

Necrotizing sarcoid granulomatosis causing compressive myelopathy at cranio-vertebral junction

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ABSTRACT

Necrotizing Sarcoid Granulomatosis is the term given to a variant of Sarcoidosis in which the granulomas show necrosis and evidence of angitis. This variant of sarcoidosis tends to involve the lungs primarily; however central nervous system (CNS) involvement has also been reported. We report here a case of extra-dural cord compression at cranio-vertebral junction with no other systemic involvement and excellent clinical response to corticosteroids alone. Recognition of this rare variant of sarcoidosis by the pathologist and prompt institution of corticosteroids by the clinician will generally give very gratifying results, as was seen in the present case.

Key words: Cranio-vertebral junction, necrotizing sarcoid granulomatosis, compressive myelopathy

INTRODUCTION

Sarcoidosis is a granulomatous, systemic inflammatory disorder of obscure etiology. Although it appears most commonly in the lungs or lymph nodes, any organ may be affected and clinical presentations are protean. The diagnosis of sarcoidosis is confirmed only by histopathology. Necrotizing sarcoid granulomatosis (NSG) is the term given to a variant in which the granulomas show necrosis and evidence of angitis.^[1] This variant of sarcoidosis also tends to involve the lungs primarily. However CNS involvement has also been reported. We report here a case of extra-dural cord compression at cranio-vertebral junction with no other systemic involvement and excellent clinical response to corticosteroids alone.

CASE REPORT

The present case report is about a 52-year-old female patient was diagnosed to have a posterior cranial-fossa mass in another hospital in January 2010. She underwent a right sub-occipital craniotomy and laminectomy of atlas and excision of mass was carried out. Histopathology

report suggested granulomatous lesion with necrosis. She was commenced empirically on anti-tubercular and anti-fungal medications. She presented to our hospital in May 2010 with progressive quadriparesis, difficulty in deglutition and dyspnea. There were no co-morbidities or any other previous medical illness. Neurological examination revealed a spastic quadriparesis with grade 3/5 motor power. There was bilateral lower cranial nerve palsy. Magnetic resonance imaging of the brain and cranio-vertebral junction with contrast showed dural based extra-axial lesion extending from clivus to C2 with cord compression at craniovertebral (CV) junction with cervicomedullary myelopathic changes [Figure 1]. No lung lesions or lymphadenopathy was detected on computerized tomography of the chest. Serology was negative for human immunodeficiency virus, antinuclear antibodies and anti-neutrophil cytoplasmic antibodies (ANCA). Mantoux test was negative. The patient underwent re-exploration with additional C2 laminectomy. The excised lesion was grayish pink in color; firmly adhered to dura with defined plane of cleavage from the neural tissue. Post-operative period was uneventful.

Routinely processed sections of the excised mass revealed dense fibrocollagenous tissue with mixed inflammatory cellular infiltrate consisting of lymphocytes, plasma cells along with ill-defined granulomas consisting of similar inflammatory infiltrate with many multinucleate histiocytic giant cells. Multiple tiny zones of necrosis were also seen [Figures 2-4]. Sections were stained with periodic acid-Schiff stain and did not reveal any fungal

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organisms. Zeihl-Neelsen stain was negative for acid fast bacilli. Cultures obtained intra-operatively failed to grow any organism. No foreign body was identified by examination with polarized light. In view of previous and current biopsy showing necrotizing granulomatosis and no evidence of infection but with the presence of vasculitis, the case was diagnosed as NSG.

The patient's clinical condition improved rapidly with corticosteroid therapy which was gradually tapered and stopped over next 18 months. After 2 years of follow-up, there is no evidence, clinical or radiographic, of recurrence or progression [Figure 5].

DISCUSSION

We report here a rare case of primary intracranial NSG with no systemic features either in the lung or in any

other organ. Necrotizing sarcoidosis differs from typical sarcoidosis by the presence of significant non-caseating necrosis and granulomatous inflammation consisting of epithelioid histiocytes, lymphocytes and plasma cells, usually with an associated vasculitis.

The term "NSG" was first used by Liebow.^[1] In his report on eleven patients, the following three characteristics were noted:

- Histologically, there were sarcoid like granulomata, a prominent and usually granulomatous vasculitis and varying degrees of necrosis
- Radiologically, prominent nodules in lungs but no enlarged hilar lymph nodes
- Clinically, a benign course even with minimal or no therapy.

It is speculated in this report that NSG may be a variant of angiocentric granulomatosis in which sarcoid-like



Figure 1: Pre-operative neuroimaging studies of brain and cranio-vertebral junction revealing a dural based extra axial lesion extending from clivus to C2 with cord compression at craniovertebral junction

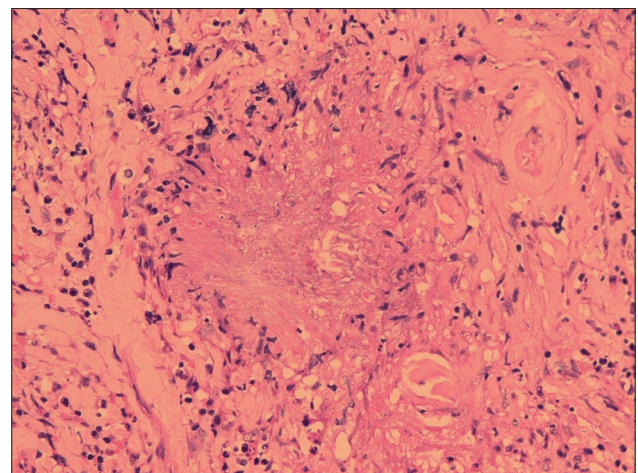


Figure 2: Dense fibrocollagenous tissue infiltrated by mixed inflammatory cells infiltrate and a granuloma with a central region of coagulative necrosis surrounded by palisading of histiocytes (H and E, x200)

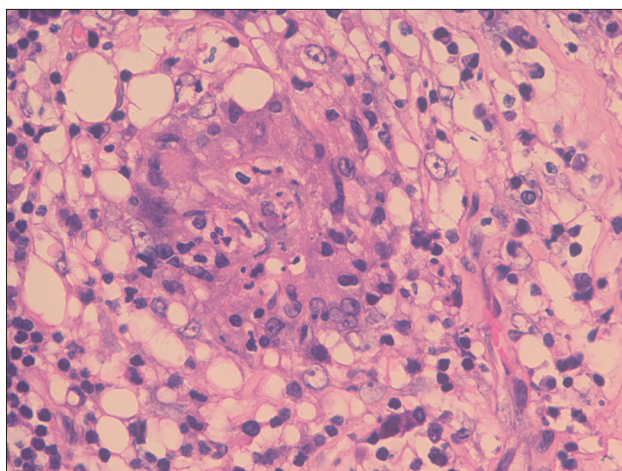


Figure 3: A granuloma with giant cells, histiocytes and karyorrhectic remains of degenerated cells (H and E, x400)

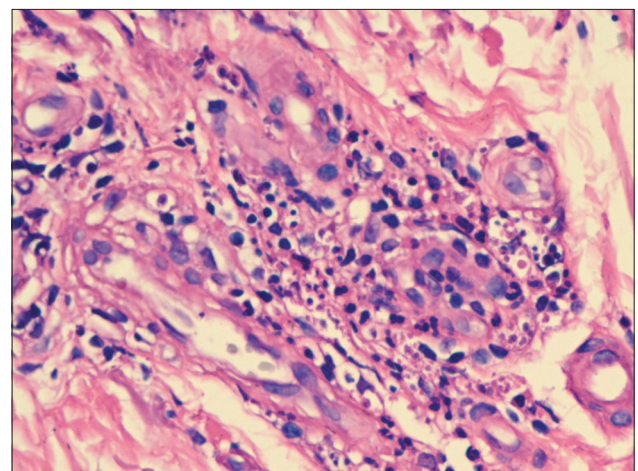


Figure 4: Vasculitis characterized by infiltrate of lymphocytes and histiocytes (H and E, x400)



Figure 5: Neuroimaging 14 months after surgery and therapy showing disappearance of the mass

features are unusually prominent or that this may be a variant of sarcoidosis. Later on Churg *et al.* in a retrospective study of twelve cases concluded that clinical behavior of NSG is unlike other angiocentric granulomatosis and that most patients with this disease can be left untreated or be treated with corticosteroids alone. In the same case-report one of the patient also had uveitis as well as non-caseating granulomas in hilar lymph nodes.^[2] This strengthens the belief that NSG is a variant of sarcoidosis. In another case report, appearance of NSG in left temporal lobe of brain has been documented in a typical case of sarcoidosis, diagnosed several years ago on the basis of parotid gland biopsy.^[3] These observations suggest that NSG is more likely a variant of sarcoidosis rather than a type of ANCA-negative granulomatous angitis.

The differential diagnosis of necrotizing granulomas includes chronic infection (typical/atypical mycobacteria and fungi), Wegener's granulomatosis and foreign body granulomatous inflammation. Diagnosis of sarcoidosis, whether necrotizing or non-necrotizing, should be made only after these causes are excluded by histochemical stains and cultures.

Wegener's granulomatosis is histologically characterized by liquefactive and/or coagulative necrosis, a large number of eosinophils, scanty benign appearing lymphocytes and multinucleated giant cells that generally do not form well-defined granulomas and a destructive angitis involving arteries and veins. Demonstration of cytoplasmic-ANCA by immunofluorescence or anti-Pr3 antibodies by ELISA constitutes a reliable serologic marker for Wegener's granulomatosis with sensitivity of about 90%, in biopsy proven Wegener's granulomatosis. Similarly, chronic

infection (typical and atypical mycobacterium and fungi) and foreign body granulomatous inflammation can be excluded by histochemical stains, cultures, polarizing microscopy and by molecular studies.

Other than the histologic analysis, clinical behavior of patients with NSG is more like those with typical sarcoidosis, i.e. a long clinical course with a favorable response to steroids. This also rules out any underlying infectious cause of necrotizing granulomatosis, particularly when supported by negative cultures.

The first case of primary CNS-NSG in the absence of systemic sarcoidosis was reported by Tobias *et al.*^[4] The lesion involved cranial base resembling an en Plaque Sphenoid Wing Meningioma. Subsequently, Markert *et al.* have reported three cases of NSG with diverse presentation as diffuse leptomeningeal involvement, a cerebello-pontine angle tumor and a thoracic spinal cord lesion.^[5] A review of literature reveals no report of NSG at CV junction.

The present report emphasizes the need to keep NSG on the list of differential diagnosis of necrotizing granuloma in the brain even without systemic manifestation. Necrotizing sarcoidosis, like typical sarcoidosis, is a diagnosis of exclusion and should not be made until all special studies including cultures have been performed to rule out an infectious cause. Recognition of this rare variant of sarcoidosis by the pathologist and prompt institution of corticosteroids by the clinician will generally give very gratifying results, as was seen in the present case.

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