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A Rare Unusual Facial Swelling

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Introduction

We report and discuss a case of unusual eosinophilic epithelioid granulomatous reaction. 13 years old male resident of Bihar, India was admitted with history of swelling left side of the face since 2 months, fever and swelling around right eye since1month.

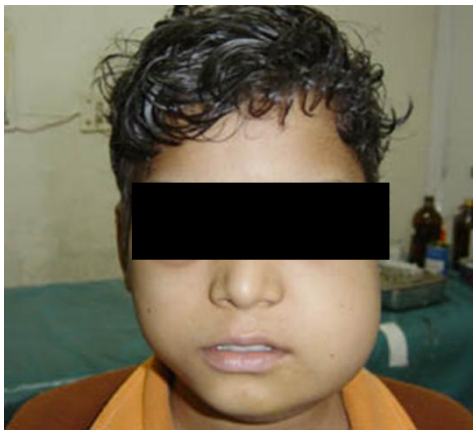
There was no history of ear discharge/ache, difficulty in mastication, excessive weight loss, chronic cough, discharging sinus and bony tenderness. No history of Joint pain/swelling, decrease visual acuity/dry eyes, testicular enlargement, tuberculosis contact, bleeding, blood transfusion. At the time of admission, vitals were temperature 102.6°F, Heart rate 100/min, Respiratory rate 30/min, Blood pressure 100/62mmHg. On general examination, no pallor, icterus, clubbing, pedal edema, petechiae. Left Axillary lymph node was 1.5 cms, soft, tender, mobile, overlying skin normal. On local examination there was a diffuse swelling over left parotid region extending up to submandibular region, firm, regular, immobile, non-tender, overlying skin normal, no sinuses, not lifting pinna. Dentition normal. Submandibular gland enlarged, mobile, no ducts discharge. There was mild proptosis of right eye, pupils were normal size reacting to light with normal movement, fundus normal, no xerosis, Schirmer's test was normal(for tear film adequacy). On abdominal examination, liver was palpable 3 cms below costal margin, no splenomegaly, no free fluid, and no lump, testicles normal, Chest was clear, in cardiovascular system S1 S2 normal. Central nervous system examination was normal and in locomotor system joints were normal.

The probable diagnosis was kept as one of the possibilities could be lymphoreticular malignancy. It was also thought as an autoimmune disorders (Mikulicz's syndrome, Sjogren's syndrome) or tuberculosis, or primary or secondary salivary glands neoplasms, also can be chronic sialo-adenitis or eosinophilic granuloma.

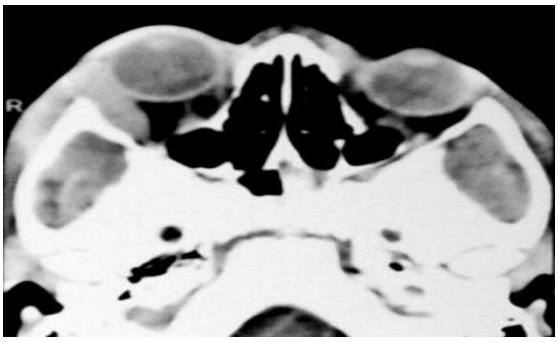
On basic investigations, Haemoglobin 11.5 g/dl, total leucocytes count 20,600/mm3, platelets count 206,000/ mm3, Peripheral Smear Neutrophil 32 Lymphocytes14 Eosinophiles 53 Monocytes1, Eosinophilia (Count 10,300/mm3) normocytic

normochromic red blood cells. Urine examination was normal, no proteinuria. Fever profile including peripheral smear for malarial parasite was negative and widal test was also negative. Blood culture was sterile after 5 days. Urine culture was also sterile. Liver function test was normal, renal function test were normal and even human immunodeficiency virus I/II ELISA were non-reactive. On radiological examination of chest, it was normal. On local examination of mandible, there was a soft tissue swellings but no lytic areas. Skeletal survey was done which revels no evidence of any bony malignancy. Primary or secondary Computed tomography Head &Neck was done which was suggestive of bilaterally enlarged and dense submandibular glands, dense deep lobe of left parotid enlarged and dense right acrimal gland, no area of necrosis. There were multiple non necrotic lymph nodes in the submandibular region. Ultrasound abdomen was normal. Fine needle aspiration cytology of left submandibular swelling was done s/o Lymphoid hyperplasia with increased number of plasma cells & eosinophils . Acid fast bacilli – Negative. Bone Marrow aspiration cytology was suggestive of mainly eosinophils & eosinophilic precursors. No abnormal cells seen. Bone marrow biopsy was also done s/o normocellular marrow maturing erythroid and myeloid series, increased eosinophilis precursor but no abnormal cells

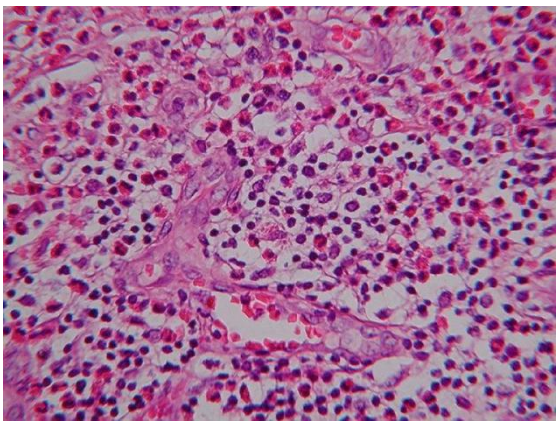
Lymph node biopsy was done suggestive of hyperplastic lymphoid follicles with prominent germinal centers, infiltration of plasma cells and eosinophils and Warth in Finkeldy type of polykaryocytes vascular hyperplasia. Serum IgE level was 2013 IU/ml. (Normal range = 1.00 - 181.00 IU/ml). In view of clinical profile, tissue and peripheral eosinophilia, HPE report and,High IgE levels, diagnosis of Kimura's Disease was kept. The patient initially showed response to broad spectrum antibiotics and had improved significantly. But again relapsed with recurrence of fever and increase in swelling, noteven responding to steroids, cyclosporine and montelukast. Hence radiotherapy was planned for this patient.



Picture 1: Swelling over the Right side of face



Picture 2: Magnetic Resonance Imaging: Bilaterally enlarged and dense submandibular glands, dense deep lobe of left parotid enlarged and dense right acrimal gland, no area of necrosis. There were multiple non necrotic lymph nodes in the submandibular region



Picture 3: Micrograph of Kimura disease, H&E stain

Discussion

Kimura disease is a rare, chronic inflammatory disorder.[1] It is a chronic inflammatory disorder of unknown etiology that most commonly presents as painless lymphadenopathy or subcutaneous masses in the head and neck region.[2] TRIAD of usually painless unilateral cervical adenopathy or subcutaneous masses predominantly in the head or neck region, blood and tissue eosinophilia, and markedly elevated Serum immunoglobulin E levels are enough to arrive at a diagnosis. The disease was first described in China by Kim & Szeto (1937).[3] It first received its name in 1948 when Kimura at el[4] and others noted a change in the surrounding blood vessels and referred to it as "unusual granulation combined with hyperplastic changes in lymphoid tissue." Kimura et al (1948) describe the epidemiological Age as Median 2nd –3rd decades, and Sex ratio Males (3.5:1), geography wise predominantly Asians. However, a study of 21 cases in the United States showed no racial preference, of 18 males and three females (male/female ratio 6:1), 8 to 64 years of age (mean, 32 years), and seven Caucasians, six Blacks, six Asians, one Hispanic, and an Arab were included.[5]Etiology is still unknown. Shetty A K et al[6] described it as a self-limiting allergic or autoimmune response triggered by an unknown stimulus, a viral or parasitic trigger with alter T-cell immune-regulation or induce IgE mediated type-1 hypersensitivity**.** Its cause still remains unknown. Reasons such as an allergic reaction, tetanus toxoid vaccination, or an alteration of immune regulation are also suspected. Other theories like persistent antigenic stimulation following arthropod bites and parasitic or candidal infection have also been proposed. To date, none of these theories has been substantiated.[7,8]

According to Hosaka N et al[8,9]eosinophils undergo accelerated apoptosis & multiple epitheloid granulomas with central eosinophilic abscesses and necrosis**.** Typical histiopatholog-ically involved lymph nodes shows prominent germinal centres, hyperplastic lymphoid follicles, variable interfollicular eosinophilia with eosinophilic micro abscesses and central necrosis, and Warthin-Finkeldey type polykaryocytes and proliferation of thin walled vessels. The clinical presentation typically is insidious onset, painless subcutaneous masses with adenopathy in the head and neck region (periauricular, axillary and inguinal) and generally involves parotid & submandibular salivary glands and rarely oral mucosa. Other unusual sites are auricle, scalp and orbit (lacrimal).

The clinical course of the disease is generally benign and self- limited. There is no potential that the lesions will become malignant.[9] It may be complicated by renal involvement. Even though it is a benign disease in a majority of cases, it can produce devastating renal and thrombotic complications.[10] Proteinuriamay occur in 12-16% cases. Nephrotic syndrome is the most common presentation in such patients; many may have normal renal function and no evidence of proteinuria. Diagnosis can be difficult. Kimura's disease can mimic other disorders such as angiolymphoid hyperplasia with eosinophilia (ALHE) which is the most common differential diagnosis .Others are Mikulicz's disease, eosinophilic granuloma, malignant lymphoma and salivary gland tumors and Sjogren's syndrome are also in the list of differencial diagnosis.. The common features shared by Kimura's disease and angiolymphoid hyperplasia with eosinophilia aremale predominance predilection for head and neck region, tendency to recur and vascular nature of the lesion with lymphoid & eosinophilic infiltrates.[11]

The diagnostic modalities includes laboratory test which revels peripheral blood eosinophilia &markedly elevated serum IgE levels. In the imaging studies, computed tomography (CT) & Magnetic Resonance Imaging (MRI) studies are useful to delineate the extent of the disease. Bio-psy is important to rule out malignant disorders.

Survival is excellent, as there is no evidence of malignant transformation and occasionally spontaneous resolution occurs. However, recurrence is common.

Treatment Modalities: Surgical excision, systemic corticosteroids, radiation and cyclosporine. Selection of mode of treatment for various patients depends upon the extent of the disease at the time of presentation. Surgical excision is the best option when it is a localized growth. Systemic corticosteroid is better if there are frequent relapses or when cases are complicated by nephrotic syndrome. Radiation therapy is needed in cases refractory to surgical and medical therapy or recalcitrant and large tumors when surgery is not feasible. Cyclosporin can be used in recurrent cases. Intravenous immunoglobulin may have value in the treatment of Kimura's disease.[12]

In one of the studies conducted by department of radiology, of Sapporo Medical University, School of Medicine, Japan in 1998[13] radiotherapy is an effective treatment with radiation field limited to the lesion & swelling of adjacent lymph nodes as much as possible. They found 26-30 Gy to be optimum dosage regardless of the tumor size. Irradiation should be considered not only in patients resistant to steroids but also in young patients in whom the long-term side effects of steroids may be more deleterious than a limited course of irradiation which may prevent relapse.[14]

Another study performed in the Department of Dermatology, at Nippon Medical School, Tokyo, Japan in 1999 found cyclosporin effective in recurrent cases as it affects the T helper-2 cells.[15]

A study is going on to assess the efficacy of tacrolimus on Kimura's disease. One case has so far been described. Tacrolimus may be an effective treatment for patients with Kimura's disease, but more research is needed to determine its long-term efficacy and safety, as well as its mechanism of action.[16]

However, regrowth of the lesion is common after discontinuing such treatments. In addition, the risks from radiation & chronic steroid therapy pose limitations. Hence there is a need for awareness of Kimura's disease by clinicians and pathologists to avoid unnecessary and potentially harmful investigations & treatment.

Key Message of this case study is that Kimura's Disease is a rare, self-limited, benign chronic inflammatory disorder of head & neck region. Disease can be diagnosed by simple procedures & if diagnosed properly, can be treated by simple procedures, but can recur.

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