Nasal Polyp : a Rare Presentation of Hypereosinophilic Syndrome

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Introduction

Hypereosinophilic Syndrome (HES) represents a heterogeneous group of leukoproliferative disorders associated with prolonged eosinophilia without any detectable cause along with multiple organ dysfunction. The eosinophil count exceeds 1500/cu mm and persists for at least 6 months unless death intervenes. It is rare in children, most cases are reported in females between 20 and 50 years. The clinical presentation is very insidious but may present as a complication for the first time. We report one case of HES that presented as nasal polyp.

Case Report

A 15 year old girl presented with bilateral nasal obstruction and left sided occasionally blood stained mucopurulent nasal discharge that had been progressive for the past 5 months. Nasal obstruction was present 2 years ago for which some mass was removed from the left nasal cavity elsewhere on two occasions. She had loss of weight and complained of easy fatigability. There was no history of allergy, headache, fever, oliguria, haematuria, joint pains, skin lesions, jaundice or worm infestation. On examination, she was thinly built, afebrile but anaemic. There was broadening of the nose due to a single, large, fleshy, polypoidal mass in the left nasal cavity deviating the septum to the right and causing bilateral total nasal obstruction. The mass was pale pink in colour, soft, smooth and did not bleed on touch. The mass was occupying the nasopharynx and was visible behind the soft palate in the oropharynx similar to a large antrochoanal polyp. Spleen was palpable 3 cms below costal margin. There was no lymphadenopathy, neuropathy or hepatomegaly. Clinical diagnosis of antrochoanal polyp with anaemia was made and she was hospitalised for surgical removal of the mass. Investigations revealed Hb-8.5 gm%, total leukocyte count - 31,000/cu mm (P-16, L-14, E-69, M-01), AEC-3800/cumm, platelets - 2.07 lac/cu mm, ESR-50 mm fall. Peripheral blood smear showed normocytic hypochromic blood picture with eosinophilia, no blast cells or haemoparasites were seen. Urine and stool examination were normal. All other biochemical and haematological tests were normal. X-ray chest, ECG, echocardiogram were normal. Ultrasound

abdomen confirmed splenomegaly. Bone marrow was markedly cellular with significant increase in eosinophils and its precursors. CT scan of the paranasal sinuses showed homogenous soft tissue opacity occupying the left antrum (Fig 1), nasal cavity and the nasopharynx (Fig 2). Punch biopsy from the nasal mass revealed a benign polyp with no respiratory epithelium. She was dewormed and given haematinics, vitamins and hetrazan for 3 weeks while the investigations were on. There was no significant reduction in eosinophil count or rise in haemoglobin level. Since eosinophilia was present for more than 6 months without any cause she was labelled as HES. She was given prednisolone 1 mg/kg once daily for four weeks and then tapered off in another two weeks. After six weeks TLC fell to 11,500/cu mm and eosinophils to 12%. Hb was 9.6 gm%. She was then operated upon for removal of antrochoanal polyp under general anaesthesia. By a lateral rhinotomy approach, the antral portion of the polyp was detached from the nasal and choanal parts and the latter delivered per orally in one piece. There was no difficulty in completely removing the antral part by traction with a Luc's forceps because the lateral wall of the nose was found destroyed due to the pressure atrophy by the polyp (Fig 1). There was no significant bleeding. All the turbinates were atrophied due to pressure of the polyp. There were no post operative complications. Histopathology of the polyp revealed a covering of stratified squamous epithelium. Subepithelial tissue was edematous and infiltrated with numerous eosinophils, few plasma cells and neutrophils. Blood vessels displayed a prominent endothelial lining and therefore stood out prominently in the stroma. She is under regular follow up. One year after diagnosis she is in remission without drugs. There is no recurrence of nasal polyp or any other nasal symptoms

Discussion

Marked eosinophilia is a common finding in many diseases [1]. Most commonly seen in allergic and parasitic conditions and less commonly during the course of many bacterial and mycobacterial infections. Occasionally eosinophilia is seen in conjunction with malignancies and blood dyscrasias. A separate syndrome has emerged over the past 60 years characterised by persistent marked eosinophilia of the

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Fig. 1 : CT scan showing soft tissue mass in left maxillary antrum and nasal cavity

blood associated with diffuse infiltration of organs with eosinophils. Various workers have given different names to these syndromes even though there have been many overlapping clinical and laboratory features. Recent workers have therefore referred to them as HES. Hardy and Anderson first described this syndrome in 1968. The three features required for the diagnosis of this syndrome were defined in 1975 by Chusid et al [2]. First, there must be a sustained eosinophil count of greater than 1500/cu mm for more than six months. Secondly, there must be no other etiologies for eosinophilia present. Thirdly, patients must have signs and symptoms of organ involvement. The last requirement excludes all benign eosinophilia that may be present for a long time without associated pathology. Initial chief complaints of these patients are quite varied. Symptoms frequently seen at the time of disease onset are weight loss with anorexia and fatigue, recurrent abdominal pain, fever with night sweats, persistent non productive cough with chest pain, various neurological abnormalities, pruritic rash and congestive cardiac failure. Cardiovascular, pulmonary, haematopoietic and nervous systems are most often involved followed by hepatic, dermatological, gastrointestinal and urinary systems (kidneys). During extensive review of literature we have come across a few cases of HES involving the middle ear and lungs [3,4] but no mention of any other otorhinolaryngological involvement. Because of the well known eosinopenic effects of corticosteroids, the therapy of hypereosinophilia has centered around the use of steroids and antileukaemic drugs [5]. A universally acceptable protocol for treatment of HES is still lacking. A patient of HES who has no symptoms or organ dysfunction despite a high AEC needs no treatment besides close follow up at 2-3 monthly intervals. Symptomatic patients must be offered steroid therapy.

Fig. 2 : CT scan showing extension of the mass in the nasopharynx

Those not responsive to steroids should be offered chemotherapy. Since organ damage can occur many years later, alfa interferon has been used in some cases with success [6]. Although mortality in HES is high, aggressive medical and surgical treatment can result in significant clinical improvement and outcome is much better than before. Our case is possibly a very rare ENT presentation of HES. Patient has achieved remission with steroids and surgery.

The HES groups together a number of conditions in which eosinophilia occurs for no apparently identifiable cause. It may be accompanied by involvement of lungs, pleura, heart, pericardium, liver, gastro-intestinal tract, peripheral nerves or skin. Involvement of the ENT region is extremely rare. Initial reports indicated a uniformly grave prognosis for HES but recent reports suggest a more favourable outcome in certain cases. This case is reported for its rarity in a 15 year old girl with a very unusual clinical presentation of a large nasal polyp.

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