Clinical picture

Unusual presentation of sarcoidosis—involving testis, spinal cord and the brain

A 37-year-old African American man with history of HTN presented with 3 months history of bilateral leg weakness, headaches, urinary retention and impotence. Initially he started to have headaches. A few weeks later he started complaining of bilateral leg numbness from the umbilicus area to the toes. On presentation his vitals were stable. He had normal cardiopulmonary and abdominal exam. The patient had decreased strength in both lower extremities (3/5 in both legs), with decreased light touch and painful sensation in both lower extremities. He had a wide-based shuffling gait. He was found to have a 1 cm firm lesion in the left testis. MRI spine showed numerous nodules in the dura, involving the cervical region and the cauda equine (Figures 1 and 2). Many of these lesions were also intramedullary. MRI brain showed multiple dural-based and leptomeningeal lesions in the left hemisphere (Figure 3). CT chest revealed cavitary left upper lobe lesion with mediastinal adenopathy. Ultrasound of the left testis revealed a 1.6 x 1.4 cm round hypoechoic mass. Lumbar puncture was suggestive of lymphocytic pleocytosis (WBC of 298/µl, glucose 29 mg/dl, protein 672 mg/dl and elevated IgG index). CSF gram stain and cultures were unremarkable. ESR was 15 mm/h, and CRP was 0.82 mg/dl. Blood and urine cultures were negative. Serum ACE level was elevated at 56 units/l (normal < 49). Biopsy of the left upper lung lesion showed necrotizing granulomatous inflammation. The excisional left testicular wedge biopsy revealed extensive necrotizing and non-necrotizing granulomatous inflammation. Stains for fungi, bacteria and acid-fast bacilli were negative. This suggested the diagnosis of sarcoidosis and he was started on high dose steroids. Before discharge, his numbness improved and he was able to walk without support. At 3 months follow-up he had started working and MRI brain and spine showed significant reduction in size of his spinal and left hemisphere brain lesions.

Sarcoidosis is a multisystem disorder of unknown etiology. A typical presentation is with bihilar

Figure 1. T1 MRI cervical spine showing numerous nodules in the dura, involving the cervical region.

Figure 2. T1 MRI lumbar spine showing numerous nodules in the dura, involving the cauda equine.
lymphadenopathy and erythema nodosum. Sarcoidosis has a 3.8-fold higher incidence among young African Americans compared with Caucasians.\(^1\)

Diagnosis requires three components that include: typical clinical and radiographic findings, exclusion of other diseases with similar presentation and histopathologic evidence of non-caseating granulomas.\(^1\) Differential diagnosis for similar case presentations should include malignancy (lung, testis and intracranial) and infections (disseminated tuberculosis, other fungal or viral etiologies). Sarcoidosis should be considered in the differential diagnosis with CNS and genitourinary masses. Typically, sarcoidosis is localized to the lungs (95%).\(^1\) Genitourinary involvement is very rare (0.2%), with testicular involvement in only 21% of these cases.\(^2,3\) Neurosarcoidosis occurs in 5% of cases, with few having spinal cord involvement (10% of the neurosarcoidosis cases).\(^1,4\) Caseating granulomas, usually seen in tuberculosis, are very uncommon in sarcoidosis.\(^5\) This can lead to misdiagnosis and unwanted side-effects if anti-tuberculosis treatment is started. This is a distinctive case that describes caseating granulomatous sarcoidosis that involves the testis, and the CNS.

Fibrosis and ductal obstruction from scrotal granulomas, and surgical management includingorchidectomy and epididymectomy can lead to azoospermia and infertility.\(^2\) In cases of testicular masses with high suspicion of sarcoidosis, testis preservation should be considered and only a biopsy for diagnosis should be performed. Progression of male genitourinary sarcoidosis is very unpredictable.\(^6\) Thus, patients with signs of oligospermia should be offered sperm banking. Caseating granulomas have a favorable response rate to steroid therapy.\(^7\)

Neurosarcoidosis can spontaneously resolve, have a relapsing-remitting course or be a progressive disease with episodic deteriorations.\(^1\) Corticosteroid therapy is the mainstay of treatment. For patients who deteriorate or cannot tolerate steroid therapy, alternative treatments may include methotrexate, azathioprine, hydroxychloroquine and cyclophosphamide.\(^1\)

Photographs and text from: M.C. Alraies and R. Desai, Cleveland Clinic Lerner College of Medicine of Case Western Reserve University, Cleveland Clinic Foundation, 9500 Euclid Avenue, NA21, Cleveland, OH 44195; A.H. Alraies, Department of Pulmonary Diseases, Critical Care, & Environmental Medicine, Tulane University Health Sciences Center, 1430 Tulane Avenue, SL-9, New Orleans, LA 70112, USA.

email: alraiec@ccf.org

Conflict of interest: None declared.

References


Figure 3. T1 MRI brain showing multiple dural based and leptomeningeal lesions in the left hemisphere.