Case Report

Ear, Nose and Throat manifestations of relapsing Polychondritis in a child

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ABSTRACT

Relapsing polychondritis is a rare disorder, that can present initially to Otolaryngologists and can easily be overlooked due to its relative rarity and atypical initial symptoms and signs. Here we report on a 12-year-old schoolboy who presented initially with ear, nose and throat manifestations, including stridor, cough, hoarseness, sore throat and fever. The other clinical signs such as nasal tip, depression and softening of right auricle, developed subsequently. The clinical features, laboratory investigations, diagnostic criteria and treatment options were discussed with a brief review of literature.

Keywords: Relapsing polychondritis, otolaryngological manifestations, children, corticosteroids.

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Relapsing polychondritis (RP)[1] is a rare, systemic, multifocal disorder, of unknown etiology, presumably an autoimmune disorder, characterized by recurrent inflammatory degeneration of various cartilaginous structures throughout the body, including auricles, ribs, joints, nasal, laryngeal and tracheal cartilages.[2,3] It is often associated with other autoimmune diseases such as systemic lupus erythematosus (SLE) and Wegener's granulomatosis.[3,4] Furthermore, an association with human leukocyte antigen (HLA)-DR4 has been described.[5] It can occur in any age but is unusual in children. Its peak incidence is between the ages of 40-60 years[3,6] with no sex predilection.[2,6] The diagnosis of RP is mainly clinical and no laboratory tests are specific but the erythrocyte sedimentation rate (ESR) is typically elevated, indicating the disease activity.[7] Many affected patients of RP have mild normocytic anemia, leucocytosis and they may show some degree of vasculitis of small and large arteries.[8] Treatment includes high dose, long term steroid therapy with a possible need for immunosuppressive agents for advanced cases.[9] We report, the first case of RP in a schoolboy from the central region of the Kingdom of Saudi Arabia, to highlight this rare and unusual condition as it presented essentially as an otolaryngological problem.

Case Report. A 12-year-old Yemeni schoolboy, presented to the Emergency Room (ER), with progressive respiratory difficulty, sore throat, intermittent fever, cough and hoarseness, for 6 weeks. He gave no history of foreign body inhalation or weight loss. Clinical examination showed marked inspiratory stridor and limited mobility of both vocal cords but no obvious mass. Other ear, nose and throat (ENT) and systemic examinations were essentially normal. The hemogram was normal except for the leukocyte count of $18x10^9/l$ and ESR of 83 mm in the first

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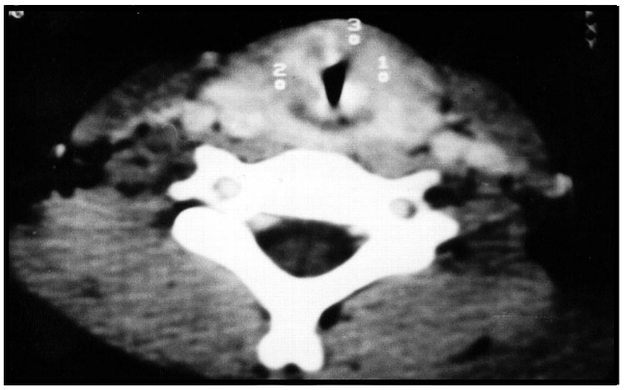

Figure 1 - Computerized tomography (CT) scan of the larynx showing diffuse mass in the paraglottic region (marked 1, 2 and 3) with narrowing of the subglottis and upper trachea.

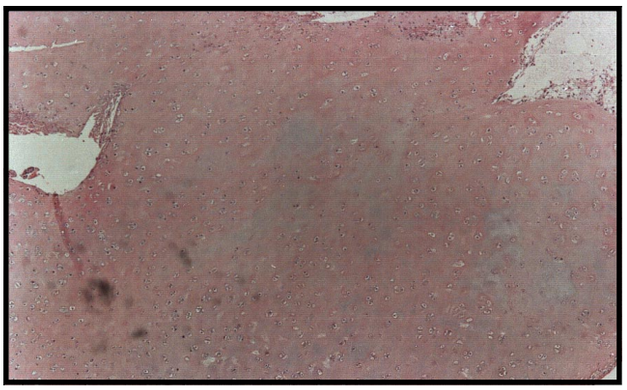

Figure 3 - Photomicrograph of the septal cartilage showing multiple degenerative (pale areas) and inflammatory (top corners) changes (Hematoxylin and Eosin stains; x 100).

hour. Plain x-ray of the neck soft tissue, showed diffuse mass in the paraglottic area and narrowing of the subglottis and upper trachea and this was reconfirmed by the computerized tomography (CT) scan of the same area (Figure 1). The chest x-ray, the neck ultrasound and radioisotope scanning of the thyroid were all normal. A working diagnosis of acute laryngotracheobronchitis (croup) was made and the patient was eventually admitted to the intensive care unit (ICU) after endotracheal intubation. He continued to improve on intravenous (IV) hydrocortisone 50mg 6 hourly and cefotaxime 500 mg 8 hourly. Due to marked improvement, the patient was discharged home on the 10th day of admission. Unfortunately, one week later he was readmitted for the same complaints of episodic fever and severe stridor and was restarted on IV hydrocortisone and ampicillin 250mg 6 hourly. Direct laryngoscopy, revealed a diffuse edematous area in the paraglottic region and the biopsies submitted from this area revealed non-specific chronic inflammation. The patient continued to run intermittent spikes of fever, joint pains and stridor. Repeat CT scan was inconclusive. Thus IV vancomycin 250 mg 6 hourly was added and tracheostomy was carried out. The battery of investigations performed included repeated leukocyte counts ranging from $13x10^9/l$-$20x10^9/l$; ESR ranging from 40-120mm in the first hour, the sputum for Acid Fast Bacilli (AFB), blood culture, Mantoux, Treponemapallidum hemagglutination (TPHA), Widal, rheumatoid factor, antinuclear factor (ANF) and the brucella tests were all negative. The liver function tests, renal and thyroid profiles showed no abnormalities. Repeat x-ray of the chest and cervical spines were also normal. Eye examination including fundi showed no abnormality at this stage but later, episcleritis was detected. The hearing tests and the echocardiogram were also normal. By this time, the patient developed nasal tip discoloration and depression with softening and collapse of the nasal septal cartilage (Figure 2). Likewise, the right auricle was noticed to be hyperemic and rather soft, and on repeat endoscopy, the laryngeal framework and tracheal rings were found to be soft and collapsing. Biopsies from the nasal septal cartilage (Figure 3) showed, multiple degenerative changes, consistent with the clinical diagnosis of relapsing polychondritis. The antimicrobials were then discontinued and aspirin 500 mg daily was started and steroid therapy was readjusted. The patient was referred to King Faisal Specialist Hospital and Research Center, (KFSH&RC) Riyadh, where the diagnosis was

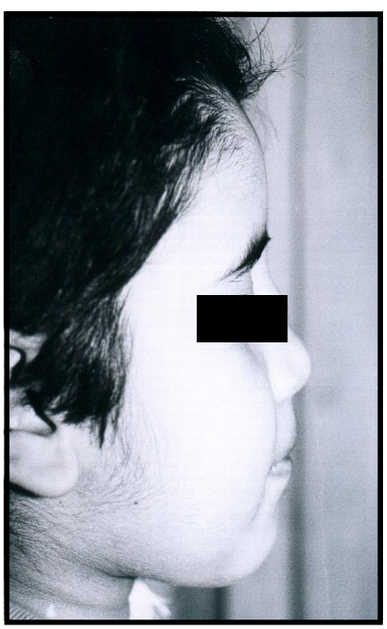

Figure 2 - The patient with evident nasal tip depression secondary to septal cartilage softening and collapse.

confirmed and methotrexate with supplemental pulse steroids and an analgesic were suggested. No relapse was observed since then after a follow-up of more than 18 months.

Discussion. Relapsing polychondritis is a multisystemic rheumatic disease, characterized by a wide-spread and potentially destructive inflammatory lesion of the cartilage. It was first described by Jaksch-Wartenhorst in 1923 who had named this entity as polychondropathia.[10] The term RP initially appeared in 1960 in an article by Pearson[1] et al and by the early 1990s, several cases of RP had been reported in literature.[9] The clinical manifestations of RP are myriad and it frequently presents initially to otolaryngologists. Thus it can be easily mistaken for upper respiratory tract infection, croup or infectious cellulitis as happened in the case under discussion. On both first and 2nd admission, the patient was diagnosed to have croup on clinical symptoms. However, when eventually more clinical signs were displayed the diagnosis of RP was thought of. The diagnostic criteria set out by McAdam[2] et al, modified by Damiani[11] et al and Michet[12] et al, held true in this child's case, by virtue of the confirmed histological changes, involvement of the multiple anatomic localities and the good therapeutic response to steroids. The various laboratory investigations carried out for this patient, revealed inconclusive evidence and none of the laboratory tests are claimed to be pathognomonic for this disorder. However, many RP affected patients were reported to have normocytic anemia and leucocytosis.[13,8] The ESR is typically elevated in RP and it is often used as an indicator of the disease activity and not for diagnosis of the disorder. The etiology of RP is still unknown. However, there is evidence suggesting immunologically mediated mechanisms in its pathogenesis. Autoantibodies against native collagens (II, IX and XI) have been reported.[14,15] Rapid fatalities can occur during the recurrent exacerbation as the result of the respiratory, renal, cardiovascular or neurological involvement. The majority of RP reported cases were managed medically. However, the choice of therapy is generally based on the severity of symptoms and the extent of the organ involvement. Unfortunately, there is no universally accepted medical regimen. This therapeutic dilemma is mainly due to rarity of this condition and its unpredicted natural history.[16] Medical treatment traditionally consists of salicylates, non-steroidal anti-inflammatory agents and dapsone for mild episodes and systemic corticosteroids and possibly immunosuppressive agents for advanced cases.[9] In addition to medical treatment, emergency tracheostomy was carried out earlier in this case as a life saving procedure and it is still adequately maintained at home.

In conclusion, in pediatric otolaryngological practice, RP poses a difficult diagnostic problem, specially when occurring atypically in a child or when the characteristic changes in nose and ear are absent at the initial stage. Therefore, it should be considered as a rare possible diagnosis whenever inflammatory features are refractory to the conventional treatment.

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