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Cranial nerve VI palsy as an initial presentation of necrotizing sarcoid granulomatosis in a 14-year-old female: Case report and literature review

Mona Doss, DO[a,*], Ronald Araneta III, MD[b], Mary Fiel-Gan, MD[b], Barbara Edelheit, MD[c]

[a] Department of Pediatrics/Medical Education, Connecticut Children's Medical Center, Medical Education, University of Connecticut, 4H, 282 Washington St, Hartford, CT 06106
[b] Department of Pathology, Hartford Hospital, Hartford, CT
[c] Department of Pediatric Rheumatology, Connecticut Children's Medical Center, University of Connecticut, Hartford, CT

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ABSTRACT

Objectives: We describe a case of pediatric necrotizing sarcoid granulomatosis (NSG) presenting with right cranial nerve VI palsy and multiple lung nodules, successfully treated with corticosteroids.
Methods: This is a descriptive case report of one patient with review of the literature.
Results: A 14-year-old Caucasian female presented with complaints of pain on inspiration and dyspnea on exertion, as well as diplopia that was worse with right gaze. The patient presented to our emergency department with persistent diplopia and was found to have stable right cranial nerve VI palsy. CTA showed multiple pulmonary nodules. Despite continued extensive multispecialty work-up, the patient's cranial nerve VI palsy had not resolved, thus tissue confirmation via lung biopsy was performed. Pathologic diagnosis revealed necrotizing sarcoid granulomatosis. The patient was subsequently started on intravenous corticosteroids, which led to the rapid resolution of her presenting symptoms.
Conclusions: Necrotizing sarcoid granulomatosis is a multisystem organ disease that is rare in children. Pathology commonly reveals epithelioid noncaseating granuloma and granulomatous vasculitis with necrosis. We report an unusual presentation involving sixth nerve palsy in a 14-year-old girl. Diagnosis was determined and confirmed by histopathology of a pulmonary nodule biopsy. This is the first case to our knowledge of NSG presenting with cranial nerve palsy in a pediatric patient.

Introduction

Necrotizing sarcoid granulomatosis (NSG) is a rare systemic disease first described in 1973 by Liebow [1]. Prior studies have shown that extrarespiratory findings are more likely in NSG. Inflammatory markers are mildly elevated, whereas ANCA and ACE are negative and normal, respectively [2]. Most common pulmonary findings include solitary or multiple lung nodules and pulmonary infiltrates, while pulmonary function tests are normal and hilar lymphadenopathy is uncommon [2].

Cranial nerve involvement has been described in sarcoidosis and neurosarcoidosis as leptomeningeal involvement or an isolated finding, most commonly involving cranial nerve VII [3,4]. Optic nerve involvement has also been described as secondary to perineural spread of sarcoidosis from sinonasal disease [4]. Cranial nerve VI palsy, as in our case, has not been described in cases of NSG.

The goal of this article is to present a case report of a 14-year-old otherwise healthy female who presented with cranial nerve VI palsy and was found to have necrotizing sarcoid granulomatosis, which to our knowledge is the first of such cases.

Methods

This is a descriptive case report of a 14-year-old female patient with dyspnea and chest pain on exertion, other accompanying clinical findings, work-up, diagnosis and management of NSG with literature review of the clinical presentation, radiologic findings, and histopathology of NSG from multidisciplinary publications.

Case report

We describe a previously healthy 14-year-old female who presented to our emergency department (ED) complaining of

* Corresponding author.
E-mail address: mdoss@connecticutchildrens.org (M. Doss).

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one week of chest pain, which was worse with inspiration and radiated to her shoulder, and difficulty breathing while playing soccer. She had no fever, shortness of breath, cough, hemoptysis, leg pain, nausea, vomiting, headaches, dysuria, or diarrhea. One day prior to visiting our hospital she complained of a sudden onset of binocular diplopia, which was worse when looking to the right.

One week prior, at the onset of her pain, she had been worked up with a chest CTA at an outside hospital ED. The diplopia brought her to her primary care physician, who recommended a brain MRI, which was normal, with no intracranial lesions.

Past medical history showed no recent illness and no chronic medical conditions. She was not on any medication. She had no significant family history of rheumatologic disease. Socially, our patient attended the local high school, played soccer, and owned a cat. She had no recent travel history. An aunt who was incarcerated was recently released.

Test results included negative or normal complete blood count, urinalysis, complement levels (C3 and C4), ACE, ANCA, coagulation factors, prothrombin time (PT), partial thromboplastin time (PTT), and immunoglobulins. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were mildly elevated at 23 mm/h (lab normal range: 0–20 mm/h) and 2.7 mg/L (0–0.49 mg/dL), respectively.

On exam she was found to have stable right cranial nerve VI palsy with no other focal neurologic deficit. Other than the diplopia, she complained of intermittent mild pain with deep inspiration radiating to her right anterior shoulder. This mild pain was relieved by oral non-steroidal anti-inflammatories or use of an ice-pack.

The patient's hospital course involved thorough evaluation from pediatric subspecialties including infectious disease, neurology, oncology, ophthalmology, pulmonology, and rheumatology services. She was found to have a negative interferon gamma release assay, PPD, HIV, fungal cultures, *Aspergillus*, and *Bartonella henselae*. EBV IgG antibody was positive while the rest of the panel was negative, consistent with previous infection or exposure. Bone survey looking for occult malignancy or metastasis as recommended by pediatric oncology was negative. Pulmonary function testing was normal.

CTA of the chest confirmed multiple peripheral nodules of the lung including a subpleural nodule in the right middle lobe, which would later be accessed for biopsy (Figs. 1–3). Brain MRI and MRA showed no signs of lesions, intracranial or space-occupying masses, or vascular abnormalities. Despite continued extensive multispecialty work-up, the patient's VI cranial nerve palsy had not resolved, thus tissue confirmation via lung biopsy was performed.

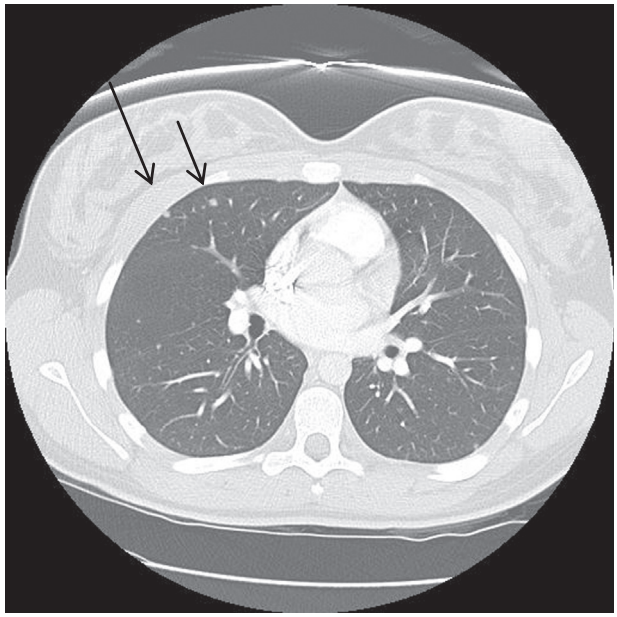

Fig. 2. CTA coronal view displaying a 3-mm (long arrow) and a 1-mm (short arrow) peripheral and inferiorly located pulmonary nodule of the right upper lobe, respectively.

Tissue biopsy from a right lung subpleural nodule confirmed the presence of necrotizing granulomas and vasculitis (Figs. 4–7). Tissue sections showed nodules of coalescent epithelioid granulomas within which were zones of necrosis. A few individual epithelioid granulomas were identified, some with and some without punctate necrosis. In addition, prominent inflammation of veins and arteries were noted, with infiltrating lymphoid cells and epithelioid histiocytes/granulomas. Some of the blood vessels were seen within the necrotic centers of the granulomatous nodules. The process appeared to follow a lymphangitic distribution (subpleural,

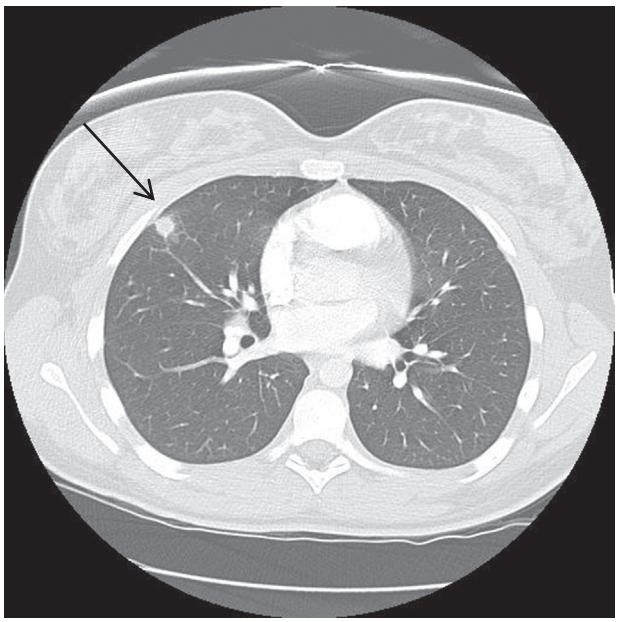

Fig. 1. CTA coronal view displaying an 8-mm pulmonary nodule anteriorly located in the right middle lobe (arrow).

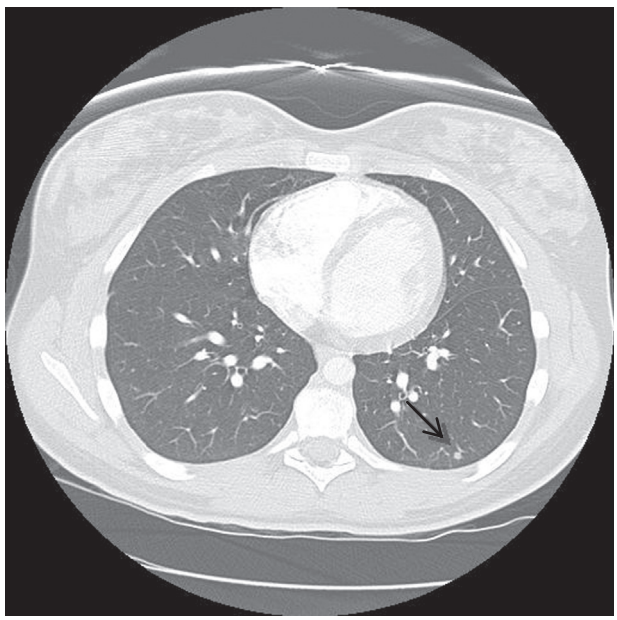

Fig. 3. CTA coronal view displaying a 3-mm posterior pulmonary nodule of the left (arrow).