Simultaneous Cavernous Sinus Syndrome and Facial Lesion as Presenting Signs of Ki-1 Positive Anaplastic Large Cell Lymphoma in a Child

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INTRODUCTION

Lymphomatous lesions in the cavernous sinus that produce visual symptoms or neuro-ophthalmologic abnormalities are rare. The skin also is an uncommon site of presentation for lymphoma. Lymphomatous skin lesions can mimic insect bites or skin abscesses, often delaying correct diagnosis and treatment. We describe a patient with lymphoma who presented simultaneously with a facial skin lesion and cavernous sinus syndrome.

CASE REPORT

Five weeks prior to presentation at our institution, an 8-year-old boy developed a crusting, elevated, erythematous skin lesion on his face, approximately 1 cm lateral to the left angle of his mouth. Nine days later, he developed ptosis of the ipsilateral upper lid. Three days after developing ptosis, an ophthalmologist diagnosed blepharitis and prescribed topical antibiotics. Two days later, an oral antibiotic was added by his pediatrician to treat the skin lesion that now was presumed to be an abscess. Failing to improve after 5 days of oral antibiotic therapy, he returned with an ipsilateral periorbital headache, photophobia, anorexia, and a temperature of 38°C. He was admitted to the hospital, and treatment for meningitis was begun, although computed tomography (CT) of the head and lumbar puncture were negative. Culture of the skin lesion also was negative. After 4 days of intravenous antibiotic treatment, his fever and photophobia resolved. The left periorbital pain, ptosis, and facial lesion, however, remained unchanged. With the diagnosis remaining in question, the patient was referred to our institution. The working diagnosis on presentation was an infectious cavernous sinus thrombosis transmitted from a facial abscess. Past medical and surgical history were noncontributory.

The patient's visual acuity was 20/20 OD and 20/30-2 OS. The pupils were symmetric in size and briskly reactive to light. A trace left relative afferent pupillary defect was present. The left eye demonstrated 4 mm of ptosis and 2 mm of proptosis. All ductions except inferiorduction were limited in the left eye (Fig 1). The remainder of his eye examination was normal. A 17-mm flaking, crusting, nonerythematous, nontender, elevated facial skin lesion was present approximately 1 cm lateral to the left side of the mouth (Fig 1). The remainder of the neurologic and physical examination was normal.

Laboratory evaluation—which included a complete blood count, purified protein derivative, VDRL, electrolytes, cryptococcal antigen, hepatitis panel, antinuclear antibody (ANA), rheumatoid factor, and Westergren erythrocyte sedimentation rate—was normal. Repeat culture of the skin lesion was negative for bacteria and fungi. Head and orbit magnetic resonance imaging (MRI) demonstrated an enhancing lesion that extended from the left cavernous sinus to the left posterior orbital apex (Fig 2). The superior ophthalmic vein appeared normal in caliber. Retrospective review of the patient's original CT scan revealed subtle abnormalities in the left cavernous sinus. Biopsy of the skin lesion demonstrated Ki-1 positive anaplastic large cell lymphoma (ALCL). Chest x-ray and abdominal CT demonstrated lung and liver dissemination; bone marrow biopsy and cerebrospinal fluid were negative.

Chemotherapy with methotrexate, vincristine, Adriamycin, and prednisone was instituted. The patient also received 450 cGy of radiation (XRT) to the left orbit and cavernous sinus with shielding of the left eye.

Remarkable improvement of his ocular findings was seen after 3 days of XRT. The visual acuity in his left eye improved to 20/25. The afferent pupillary defect and ptosis resolved completely. Mildly reduced supraduction of the left eye was the only remaining motility disturbance. Systemically, the patient did well following chemotherapy; however, he had a relapse 15 months later. He underwent an autologous peripheral blood stem cell transplant and has since been in remission.

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PATHOLOGIC FINDINGS
A gross examination revealed the tissue to be tan and rubbery, measuring 6 mm × 4 mm × 3 mm. Microscopically, the tissue was densely infiltrated by large cells and foci of necrosis. The cells had abundant cytoplasm, pleomorphic nuclei, and internal wreath-like shapes (Fig 3). The chromatin pattern was vesicular, the nucleoli were frequent, and mitotic activity was abundant and uniformly distributed. Immunohistochemical studies classified the tumor as an ALCL.

DISCUSSION
Lymphoma presenting as cavernous sinus syndrome is rare, with only 10 cases reported in the literature.\textsuperscript{1-6} The typical presentation is characterized by sharp periorcular pain, periorbital paresthesias, and various ophthalmoplegias. Differential diagnoses include infectious cavernous

Fig 1: Pretreatment extraocular motility.
sinus thrombosis (fungus, tuberculosis, bacteria); sarcoidosis; orbital inflammatory disease (ie, Tolosa-Hunt syndrome); primary orbital tumors such as glioma, schwannoma, hemangioma, or lymphoma; and metastatic disease. In children, rhabdomyosarcoma and metastatic neuroblastoma also must be considered. All previously reported cases (n=10) of lymphoma presenting as cavernous sinus syndrome were of the large-cell variety.1-6 The mean age at presentation was 45 years. Unlike large cell lymphoma presenting in other sites, those that presented in the cavernous sinus have a grave prognosis with a mean survival time of 8.7 months.

ALCL is a rare high-grade lymphoma accounting for 2% to 7% of non-Hodgkin’s lymphomas.6 Despite a mean age of 45 years for patients initially presenting with cavernous sinus syndrome, ALCL occurs in all age groups, but more often in children and young adults. The male-to-female ratio is 2.2:1.7 “Ki-1” refers to a Hodgkin’s-disease–associated antigen that is found in Hodgkin’s lymphoma, several other forms of lymphoma, and a small percentage of carcinomas. The proJ10 sentation of ALCL is variable with lymph nodes, skin, bone, and the gastrointestinal tract being the most commonly affected sites. When ALCL involves the orbit, it almost always is due to a dissemination from another primary site. Often, the ALCL skin lesion simulates an insect bite or an abscess, as in our patient. Isolated cutaneous involvement at presentation in ALCL usually is associated with a more favorable prognosis. In two published studies,7,8 all patients (n=11) with skin involvement did well with an indolent course and excellent response to treatment. All but one were alive and well with follow up ranging from 8 to 30 months. The presence of cavernous sinus disease, however, portends a grave prognosis.

This is the first reported case of ALCL presenting simultaneously as cavernous sinus syndrome and a facial skin lesion, and is notable for two reasons: (1) the tumor presented in a child and (2) the tumor masqueraded as an infectious cavernous sinus thrombosis secondary to retrograde spread from a skin abscess, leading to a delay in diagnosis.

CONCLUSIONS

Large-cell lymphoma can present a difficult diagnostic dilemma because of the myriad of possible presentations, ranging from minor-appearing skin involvement to significant CNS dissemination. Although rare in children, large-cell lymphoma should be considered in the differential diagnosis of a nonhealing skin lesion or cavernous sinus syndrome.

REFERENCES