HEERFORDT-WALDENSTROM SYNDROME UVEOPAROTID FEVER: REVIEW OF THE LITERATURE AND A CASE PRESENTATION

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ABSTRACT

Heerfordt-Waldenström syndrome is also referred to as uveoparotid fever. In our patient physical examination showed bilateral parotid gland enlargement. Chest X-ray showed bilateral hilar lymph adenopathy. Biopsy specimen from the right parotid gland revealed scattered granulomas with focal central necrosis. Stains for acid-fast bacilli and fungi were negative. He was diagnosed as a case of Heerfordt-Waldenström syndrome, a rare form of sarcoidosis in which the compression of the facial nerve results in palsy. He was treated with 60 mg of prednisone daily, and at follow-up after two weeks later, the swelling and uveitis was resolved.

Keywords: Heerfordt-waldenstrom syndrome, Uveoparotid fever.

INTRODUCTION

Heerfordt syndrome also called as uveoparotid fever is a rare type of sarcoidosis which presents with fever, uveitis, parotid gland enlarement and cranial nerve palsies most commonly facial nerve[1]. In 1909, the condition was first described by Danish ophthalmologist Christian Frederick Heerfordt, for whom the syndrome is now named[2].

CASE REPORT

We report a case of 24 years old student who presented with 2 weeks history of fever, cough, and facial palsy, difficulty in swallowing and blurred vision (fig-1). Physical examination revealed bilateral facial nerve palsy and the eyes were red and swollen. Ophthalmologic examination showed anterior uveitis. Fundi were normal in appearance. Blood picture revealed haemoglobin 13 gm%, total leucocyte count 8500 /cu mm, neutrophils 65%, lymphocyte 32%, eosinophil 2% , monocytes 1% and ESR 40 mm/hr. Blood glucose, urea, creatinine and liver function tests were normal. Serum angiotensin-converting enzyme level was 150 U/L (reference range 8-65 U/L). Serum calcium was 9.7 mg /dl. X-ray chest (PA) view showed bilateral hilar lymphadenopathy (fig-2). Fine needle aspiration of parotid gland revealed noncaseating granulomas. The patient was

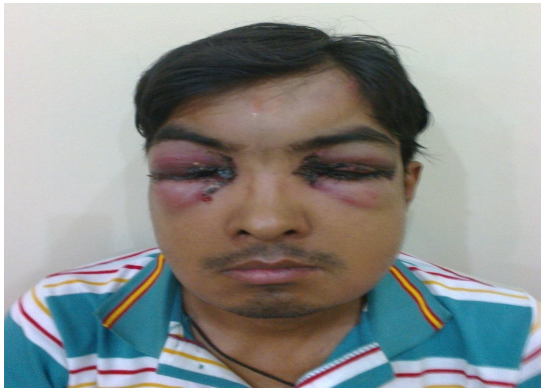

Figure-1:Showing the bilateral enlargement of parotid glands and anterior uveitis in this patient of Heerfordt-Waldenström syndrome.

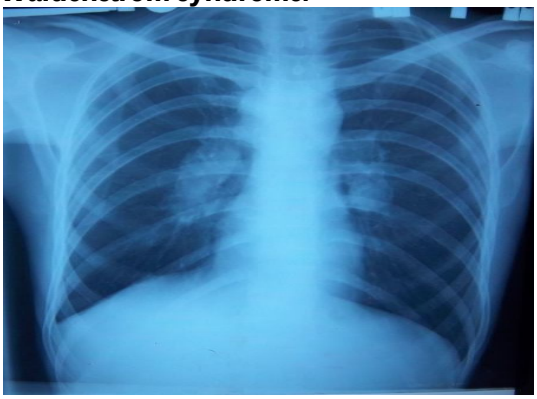

Figure-2: X-ray chest (PA) view shows bilateral hilar lymph adenopathy.

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diagnosed as a case of Heerfordt-Waldenström syndrome, or uveoparotid fever which is a rare initial presentation of sarcoidosis. Treatment with a high-dose steroid improved his parotid gland enlargement and anterior uveitis.

Sarcoidosis is a multisystem disease characterized pathologically by the formation of non-caseating granulomas. Heerfordt-Waldenström syndrome is a rare manifestation of neurosarcoidosis and has to be included in the differential diagnosis of facial nerve palsy. Involvement of lymph nodes, lungs, skin, spleen, liver, and the uveoparotid region have been described[3,4]. Anterior uveitis classically presents with infected conjunctiva, blurred vision, or eye pain[5].

Sarcoidosis has a prevalence of approximately 10 cases per 100,000 in whites and 36 cases per 100,000 in blacks in the United States[6]. Heerfordt-Waldenström is present in 4.1–5.6% of those with sarcoidosis[7].The tuberculin skin test is frequently used to distinguish it from tuberculosis since a negative tuberculin skin test is highly predictive for sarcoidosis[5,8].

ACE levels are raised in sarcoidosis are ,however the serum ACE level are not always related with disease activity[9,10].

This patient had marked improvement of symptoms after 2 weeks of prednisolone therapy. One month after discharge, his parotid gland swelling was regressed, and uveitis had improved.

CONCLUSION

Early diagnosis of Heerfordt-Waldenström is therapeutically rewarding by preventing ophthalmologic complications though the disease may be self-limiting. Increased awareness of rare manifestations will facilitate better management of these patients.

CONFLICT OF INTEREST

This study has no conflict of interest to declare by any author.

Permission for publication of photograph of the patient obtained.

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