Case Report
Inflammatory Pseudotumour

Manish A Chhabria*, Reeta J Dalal**, Shailendra R Maheshwari***, Chitra V Madiwale****

Abstract
A 29 year old female patient with submandibular swelling, anaemia, fever was diagnosed to have inflammatory pseudotumour of neck. We are reporting this case as it can affect diverse locations in body with varied clinical manifestations mimicking malignancy and posing a diagnostic challenge.

Introduction
Inflammatory pseudotumour (IPT) is a rare benign condition of unknown aetiology.1 The term pseudotumour was used as these lesions mimic invasive malignant tumours both clinically and radiologically.2 It was first described in 1905 by Birch-Hirschfield and remains an enigma despite several reports. Diagnosis is based on combination of clinical history, biochemical findings, histopathological and imaging features of an infiltrating soft tissue mass. If a diagnosis of IPT is confirmed, steroid therapy is advocated. It is important to avoid radical surgery in these cases.3

Case History
A 29 year old female was admitted to our hospital with recurrent fever of six months duration, submandibular swelling and inability to open her mouth since two months. She was detected to have severe anaemia two years ago and was administered two blood transfusions. A year ago she was admitted elsewhere for right abdominal pain. She underwent appendicectomy but had no relief. She developed fever hence was empirically put on anti tuberculous treatment (Isoniazid, Rifampicin, Ethambutol, Pyrazinamide) with a diagnosis of abdominal tuberculosis. However the fever persisted and she developed submandibular swelling, trismus, dyspnoea and was admitted to another hospital and treated as “Ludwigs angina”. As her symptoms worsened she was referred to our hospital. On admission, she was tachycardic, febrile, pale, had trismus (mouth opening < 1 cm), diffuse firm swelling on right side of neck and submandibular region, mildly tender submandibular lymph node – 1.5 x 1.5 cm and tender nodules on right arm. Chest examination revealed bilateral lower zone crepitations. She denied history of arthralgias, dry eyes and mouth. Flexible laryngoscopy was normal. Investigations revealed haemoglobin 4.5 gm/dl (hypochromic microcytic), white blood cell count of 30,370 / mm$^3$ with neutrophils of 90%, platelets 3,49,000 / mm$^3$, ESR of 140, CRP of 192 mg/L, total proteins were 7.8 gm/dl (albumin of 1.4 gm/dl), liver enzymes were normal, serum protein electrophoresis showed no ‘M’ band, renal profile was normal, HIV, HBsAg, HCV serology, ANA and anti neutrophil cytoplasmic antibody (ANCA) were negative, serum angiotensin converting enzyme (ACE) levels were 33 U/L. CT scanning of neck, chest and abdomen showed infiltrating fibroinflammatory soft tissue in right parapharyngeal space encasing the carotid sheath with infiltration of right side of neck extending across the midline, presence of infective consolidations in lower lobes of lung bilaterally, retroperitoneal and perirenal fascial thickening (Figure 1). Biopsy of cervical nodes (Figure 4) showed lymphoid tissue, mild fasciitis and fibrosis and biopsy of arm nodule (Figures 2 and 3) showed

Fig. 1: Ill defined infiltrating enhancing soft tissue thickening surrounding right carotid sheath with infiltration in right half of neck extending across midline to left

Fig. 2: Forearm Nodule

Fig. 3: Forearm Nodule

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is a possibility. Immuno histochemical studies of T- and B-cell subpopulations may be helpful in distinguishing lymphoma from IPT.5

Recently high levels of serum IgG4 have been documented in these systemic fibroinflammatory conditions. These have been labelled as IgG4 syndrome or IgG4 related systemic sclerosing disease. This condition is characterised by elevated serum IgG4 levels, tissue infiltration by IgG4 positive plasma cells accompanied by fibrosis and sclerosis.7 In the case reported IgG, its subtypes and IgE levels were not done.

Patients with IgG4 syndrome have clinical features of two or more organ involvement simultaneously or sequentially. A third of patients with IgG4 related systemic disease have dry eyes, dry mouth and arthralgias resembling Sjogren’s syndrome (SS).7 Despite similarities they are separate conditions with marked clinical and pathological differences.7 The pathogenesis of IgG4 syndrome is controversial (autoimmune versus allergic response).7 IgG4 is detected in several conditions and is not specific for this syndrome.7 Natural history of IgG4 related systemic disease is unknown. Spontaneous improvement followed by relapse has been reported. However, many progress to other organ system involvement which may be irreversible. Hence, there appears to be a limited window period when therapy can be effective.

Corticosteroids have been considered to be the standard treatment of these fibroinflammatory disorders.5 The studies reported in literature have short follow up periods, hence there is no uniform recommendation of dosage and duration of steroid therapy. The starting dose of prednisolone varies from 0.6 mg/kg/day to 1 mg/kg/day for 4-8 weeks, then tapered to maintenance dose for 4 months to 3 years. The high incidence of relapses on withdrawal of steroids is a major issue in their management. Steroid sparing agents like Azathioprine (2-2.5 mg/kg/day) or Mycophenolate mofetil (750 mg twice daily) have been used successfully in some cases.6 Tamoxifen and recently Rituximab have been used in steroid refractory cases.5

**Discussion**

Inflammatory pseudotumour is a benign inflammatory lesion of unknown origin.14 Numerous stimuli may act as triggers like autoimmune reactions, hypersensitivity reactions, unrecognised microorganisms. A localised derangement in the immune response after the initial insult may be an underlying mechanism for its development,5 others believe that most of the features of IPT are due to production of mediators of inflammation, such as cytokines particularly interleukin-1 which cause proliferation of fibroblasts.5 The typical features of IPT are co-existence of inflammatory cells and spindle cells, comprising fibroblasts and myofibroblasts with varying degree of fibrosis. The wide spectrum of histological appearances has led to a number of terms for this condition being used synonymously, including plasma cell granuloma, idiopathic fibrosing tumours, xanthomatous pseudotumour, and inflammatory fibromyxoid tumour.

IPT is reported to occur at any site in the body including the central nervous system, orbit, head and neck, lungs, pancreas, retroperitoneum and pelvis producing cranial nerve neuropathies, laryngeal obstruction, Riedel’s thyroiditis, mediastinal or pulmonary fibrosis, sclerosing cholangitis, sclerosing cervicitis and retroperitoneal fibrosis. The most frequent symptom in all locations is swelling and pain. Constitutional symptoms, such as fever, anorexia, weight loss, malaise, have been reported in 15% to 30% of cases of IPT. An extensive work up for collagen-vascular, infectious, neoplastic and granulomatous diseases may be negative. Whenever an infiltrating mass is seen in extracranial head and neck, lymphoma...