(history, fever, skin rash) and a positive serum IFA test to anti-*O. tsutsugamushi* IgM antibody (1:160). Diagnostic IFA must meet the following criteria: (i) the total antibody titer for Karp, Kato, and Gilliam strains of *O. tsutsugamushi* must increase fourfold (or greater) in paired serum samples, or (ii) the IgM antibody titer must be $\geqslant$1:80.[8,9] In this patient, therefore, diagnosis of acute scrub typhus infection was conclusive by clinical and serological evidence.

Several movement disorders, including parkinsonism, may occur as a consequence of CNS infection, as either an acute or chronic manifestation.[10,11] CNS involvement, ranging from aseptic meningitis to encephalomyelitis, is a known complication of scrub typhus infection.[4–6] Altered mental status, tremor, restlessness, and focal neurologic signs, including diplopia, gaze palsy, nystagmus, dysarthria, dysphagia, quadriparesis, and sensory dysfunction, can be observed. Silpapojakul et al. reported that nine of 72 scrub typhus patients had neurological symptoms; most had varying degrees of delirium, decreased levels of consciousness, and/or behavioral changes; only one had focal neurological signs. However, to our knowledge, this is the first reported patient with transient parkinsonism with myoclonus secondary to acute scrub typhus infection.

Definitive diagnosis of infection-related encephalitis could not be made in the current patient, because lumbar puncture was not performed due to lack of consent. Brain MRI was not abnormal, however, and according to the clinical course and serological evidence, the scrub typhus encephalitis diagnosis was favored, with parkinsonism (mask-like face, bradykinesia, tremor, rigidity, small-stepped gait) and myoclonus as main features. Therefore, doxycycline, clonazepam and amantadine were administered for infection and symptomatic control. The fever subsided first, and neurological symptoms improved gradually. Responses to treatment provided further affirmation of the diagnosis of scrub typhus-related parkinsonism with myoclonus.

The involuntary movement developed 2 days after onset of fever. This time course favors infection *per se,* rather than post-infectious immunological mechanisms, as the etiology of parkinsonism with myoclonus. Post-infectious immunological mechanisms leading to neurological symptoms usually require 1 to 2 weeks, as previously reported for patients with acute disseminated encephalomyelitis or Guillain-Barré syndrome.[12–14]

We report this case of parkinsonism with myoclonus as a rare presentation of scrub typhus infection, and suggest that early diagnosis and timely initiation of appropriate therapy can reduce ensuing complications and improve prognosis.

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Solitary juvenile xanthogranuloma mimicking intracranial tumor in children

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ABSTRACT

Juvenile xanthogranuloma (JXG) is primarily a benign cutaneous disorder of non-Langerhans hystiocytic proliferation. Systemic involvement occurs in 4% of patients; isolated central nervous system (CNS) lesions are rare. We report solitary CNS-JXG lesions in two patients. A 3.5-year-old boy with a parietal-occipital lesion underwent total resection with no surgical morbidity and no recurrence at 16-month follow-up. A 3.5-year-old girl underwent subtotal resection of a tumor extending from the left Meckel's cave and invading the cavernous sinus and left orbit with extensive cranial nerve involvement. Tumor regrowth with leptomeningeal spread at 9-month and 12-month follow-up was managed with steroids and chemotherapy (vinblastine and later cladribine). We present our experience and review the literature pertaining to rare reports of solitary CNS-JXG.

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1. Introduction

Juvenile xanthogranuloma (JXG) is primarily a benign cutaneous disorder of non-Langerhans hystiocytic proliferation, usually confined to childhood. Most patients present in the first year of life.[1] An underlying genetic defect, viral exposure, or dysregulation of inflammatory mediators may have a causative role;[2] however, the etiology of JXG has not been fully elucidated. Histological features include "Touton cells", which are multinucleated histiocytes with finely vacuolated foamy, xanthomatous cytoplasm. The diagnosis is confirmed by positive staining for factor XIIIa, fascin, and CD68, with no reaction to S100 or CD1a.[2,3]

Most patients with JXG present with single or multiple yellow–red nodular skin lesions, commonly in the head and neck region. Cutaneous JXG is usually benign and self-limiting.[4] In approximately 4% of patients,[1] systemic evaluation reveals involvement of other organs, including the liver, spleen, lungs, kidneys, eye, subcutaneous soft tissue, bones, and central nervous system (CNS). Patients with systemic involvement may have a severe or even fatal clinical course, despite the biologically benign nature of these lesions.[4,5] A smaller subgroup of patients presents with isolated CNS disease and no systemic involvement. This subgroup appears unique in patient characteristics, treatment, and outcome. Due to the rarity of isolated CNS disease and the short follow-up periods reported, the preferred treatment and overall prognosis of these patients remain unclear.

We report our experience with two patients presenting with solitary intracranial JXG lesions, and review reported patients with isolated intracranial disease. Our Institutional Review Board waived the requirement for informed consent for this retrospective file review.

2. Case reports

2.1. Patient 1

A 3.5-year-old boy, previously healthy aside from neonatal jaundice, was admitted to the hospital after experiencing, in a 6-month period, two generalized seizures with gaze shift and tonic-clonic convulsions. Both seizures were prolonged for a few minutes and followed by a postictal period. On examination, the patient was fully conscious. His physical and neurological examinations as well as laboratory blood test results were unremarkable.

Two months after the second seizure head CT scan and MRI revealed a well-demarcated space-occupying lesion in the right parieto-occipital cortex, measuring 23 mm in diameter. The lesion exerted a mild mass effect and was surrounded by mild edema, with mild homogenous enhancement on T1-weighted MRI with gadolinium (Fig. 1A–C).

High-dose corticosteroid and anticonvulsive treatment were administered. The patient underwent right parietal craniotomy with gross total resection. After surgery he was asymptomatic, with no new neurological deficit.

Pathological examination revealed histiocytic infiltrates invading the brain parenchyma, spreading along blood vessels through the Virchow-Robin spaces. Most cells had small purple nuclei, but some revealed a pleomorphic appearance. Nuclei were surrounded by a large amount of well-circumscribed cytoplasm. Spindle-shaped cells were also seen. No giant cells or necrosis were evident. A few mitoses were present. Immunohistochemical stains were positive for CD68 and factor XIIIa, but were negative for S100 and CD1a. This appearance and phenotype were in keeping with the diagnosis of JXG.

Due to the lack of specific and localizing systemic complaints, the patient underwent only a partial systemic workup, including abdominal ultrasound and bone scan, both found to be normal; a full neuro-axis MRI and lumbar puncture were not performed. MRI performed 8 months after surgery demonstrated good tumor resection with no evidence of residual or tumor regrowth (Fig. 1D–E). The patient was followed with close observation, without administration of adjuvant therapy. At the 28-month follow-up, there was no evidence of tumor recurrence or systemic involvement, and no new neurological deficit.

2.2. Patient 2

A 3.5-year-old girl, previously healthy, was admitted to the hospital due to left eye ptosis and diplopia of 2.5-months duration.

On examination in the outpatient clinic, the patient had left partial ptosis and cranial nerve (CN) III paresis, with normal visual acuity. With the exception of these deficits, her physical and neurological examinations were unremarkable. Laboratory blood test results were within normal limits. Steroids were administered with partial clinical improvement, but minimal left ptosis and weakness of CN IV were noted on repeat examination.

Head CT scan and MRI revealed a well-demarcated 35-mm extra-axial dumbbell-shaped lesion extending from the left Meckel's cave to the cavernous sinus and posterior fossa (Fig. 2). The mass was hyperintense on T1-weighted MRI with weak enhancement after gadolinium injection. A prominent mass effect was noted on the midbrain and left temporal lobe. The anterior part of the mass invaded the orbit, compressing the optic nerve and lateral rectus muscle and appeared to widen the foramen rotundum and ovale. The lesion contained two hypointense oval areas, suggesting cystic

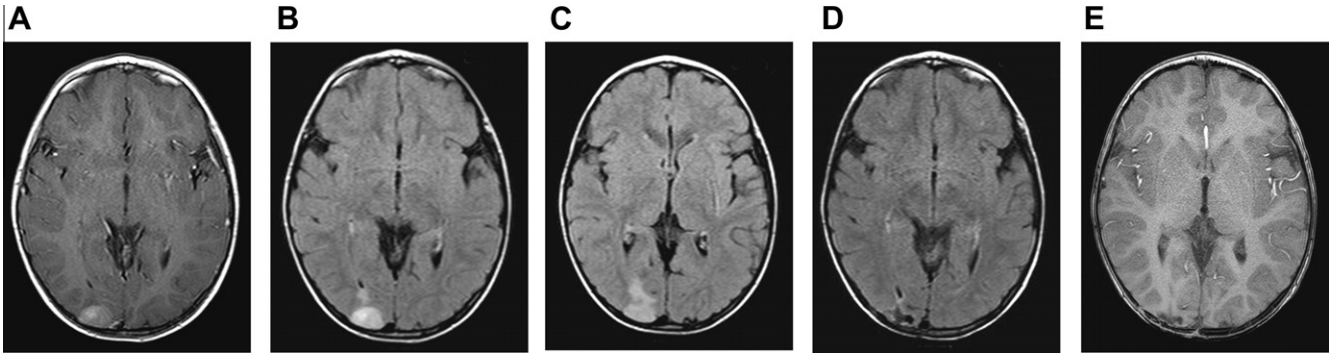

Fig. 1. Axial imaging studies in a 3.5-year-old male (patient 1): (A–C) preoperative T1-weighted, gadolinium-enhanced MRI showing a round hyperintense enhancing mass in the right parieto-occipital region; and (B, C) preoperative T2-weighted fluid-attenuated inversion recovery (FLAIR) MRI; and (D, E) postoperative (D) FLAIR and (E) T1-weighted, gadolinium enhanced MRI showing total resection of the tumor.