An interesting case of neck swelling in an immunodeficient host.

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Abstract: There are multiple causes of neck swelling in an immunodeficient host. Finding out the exact cause of the neck swelling will greatly aid in the treatment and reducing the overall morbidity. Here we present such a case of massive neck swelling in a patient infected with human immune deficiency virus.

Keyword: Cervical Adenopathy, Antiretroviral Therapy, Anti Tuberculosis Therapy, CD4 Count, Fine Needle Aspiration Cytology, IRIS

Introduction: One of the most common causes of neck swelling is cervical adenopathy, which can be due to infective or non-infective causes. Infective causes include tuberculosis, oral sepsis, fungal infection of scalp, viral infections etc. Non-infective causes include malignancy of upper respiratory tract, lymphoma, sarcoidosis etc[1]. In patients with HIV infection neck swellings are a common occurrence and may be one of the reasons a person first visits a doctor. Finding the exact cause of such a swelling and distinguishing it as a malignant or non-malignant condition will greatly help in improving the quality of life of a HIV positive individual.

Case history
Mrs. S, a 34 year old female hailing from Krishnagiri got admitted in our hospital on 3/3/12 with the complaints of recurrent swelling in the neck for past 5 months. It started as diffuse swelling in the lower part of neck. It was initially painless and progressively increased in size to attain the present size.

Local examination of the swelling: Diffuse swelling extending from angle of mandible to clavicle vertically, horizontally from midline of neck to cervical spine posteriorly measuring 9*8 cms. It was firm and not moving with deglutition. There were multiple, firm nodules mostly in posterior region and entire left part of neck involved but right side was spared. There were no scars, sinuses, ulcers over swelling. It was not warm but tender on palpation.

Retroviral disease history: She was diagnosed HIV positive in September 2008. In Nov 2011, Antiretroviral Therapy[ART] was started[Zidovudine, Lamivudine, Nevirapine regimen] at ART centre, Krishnagiri when the CD4 was 282 cells. The patient noticed the swelling about a month after initiation of Antiretroviral therapy.

Antitubercular treatment[ATT] history: The patient was started on empirical ATT for a small neck swelling, in April 2010, on the left side in submandibular region. She did not have an FNAC done then as the swelling was minimal. She stopped taking ATT after 3 months since she felt better and the swelling decreased in size.
The patient underwent routine investigations.[table-1].

**Imaging:** Chest x ray was apparently normal. Ultrasound of chest and abdomen was normal. Ultrasound of the left side of neck revealed a mixed echo pattern. CT scan of the chest report-confluent lymph node masses of left side of neck, for histopathological correlation.[image-4,5,6]

**image 6**

In summary we had a 35 year old female, with human immunodeficiency virus infection for 4 years, on antiretroviral therapy for 6 months, massive cervical swelling for 5 months, probably of lymph node origin. Since the patient had history of ATT intake and was already started on ART and the swelling was not decreasing in size but actually increasing in size and nodularity, we had a high suspicion of two conditions: IRIS or malignancy. IRIS-(immune reconstitution inflammatory syndrome)- It is a phenomenon seen in HIV infected persons when these patients are started on antiretroviral therapy, there is sudden worsening of the patient’s opportunistic infections. This is accompanied by an improvement in the patient’s immune status which is seen as an increase in CD4 count.[10]. IRIS is classified into two types: unmasking type and paradoxical type[10]. The serial CD4 counts of the patient from the time of diagnosis is given in table-2.

As noted earlier the patient noticed the swelling after the initiation of ART. The CD4 count increased from 282 to 394 while on ART. So there was a high probability of IRIS in our case.

After getting due consent from the patient, we performed a fine needle aspiration cytology [FNAC] in a hard, nodular part of the swelling[7]. FNAC report was given as tuberculous lymphadenitis of the hyperplastic variety. This particular type of tuberculous adenitis is seen only in persons with good immunity and rare in an immunodeficient person.[2]. So our final diagnosis was hyperplastic cervical adenitis in HIV infected host with immune reconstitution inflammatory syndrome. After the FNAC report, the patient was started on category -2 ATT and ART was continued following which the patient improved.

**Discussion:** In tuberculous lymphadenopathy, cervical nodes most commonly affected (63%) with mediastinal (27%) and axillary nodes (8%) coming next. Nodes may be hard or fluctuant, not usually tender, have limited inflammatory changes in the overlying skin and there is a predilection for nodes in the posterior triangle (51%) & deep upper cervical (48%). In the majority of cases lymphadenitis is unilateral[9].

Four different types of clinical presentation of tuberculous lymphadenitis[8]

**Acute type** – This is seen in infants and children below five years. The glandular enlargement is painful and tender. The overlying skin is red and edematous. The child has moderate fever.
Caseating Type – This is most common type of clinical presentation seen in young adults. Multiple glands are moderately enlarged and matted together. The caseation leads to cold abscess and sinus formation.

Hyperplastic Type – In patients with good body resistance. Lymph nodes shows marked degree of lymphoid hyperplasia. Tuberculous granulation tissue is most productive with least caseation. Lymph nodes are enlarged freely mobile. Constitutional disturbances are minimal.

Atrophic Type – These are seen in elderly individuals in whom the lymphoid tissue undergoes degeneration. The glands are small and tend to caseate and burst.

In HIV infected persons, extra pulmonary TB more common. Node AFB smear positivity is more when compared with non HIV infected persons[2,3,4]. The immune reconstitution inflammatory syndrome (IRIS) in HIV-infected patients initiating antiretroviral therapy (ART) results from restored immunity to specific infectious or non-infectious antigens. A paradoxical clinical worsening of a known condition or the appearance of a new condition after initiating therapy characterizes the syndrome. One form of ir is that during treatment lymph nodes may increase in size . This phenomenon is known as paradoxical IRIS[immune reconstitution inflammatory syndrome]. The other type is the unmasking type ,where underlying infection is revealed or manifest only after starting ART. Potential mechanisms for the syndrome include a partial recovery of the immune system or exuberant host immunological responses to antigenic stimuli. The overall incidence of IRIS ranges from 10 to 37%, but is dependent on the population studied and its underlying opportunistic infectious burden[10,11]. The infectious pathogens most frequently implicated in the syndrome are mycobacteria, varicella zoster, herpesviruses, and cytomegalovirus (CMV). No single treatment option exists and depends on the underlying infectious agent and its clinical presentation.[10].

Diagnosis: A high index of suspicion is needed for the diagnosis of mycobacterial cervical lymphadenitis. A thorough history and physical examination, tuberculin test, staining for Acid-fast bacillus, radiologic examination and fine-needle aspiration cytology (FNAC)[7] will help to arrive at an early diagnosis of mycobacterial lymphadenitis which will allow early institution of treatment before a final diagnosis can be made by biopsy and culture[5]. The differential diagnosis includes tuberculosis, other infective or neoplastic causes (lymphoma or sarcoma metastatic carcinoma), non-specific reactive hyperplasia, sarcoidosis, toxoplasmosis, cats-scratch fever, collagen vascular diseases and diseases of reticuloendothelial system.[9]

Treatment
ATT is the mainstay of treatment. The role of steroids is controversial. Surgical management [modified radical neck dissection],[5] is done very rarely in rare occasions if nodes are massive, do not respond to treatment or for relief of pressure symptoms.

References: