

Relapsing Polychondritis Presenting As Sub-Glottic Stenosis

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Citation

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Abstract

Relapsing Polychondritis is a rare disease of unknown aetiology, characterized by recurrent inflammation of the cartilages in different organs. Inflammation of the pinnae and nasal cartilages are the most common features, but the eustachian tubes, trachea, larynx, bronchi, cochlea and vestibule, and costochondral joints are frequently involved. An arthropathy and an ocular inflammation are often seen. The disease is regarded as an autoimmune disorder because of the co-existence of Rheumatoid Arthritis, Sjogren's Disease, Systemic Lupus Erythematosus, Systemic Vasculitis, Behcet's Disease, Reiter's Disease and Ankylosing Spondylitis in about one third of patients with Relapsing Polychondritis. Laryngeal involvement is the presenting feature in about 15% of patients. This rare case of a young black female presented with sub-glottic stenosis, requiring emergency tracheostomy following bronchoscopy.

INTRODUCTION

Relapsing Polychondritis is a rare disease of unknown aetiology, characterized by recurrent inflammation of the cartilages in different organs. Inflammation of the pinnae and nasal cartilages are the most common features, but the eustachian tubes, trachea, larynx, bronchi, cochlea and vestibule, and costochondral joints are frequently involved. An arthropathy and an ocular inflammation are often seen. The disease is regarded as an autoimmune disorder because of the co-existence of Rheumatoid Arthritis, Sjogren's Disease, Systemic Lupus Erythematosus, Systemic Vasculitis, Behcet's Disease, Reiter's Disease and Ankylosing Spondylitis in about one third of patients with Relapsing Polychondritis.

Laryngeal involvement is the presenting feature in about 15% of patients. This rare case of a young black female presented with sub-glottic stenosis, requiring emergency tracheostomy following bronchoscopy.

CASE REPORT

A 29-year old black female was admitted as an emergency in moderate respiratory distress. She had a cough productive of white sputum and progressive shortness of breath for 3 weeks, accompanied by hoarseness of the voice, painful swallowing as well as anorexia and weight loss of 10 pounds over the previous 3 weeks. There was no fever, haemoptysis,

pleuritic pain, joint pain or headache and no family history of asthma, hay fever or eczema. She was treated by her general practitioner for asthma, with oral bronchodilators and antibiotics with no effect.

Clinical examination revealed a young lady in moderate respiratory distress, acyanotic, prolonged inspiratory phase and using the accessory muscles of respiration. Auscultation revealed widespread inspiratory rhonchi, but no crepitations. She was treated with an oxygen/salbutamol nebulizer, but did not respond to treatment. She was then seen by the Ear, Nose & Throat Surgeon (ENT) who diagnosed laryngeal obstruction due to oedema or spasm. Plain radiographs of the larynx (Figures 1 & 2) showed sub-glottic airway narrowing at the level of the 4th and 5th cervical vertebrae. A plain chest radiograph was normal.

Figure 1

Figure 1



Figure 2

Figure 2



LEGEND: Plain radiographs of the larynx (Figures 1 & 2) showed sub-glottic airway narrowing at the level of the 4th and 5th cervical vertebrae.

She was treated with intravenous hydrocortisone and antibiotics and improved although the hoarseness persisted. Direct micro-laryngoscopy showed both vocal cords were normal as was the anterior sub-glottic region. Following bronchoscopy she developed respiratory failure and an emergency tracheostomy was performed. A computed tomography scan suggested a sub-glottic stenosis.

She was re-admitted 2 months later with left-sided chest pain, cough and fever. There was tenderness over the costochondral joints anteriorly and over the manubrium and xiphoid regions. She was treated with antibiotics and discharged. Thereafter, biopsy of the sub-glottic stenotic area showed normal epithelium with sub-mucosal fibrosis and chronic inflammation.

One month later, she developed pain and swelling of both knee joints, with bilateral effusions. Aspiration was carried out and serosanguineous fluid was obtained, microscopy of

which demonstrated a chronic inflammatory exudate.

Two months later she experienced pain in both auricles simultaneously with a recurrence of painful knee joints. Clinical examination revealed a young lady in no cardio-pulmonary distress and a tracheostomy in-situ. There was pigmentation and tenderness on palpation of both auricles with sparing of the ear lobes. She had a saddle nose deformity. A diagnosis of Relapsing Polychondritis was made and a biopsy showed normal epithelium with sub-mucosal fibrosis and chronic inflammatory cells.

Investigations revealed an erythrocyte sedimentation rate (ESR) of 140 mm/hr, haemoglobin of 9.6 g/dl, white cell count of 17 000 with a normal differential count.

Rheumatoid factor levels were 1:80 and the anti-nuclear factor, serum C-reactive protein, plasma protein, calcium, phosphorous, alkaline phosphatase, creatinine, electrolytes, blood urea nitrogen and LE cells were all normal.

Prednisolone 40 mgs daily was commenced along with misoprostol and this resulted in amelioration of her symptoms with less painful knee joints and auricles. The ESR reduced to 30 mm/hr on a maintenance dose of prednisolone 20 mgs daily.

DISCUSSION

Jaksch-Wartenhorst in 1923 first described the disease under the name "Polychondropathia"[1], and Pearson, Kline and Newcomer (1960) coined the term Relapsing Polychondritis [2]. Up to 1972, 114 cases had appeared in the literature (Hughes et al 1972) [3] and a further 338 cases reported between 1972 and 1986 (Herman et al) [4]. Since then hundreds of cases have appeared in the last 14 years. The disease is commonest in caucasians, uncommon in orientals and very rare in blacks.

The commonest presenting symptom is bilateral auricular chondritis in 95% of cases. Nasal chondritis, ocular involvement, laryngeal and tracheal chondritis, cochlear and vestibular damage and inflammation of the costal cartilages are the other sites mainly affected. There are no other laboratory tests specific for Relapsing Polychondritis. A diagnostic criteria for the disease was defined by Mc Adam et al in 1976 [5]. [Table 1]

TABLE 1:

Mc Adam's diagnostic criteria⁵ for Relapsing Polychondritis:

1. Recurrent chondritis of both auricles
2. Non-erosive inflammatory arthritis
3. Chondritis of nasal cartilages
4. Ocular inflammation – conjunctivitis, keratitis, scleritis, episcleritis and uveitis
5. Cochlear and/or vestibular damage, sensorineural hearing loss, and/or vertigo

A definite diagnosis of Relapsing Polychondritis is made when 3 or more of the signs are present and there is no need for histological confirmation of the diagnosis. Histology reveals loss of basophilic staining of the matrix, associated with infiltration of leucocytes, lymphocytes, monocytes and plasma cells. Laboratory investigations may reveal an elevated erythrocyte sedimentation rate (ESR), leucocytosis, thrombocytosis, eosinophilia and a normochromic, normocytic anaemia. Arthritis can involve any joint and may resemble rheumatoid arthritis due to its migratory nature.

Involvement of the inner ear in Relapsing Polychondritis may result in vestibular symptoms (nausea, vomiting, vertigo) or auditory symptoms (decreased hearing and/or tinnitus). One key feature of auricular chondritis is that the inflammation usually spares the fleshy ear lobe and without specific therapy, it may resolve although deformities such as cauliflower ear may ensue. Scleritis, episcleritis and conjunctivitis are the commonest ocular manifestations, but the lids and adnexae may also be involved.

Nasal chondritis may occur with acute painful swelling and tenderness of the septum, accompanied by epistaxis; untreated or severe Relapsing Polychondritis may result in a saddle-nose deformity.

A proliferative glomerulonephritis sometimes occurs with azotemia, proteinuria and a raised serum creatinine occasionally requiring dialysis. Cardiac involvement in relapsing polychondritis is a potentially fatal complication resulting from aortic and valvular lesions, which may lead to aortic dissection and aortic or mitral regurgitation. Cardiac arrhythmias may occur.

Skin manifestations are seen in about 20% of patients with Relapsing Polychondritis and in a few cases skin biopsies show lesions such as erythema nodosum, livedo reticularis, kerotoderma blenorrhagicum and leucocytoclastic vasculitis. Cutaneous polyarteritis nodosa has also been

described.

Almost any acute or subacute mono or polyarthropathy and neurological manifestations in the form of cerebral and cerebellar dysfunction can occur in about 20% of patients as a result of the vasculitis [6, 7]. Sauvaget et al described an uncommon complication of relapsing polychondritis called mesenteric panniculitis in a woman presenting with fever and abdominal and pelvic pain [8]. Abdominal pain may also occur as a result of vasculitis or ulcerative colitis, with which the disease may be associated.

Otorhinolaryngological complications are common and involvement of the upper airway is a potential life-threatening event, involving the trachea or main bronchi and necessitating tracheostomy. Inflammation of the glottis and sub-glottic region causes cough, hoarseness and dyspnoea, which may lead to stridor and airway collapse. Although involvement of the upper airway is always symptomatic and easily diagnosed, lower airway disease may be asymptomatic and detected only by computed tomography and pulmonary function tests.

Corticosteroids remain the mainstay of treatment and alleviates acute laryngeal inflammation. Other drugs used include azathioprine, cyclophosphamide,

6-mercaptopurine, penicillamine, cyclosporin and dapsone. Plasmapheresis may also have a role in the treatment of patients with relapsing polychondritis due to antibody involvement in the pathogenesis of the disease. Gaffney et al used nebulised racemic ephedrine for sub-glottic oedema in their case of relapsing polychondritis with good results [9].

Surgical management includes the use of an endotracheal prosthesis and extra-luminal splinting in dynamic airway collapse. Recently, Dunne and Sabanathan used metallic stents in the management of a patient with tracheo-bronchial involvement [10].

Sudden death may occur because of tracheal cartilage collapse and tracheostomy should be done early in those patients with sub-glottic involvement [11]. Bronchoscopy

and endoscopy with intubation are however fraught with hazard and may lead to acute respiratory failure as occurred in this case.

Due to increased awareness by physicians, relapsing polychondritis is being diagnosed more often. Laryngeal involvement is characterized by acute attacks of laryngeal oedema leading to sub-glottic stenosis, which can be life-threatening with recent studies by Okuyama et al [12] suggesting a role for gallium uptake studies to evaluate inflammatory activity in Relapsing polychondritis.

Bronchoscopy is an essential procedure in this disease, but may lead to sudden respiratory embarrassment, which may be fatal. We suggest that patients with sub-glottic oedema/inflammation or stenosis, should have early tracheostomy performed to avoid any respiratory embarrassment.

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