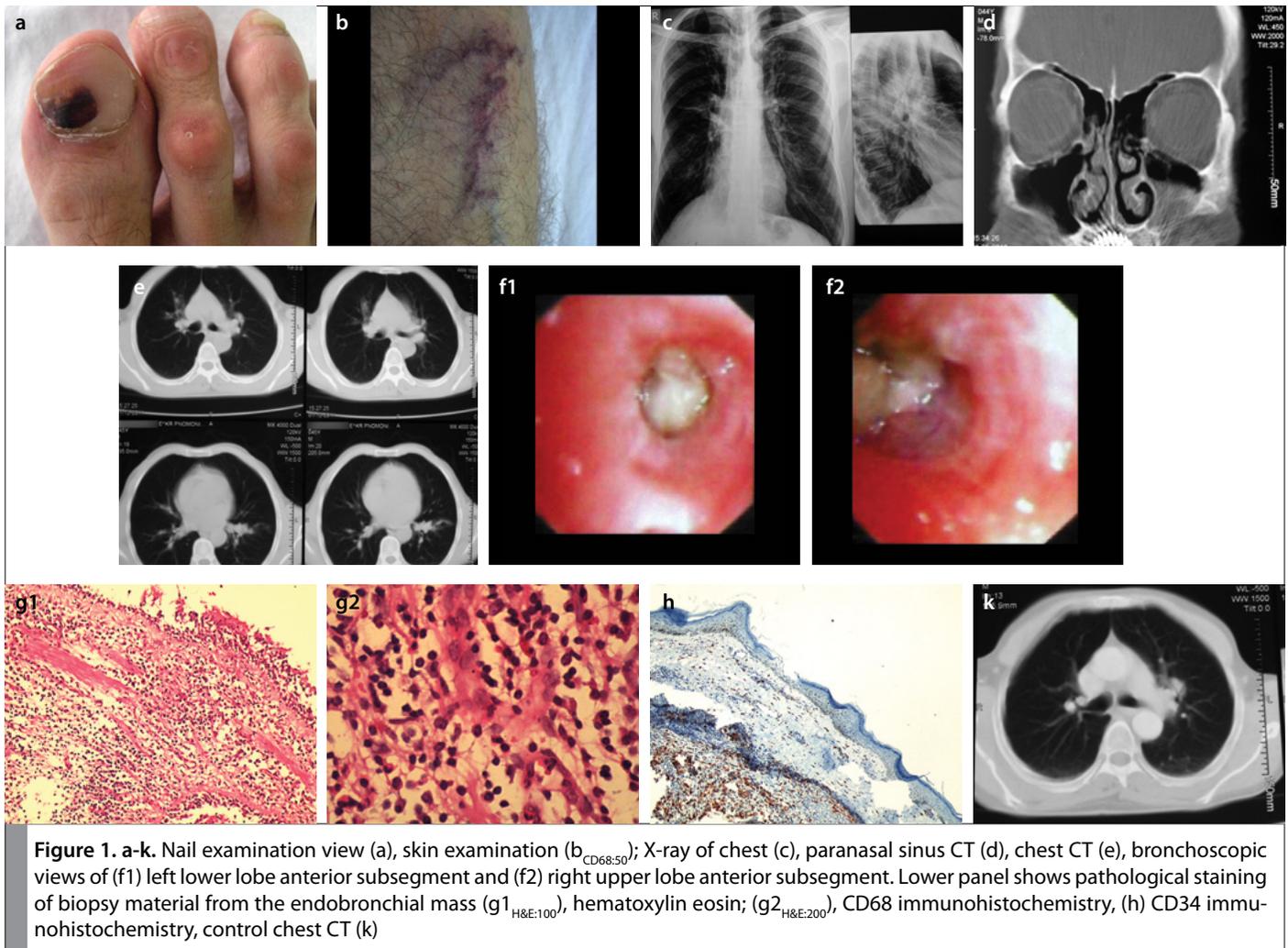


Figure 1. a-k. Nail examination view (a), skin examination (b_{CD68:50}); X-ray of chest (c), paranasal sinus CT (d), chest CT (e), bronchoscopic views of (f1) left lower lobe anterior subsegment and (f2) right upper lobe anterior subsegment. Lower panel shows pathological staining of biopsy material from the endobronchial mass (g1_{H&E:100}), hematoxylin eosin; (g2_{H&E:200}), CD68 immunohistochemistry, (h) CD34 immunohistochemistry, control chest CT (k)

growth. Eosinophilia (30%) was present in bronchoalveolar lavage. Respiratory function tests showed obstructive type respiratory insufficiency [FVC: 3.13 (63%), FEV₁: 1.81 (45%), FEV₁/FVC: 57%]. Rheumatoid factor, anti-nuclear antigen, anti-myeloperoxidase, and anti-proteinase 3 were negative. Skin test for *Aspergillus fumigatus* was negative. Stool tests for parasites were negative. Pathologic examination of skin biopsy material from the deltoid region showed inflammatory granulomatous infiltrations with giant cell populations in upper and lower reticular dermis. Staining for CD68 and CD34 verified granulomatous inflammation and vasculitis, respectively (Figure 1h). CSS was considered. Two scoring systems have been developed to assess the disease activity in patients with CSS and other vasculitis syndromes: five-factor score (FFS) and Birmingham Vasculitis Activity Score. These scoring systems are used to guide initial therapy.

FFS includes cardiac insufficiency, renal insufficiency (creatinine > 1.7 mg/dL), gastrointestinal involvement, age > 65 years, and ear-nose-throat manifestations. One point is given for the presence of each factor. FFS ranges from 0 to 2. A score of zero is given when none of the factors are present, a score of 1 for one factor, and a score of 2 for two or more score. If FFS equals to 0, then only steroids are started. This scoring system has been correlated with prognosis.

Five-factor score in our patient was zero. According to guidelines, methylprednisolone 60 mg/day (0.5–1 mg/kg/day) was started. The

dose was decreased to 40 mg/day after 3 days. Then, dose was adjusted based on the course of disease. Deltoid redness and black spot on the thumb regressed in one month. Improvement of infiltrations was verified by chest CT (Figure 1k). The patient has been continuing prednol treatment for 1 year.

DISCUSSION

Churg–Strauss syndrome is a rare syndrome characterized by asthma, necrotizing vasculitis, eosinophilic tissue inflammation and extravascular granulomas, fever, and peripheral eosinophilia (1). The incidences in general population and in asthmatic patients were 2.4/million/year and 6-18/million/year, respectively (2). CSS has three clinical stages. The initial stage is characterized by late-onset allergic disorders. Familial history of atopia is negative. Asthma, drug reactions, and allergic rhinitis occur. The second stage is characterized by eosinophilia in blood and tissues. The third stage is vasculitis. Systemic symptoms such as fever and weight loss occur years after the onset of allergic symptoms. The rate of involvement is 38%–40% for the lungs, 33%–48% for heart, 69%–75% for CNS, 70% for skin, 50% for kidneys, and 60% for gastrointestinal system (3). Our patient was in the vasculitis stage, and he had skin and lung involvement. According to the classification of American College of Rheumatology, the presence of more than four criteria (asthma, eosinophilia > 10%, mononeuropathy or polyneuropathy, transient lung infiltrations, paranasal sinus abnormalities, ex-

travascular eosinophilia in biopsy materials) provides 99.7% specificity (4, 5). Our patient showed asthma, eosinophilia, paranasal sinus abnormalities, extravascular eosinophilia, and lung infiltrations. In CSS, lung infiltrations are nodular or cavitary. Endobronchial mass appearance is unexpected (6). Szczeklik et al. (6, 7), reported 70% parenchymal abnormalities, 25% pleural effusion, 4% bronchial wall thickening, and 30% normal lung imaging in 17 patients with CSS. High resolution CT revealed ground-glass appearance and consolidation (86.7%), and centrilobular pulmonary nodules (25%). Infiltrations are randomly dispersed in most of the cases.

CONCLUSION

Churg–Strauss syndrome should be considered in patients presenting with an endobronchial mass.

Informed Consent: Informed consent was obtained from patient who participated in this case.

Peer-review: Externally peer-reviewed.

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REFERENCES

1. Rochester CL. The eosinophilic pneumonias. In: Fishman AP (ed). Fishman's Pulmonary Diseases and Disorders. M New York: Mc Graw-Hill Company, 1998: 1133-50.
2. Cordier JF. Pulmonary vasculitis. In: Olivieri D, duBois RM, editors. Interstitial Lung Diseases. Monograph of European Respiratory Society 2000; 14: 226-43.
3. Ozlen B, Ozdemir L, Eskiütüncü B, Cevirme L, Kurtar N, Soy M, et al. Churg-Strauss syndrome associated with montelukast. *Tuberk Toraks* 2008;56: 434-8.
4. King TE. UpToDate 2013: Clinical features and diagnosis of eosinophilic granulomatosis with polyangiitis (Churg-Strauss). Topic 4346 version 11.0 www.uptodate.com
5. Jeong YJ, Kim KI, Seo IJ, Lee CH, Lee KN, Kim KN, et al. Eosinophilic lung diseases: a clinical, radiologic, and pathologic overview. *Radiographics*. 2007; 27: 617-39. [\[CrossRef\]](#)
6. Castener E, Alguersuari A, Andreu M, Gallardo X, Spinu C, Mata JM. Imaging finding in pulmonary vasculitis. *Semin Ultrasound CT MR* 2012; 33: 567-79. [\[CrossRef\]](#)
7. Szczeklik W, Sokołowska B, Mastalerz L, Grzanka P, Gorka J, Pacult K, et al. Pulmonary findings in Churg-Strauss syndrome in chest X-rays and high resolution computed tomography at the time of initial diagnosis. *Clin Rheumatol* 2010;29:1127-34. [\[CrossRef\]](#)