HTLV Associated T-cell Lymphoma Presenting as a Sinus Mass with Proptosis: a Case Report

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ABSTRACT

Objectives: Describe a case of human T-cell lymphotropic virus (HTLV) associated T-cell lymphoma of the paranasal sinuses presenting with sinus destruction and severe proptosis.

Study Design: Case report.

Methods: The patient’s clinical course was reviewed, including photographs, radiographic studies, histopathologic slides. The relevant literature was reviewed.

Results: A 48 year old woman from Guyana presented with bilateral eye swelling and unilateral vision loss progressing over 4 weeks. On examination there was severe bilateral proptosis and chemosis. Imaging studies showed a large mass centered in the ethmoid sinuses with bilateral orbital invasion but no intracranial extension. An orbital biopsy revealed a large T-cell lymphoma. Serologic tests for HTLV were positive. She was treated with chemotherapy and responded well with improvement of vision in both eyes.

Conclusions: HTLV associated T-cell lymphoma is a rare etiology of a paranasal sinus lesion, but it should be considered in the differential diagnosis.

INTRODUCTION

We present a case report of a woman who presented with bilateral orbital proptosis, and was found to have HTLV associated T-cell lymphoma centered in the ethmoid sinuses.

CASE PRESENTATION

A 48 year old woman from Guyana presented complaining of bilateral eye swelling over 4 weeks with progressively worsening vision. She was treated in her home country with antibiotics, with no improvement. She denied headaches, anosmia, nasal obstruction, weight loss, or night sweats.

Physical examination revealed marked bilateral eye proptosis, left greater than right. The right eye had restricted movement, while the left eye was fixed. Vision was 20/50 in the right eye and she had no light perception in the left eye. Both eyes were chemotic with thickened, scaly lesions on the lids. Nasal endoscopy was unremarkable, with no lesions visible.

Laboratory testing was positive for HTLV-III. She was negative for HIV. An MRI of the brain and orbits with contrast is shown in Figures 1-3.

Biopsy of the lesion was positive for a large T-cell Lymphoma. She underwent chemotherapy with an EPOCH (etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin) regimen, and had a complete response, with resolution of the proptosis (Figs. 5-6), good vision in the right eye and limited vision in the left eye. Current follow-up is 14 months.

DISCUSSION

Adult T-cell leukemia-lymphoma (ATLL) is extremely rare in the paranasal sinuses. A large case series of 58 patients with lymphoma of the nasal cavity or paranasal sinuses was analyzed by Cuadra-Garcia(1) out of which only one was found to have ATLL. The one patient was a 39 year old Jamaican man, with ethmoid and sphenoid sinus involvement, who achieved complete remission but then relapsed with disseminated disease and died 3.5 years after presentation.

ATLL is a unique peripheral T-cell malignancy which is associated with human T-cell lymphotropic virus type 1 (HTLV-1), a retrovirus. The World Health Organization has subdivided this disease into four clinical categories: acute, chronic, smoldering, and lymphoma. The most common variant is acute, accounting for around 65% of patients. The lymphoma variant, which the patient presented had, is less common.

ATLL is predominantly seen in areas that are endemic to HTLV. The endemic areas are Japan and the Caribbean islands, although there are some cases reported in areas of South America and Africa, as well as Europe. The diagnosis is confirmed by histologic features, immunophenotyping of the T-lymphocytes, and serologic confirmation of HTLV-1 positivity. Other laboratory tests that may be abnormal are elevated LDH and calcium levels, as well as peripheral lymphocytosis, seen in the acute and chronic variants.

The mainstay of treatment is multidrug chemotherapy. For years the standard regimen was CHOP: biweekly cyclophosphamide, doxorubicin, vincristine, and prednisone. More recently, a phase III study (6) has shown superior results with a more aggressive regimen, which also involves carboplatin, etoposide, and ranimustine, among others (VCAP-AMP-VECP). This improved complete response from 25% to 40% and overall survival at 3 years from 13% to 24% compared to the CHOP regimen, although with increased toxicities. In those patients with poor clinical response, autologeneic stem-cell transplantation is considered. There is a 45% overall survival rate with this therapy, which some speculate may be due to a graft-versus-ATLL effect. There is current research into the use of interferon, as well as antiviral drugs (7).

The prognosis in ATLL is very poor, and the acute and lymphoma subtypes are the most lethal, with 5% survival at 4 years post-diagnosis. The chronic and smoldering subtypes have 27% and 62% 4-year survival rates, respectively (4).

In a literature review using PubMed, there were a total of 14 cases of ATLL of the paranasal sinuses identified. With the exception of the case identified in the case series already discussed, every other case reported was in Japan (2,3,5). These patients generally died within 1-2 years, but in a more recent case report, one patient was alive at 5-year follow-up, although with disease burden (5). This may reflect the more recent advances in therapy.

CONCLUSIONS

ATLL of the paranasal sinuses is very rare. Clinical suspicion of this entity is necessary, and the keys to diagnosis are tissue biopsy with immunophenotyping, and HTLV serology. The standard of treatment is multidrug chemotherapy, but while complete remission may be initially achieved, there is a very high risk for relapse with disseminated disease. Active research in this area may improve survival.

REFERENCES