A case of granulomatosis with polyangiitis (Wegener's granulomatosis) with marked infiltration of Tracheo-bronchial tree

Sir,

We hereby report a case of 36-year-old man who presented with history of pain in small and large joints, along with a history of gum ulcers for 3 years. He also complained of dyspnea on exertion [Grade III], mild intermittent scanty hemoptysis with hoarseness of voice for 6 months and intermittent low grade fever for 2 months. Past medical records revealed that he was treated as seronegative arthritis and was started on methotrexate along with steroids and non-steroidal anti-inflammatory agents with marginal symptomatic benefit. He had past history of chronic suppurative otitis media 10 years back for which he underwent typmanoplasty. On examination he was anxious looking man with mild pallor and had features suggestive of right facial nerve palsy. Complete blood count showed hemoglobin of 11.8 gm/dl, white blood cell count of 11,600 ku/ml and normal platelet count. Serum creatinine was elevated (1.67 mg/dl). Urine routine microscopy revealed 2 + proteins and 3 + blood, 20-25 RBC's occasional granular casts and 1-2 pus cells. Twenty-four-hour urinary protein collection revealed mild proteinuria (290 mg/24 hrs). Liver function tests were normal except for hypoproteinemia (Total proteins 5.68 gm/dl and Serum albumin 2.94 gm/dl). ESR was elevated (68 mm/1st hr). Serum electrolytes were normal with serum calcium of 8.7 mg/dl. Mantoux test was negative. Chest radiograph was apparently normal. Echocardiography was normal. Ultrsound abdomen showed mild fatty liver and features of bilateral medical renal disease. CT chest showed bronchial wall irregularities and thickening of tracheobronchial tree with sub-centrimetric nodules in left upper lobe and bilateral lower lobes with mediastinal adenopathy. Fibreoptic bronchoscopy revealed sluggish movement of left vocal cord and subglottic area showed infiltration and mucosal irregularities [Figure 1]. Bilateral bronchial tree showed mucosal infiltrates and growth with almost complete occlusion of right middle lobe [Figure 2] and biopsy revealed granulomatous inflammation. Other results were as following: ANA profile, RA factor: Negative, serum ACE levels (22.7 U/L/NR: 16-85 U/L) C-ANCA positive p - ANCA negative and anti-PR3 value (63.3 U/ml: Normal value 6-8 U/ml). In view of granulomatous inflammation on histopathology, proteinuria, in association with positive C-ANCA and anti-PR3, a diagnosis of granulomatosis with polyangitis was made (Generalized Active Stage). He was started on pulse methylprednisolone with and IV cyclophosphamide with maintenance treatment with oral prednisolone. He had significant improvement in joint pain and hemoptysis subsided. His repeat bronchoscopy was done after 4 months of treatment which showed complete resolution of nodules and ulceration of tracheobronchial tree [Figure 3].

Involvement of the respiratory mucosa can occur along the entire length of the lower and upper airways in 15%-55% of the patients with GPA. In 25% of the patients with GPA, involvement of the airways can be the only manifestation.[1] GPA can cause alterations in any segment of the airways, including inflammation, ulceration, pseudomembranes, tracheobronchomalacia, and destruction of cartilages, endobronchial masses, and laryngeal tracheobronchial stenoses.[2] Subglottic stenosis is the most common manifestation in patients with airway involvement as seen in present case.[3] Other causes of subglottic stenosis are post-intubation stenosis, post-infectious stenosis, and other systemic diseases like Crohn's disease, sarcoidosis, and Behcet's syndrome.[2] Bronchoscopy helps in the diagnosis and follow up of such alterations, as well as treatment through the re-establishment of the functional airway patency.[4] Our patient also presented with predominant endobronchial involvement with hemoptysis and subglottic stenosis, diagnosis of GPA was confirmed through bronchoscopic biopsy. If stenosis is causing significant compromise of airways, it can be treated with intra-lesional injection of corticosteroids, balloon dilatation, dilatation by use of metal tubes,

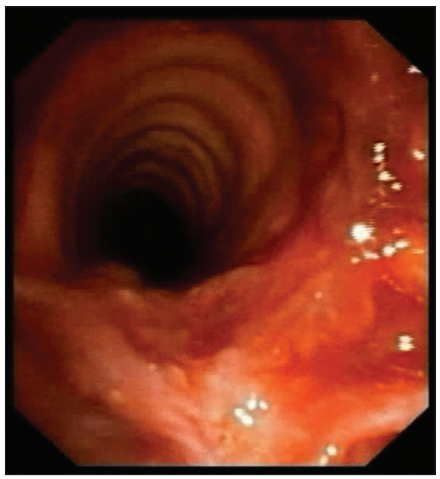

Figure 1: Video bronchoscopic image showing subglottic infiltration and stenosis

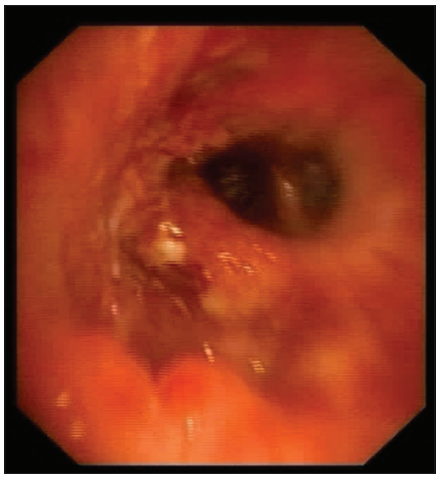

Figure 2: Video bronchoscopic image showing right middle lobe infiltration, narrowing and ulceration

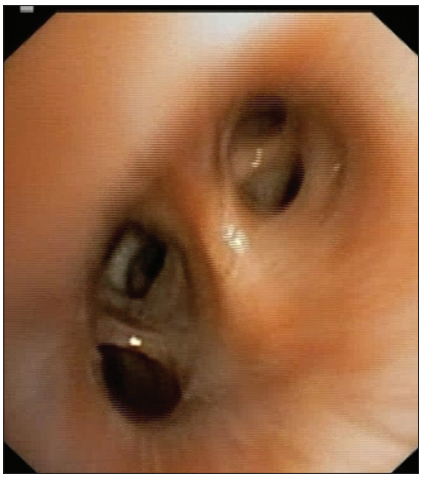

Figure 3: Video bronchoscopic image showing complete resolution of right middle lobe infiltration after treatment

laser, endoprosthesis, tracheostomy, surgical resection, and reanastomosis.[5] The rarity of present case is wide spread involvement of subglottic area, right and left bronchial tree with relative sparing of trachea. High-dose corticosteroids along with other immunosuppressants like cyclophosphamide resulted in significant resolution in our case. He had significant symptom relief with the same and is under follow-up since thereafter.

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REFERENCES

1. Gluth MB, Shinners PA, Kasperbauer JL. Subglottic stenosis associated with Wegener's granulomatosis. Laryngoscope 2003;113:1304-7.
2. Polychronopoulos VS, Prakash UB, Golbin JM, Edell ES, Specks U. Airway involvement in Wegener's granulomatosis. Rheum Dis Clin North Am 2007;33:755-75, vi.
3. Prince JS, Duhamel DR, Levin DL, Harrell JH, Friedman PJ. Nonneoplastic lesions of the tracheobronchial wall: Radiologic findings with bronchoscopic correlation. Radiographics 2002;22:S215-30.
4. Fauci AS, Haynes BF, Katz P, Wolff SM. Wegener's granulomatosis: Prospective clinical and therapeutic experience with 85 patients for 21 years. Ann Intern Med 1983;98:76-85.
5. Shapshay SM, Valdez TA. Bronchoscopic management of benign stenosis. Chest Surg Clin N Am 2001;11:749-68.

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