Neurosarcoid Presents Differently in Children Than in Adults

Robert J. Baumann, MD, and William C. Robertson, Jr, MD

ABSTRACT. Background. Neurosarcoid is seldom recognized in children. In the absence of any large pediatric series, it has been assumed that the presenting signs and symptoms are identical in adults and children.

Objective. To test the hypothesis that childhood neurosarcoid differs in presenting signs and symptoms from neurosarcoid in adults.

Methods. We tabulated the initial neurologic signs and symptoms in all reported cases of childhood sarcoid with evidence of central nervous system involvement. These data then were compared with published studies of adult neurosarcoid.

Results. Twenty-nine cases (from the English, French, and German literature) had descriptions of presenting signs and symptoms. Ages were 3 months to 18 years; 48% (14 of 29) presented before 13 years. Seizures were the most common presenting symptom (38%, 11 of 29), and 73% of these children (8 of 11) were <13 years old at presentation. Twenty-one percent (6 of 29) had cranial nerve involvement at presentation, and all were ≥12 years old. Twenty-one percent (6 of 29) had hypothalamic dysfunction. Five children presented with headache, 4 with motor signs, and 3 with papilledema. Twenty-four percent (7 of 29) had mass lesions on imaging.

Conclusions. Children with neurosarcoid present differently than do adults. Children are more likely to have seizures, less likely to have cranial nerve palsies, and perhaps more likely to have a space-occupying lesion. Our analysis of the cases available for review in the published literature suggests that children evolve to an adult pattern as they progress through adolescence. Pediatrics 2003;112:480–486. URL: http://www.pediatrics.org/cgi/content/full/112/6/480; adolescence, child, cranial nerve injuries, manifestations neurologic, sarcoidosis, seizures.

ABBREVIATIONS. CNS, central nervous system; CSF, cerebrospinal fluid; MRI, magnetic resonance imaging; ACE, serum angiotensin-converting enzyme; CT, computed tomography.

Physicians seldom include sarcoid in the differential diagnosis of progressive encephalopathies of childhood.1,2 This apparently occurs because sarcoid is seldom diagnosed in the young and is therefore considered rare.3,4 The omission is surprising, because sarcoid is both treatable and known to have protean manifestations. Moreover, if untreated, CNS sarcoid can result in death or permanent disability.

In the absence of any large pediatric series, it has been assumed that the presenting signs and symptoms of CNS sarcoid are identical in children and adults.3,5 We recently cared for a child who did not fit the usual adult pattern and decided to reexamine this premise. We searched for all reported cases in the world literature and analyzed them for presenting signs and symptoms. Our review suggests that prepubertal children with neurosarcoid present differently than adults, and this pediatric pattern evolves to an adult pattern during puberty.

CASE REPORT

After 3 days of headache and malaise, a previously healthy 9-year-old white girl from rural Kentucky had a generalized tonic-clonic seizure. Cerebrospinal fluid (CSF) examination revealed a white blood cell count of 350 per mL (52% lymphocytes and 48% polymorphonuclear cells), normal glucose, and protein of 68 mg per dL. Routine cultures were sterile. After intravenous phenytoin there were no additional seizures. Her hospital course was characterized by fever to 101°F, confusion, and headache. Electroencephalograms showed symmetrical slowing compatible with a nonspecific, diffuse encephalopathy. Precontrast magnetic resonance imaging (MRI) of the head was unremarkable, but gadolinium-enhanced images revealed 2 irregular areas of enhancement involving the anterior aspect of the left frontal lobe. After 10 days she was afebrile, symptom-free, and discharged on phenytoin with a presumptive diagnosis of viral encephalitis.

After 3 medically uneventful months (though marred by poor school performance), the child became acutely confused and had another generalized tonic-clonic seizure. On readmission, routine laboratory studies were unremarkable. Gadolinium-enhanced MRI showed diffuse, fine punctate lesions with increased signal from cortical gyri on the late T2 sequence. Magnetic resonance angiogram was unremarkable. The erythrocyte sedimentation rate was 11, and the phenytoin level was therapeutic. Numerous laboratory studies including serum aldolase, antineutrophil cytoplasmic and perinuclear antibodies, antinuclear antibodies, anti-Smith antibodies, factor V Leiden, anti-smooth muscle antibodies, and anticardiolipin antibodies were negative. Homocysteine, protein C, and protein S were unremarkable. Spinal fluid analysis revealed 4 mononuclear cells, 1 red blood cell, glucose of 64 mg per dL, and protein of 53 mg per dL. CSF cultures including those for fungi.

From the Departments of Neurology and Pediatrics, University of Kentucky, Lexington, Kentucky.

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Address correspondence to Robert J. Baumann, MD, Kentucky Clinic, L409, Speed Sort 0284, Lexington, KY 40536-0284. E-mail: baumann@uky.edu

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tuberculosis, and viruses were sterile. Other CSF studies including venereal disease research laboratory test, pyruvate, oligoclonal bands, and myelin basic protein were unremarkable. Valproic acid was added to the patient’s medication program. She improved and was discharged after 6 days of hospitalization.

After discharge the patient had headaches daily. Over the next 3 weeks she became unsteady and had difficulty remembering numbers. After 2 generalized seizures the child was readmitted. Examination revealed confusion, a right 6th-nerve palsy, extensor plantar responses, and truncal ataxia. Multiple laboratory studies again were normal. Brain MRI now showed large areas of increased T2 signal outlining multiple, swollen cerebral cortical gyri and the caudate nuclei (Fig 1). The dot-like enhancement previously seen with gadolinium enhancement was more prominent and again corresponded to areas of increased T2 signal (Fig 2).

The child’s headache was unrelenting. She developed bilateral 6th-nerve palsies, left lower facial weakness, and slow speech. Brain and meningeal biopsy revealed discrete noncaseating epithelioid granulomas consistent with sarcoid (Fig 3). The serum angiotensin-converting enzyme (ACE) level was 15 international units (normal: 41–126), and chest radiograph was normal.

The child had marked clinical improvement within 48 hours of high-dose steroid therapy. After 5 months of tapering doses of prednisone, MRI findings had completely resolved, and steroids were discontinued. There were no cranial nerve findings or focal neurologic deficits; however, the child did have frontal lobe indifference and mild cognitive difficulties. Four years later these cognitive deficits have persisted, but there has been no recurrence of sei-

METHODS

We attempted to identify all reported childhood cases of neurosarcoid. We searched PubMed for the common manifestations of

Fig 1. Axial T2-weighted image demonstrates large areas of increased signal involving primarily frontal cortex and subcortical white matter.

Fig 2. Coronal MRI with gadolinium shows enhancing punctate lesions within sulci and cortical gray matter.

Fig 3. Biopsy specimen shows noncaseating granuloma with multinucleated giant cells in the frontal cortex (hematoxylin/eosin, original magnification ×100).
sarcoid in childhood (pulmonary and artritic) as well as for
neurosarcoid. Each reported case was examined for evidence of
CNS manifestations, and reference lists were scanned for reports
of additional cases. We also reviewed child neurology and neu-
oradiology textbooks for cases described in the texts but not
reported otherwise.

RESULTS

Including our patient, there were 29 cases of child-
hood neurosarcoid with descriptions of presenting
signs and symptoms (Table 1). An additional 15 cases
lacked sufficient data about presenting signs and
symptoms and were omitted. Because on rereview
of the original articles we could not find evidence of
neurologic involvement, we omitted 2 of the 23 chil-
dren listed by Weinberg et al in their 1983 review of
pediatric neurosarcoid. In the paper by Beier and
Lahey7 we could not find the child with a 7th-nerve
palsy referenced by Weinberg et al. In addition, in
the 1956 series collected by McGovern and Merritt8
we were unable to identify the child tabulated by
Weinberg et al as having “facial dysphagia.”

Cases were found in the English, French, and Ger-
man language literature. Only 13 cases were specif-
ically published in peer-reviewed journals as neuro-
sarcoid. Eleven other children were reported as hav-
ing systemic sarcoid, but our review of the pub-
lished details indicated nervous system involvement.
Two other cases were reported as examples of hypo-
thalamic complications of sarcoid. Another was
found in an article on the radiologic manifesta-
tions of sarcoid, 1 was reported as an ophthalmologic com-
plexion of sarcoid, and the final case was dis-
covered in a child neurology textbook. Ages ranged
from 3 months to 18 years with 48% (14/29) present-
ing before 13 years. Seizures were the most common
presenting symptom (38% of subjects, 11 of 29). Of
the children presenting with seizures, 73% (8 of 11)
were <13 years old at presentation. Twenty-one per-
cent (6 of 29) had cranial nerve findings at presenta-
tion that appeared to be caused by direct nerve in-
volvement and not as a secondary effect such as
6th-nerve palsies associated with increased intracra-
nial pressure. All children with cranial nerve palsies
were ≥12 years old. Only 1 child had involvement of
multiple cranial nerves (age 13). Among the others, 2
had optic nerve and facial nerve palsies, and 1 had a
superior oblique palsy. Twenty-one percent (6 of 29)
evidenced hypothalamic dysfunction with diabetes
insipidus (4 of 6), short stature (3 of 6), and sexual
immaturity (2 of 6). Hypothalamic dysfunction
showed no age predilection. Five children presented
with headache, 4 with motor signs, and 3 with pap-
illedema.

Twenty-four percent (7 of 29) had mass lesions or
focal edema on imaging: One 8-year-old child with
seizures had a “ring cortical lesion” on computed
tomography (CT)9; our case (age 9) had papilledema
and swollen frontal gyri on MRI; one 12-year-old
child had nodular lesions in the optic chiasm and
right sylvian fissure on MRI10; a 13-year-old child
had a pontine mass on MRI11; a 6-year-old child
had hemiparesis and a left frontal mass12; a 17-year-old
child with seizures had a temporal mass on MRI13;
and an 18-year-old child had hemiparesis and pap-
illedema with a mass on angiogram.14

DISCUSSION

Our patient presented with an encephalopathy
that lacked unique or distinguishing features, similar
to many of the cases of childhood neurosarcoid re-
ported in the literature. She lacked evidence of pul-
monary sarcoid, had no adenopathy, and had a nor-
mal ACE level. Neurosarcoid was not considered in
the differential diagnosis, although there were hints
as to the correct diagnosis on MRI (punctate, enhanc-
ing lesions with gadolinium). She developed signs of
increased intracranial pressure (papilledema and bi-
lateral 6th-nerve palsies), leading to a biopsy of the
mass-like, intracranial lesion and the correct diagno-
sis. Her response to corticosteroid therapy was prompt
and probably life-saving. She has been off
medication and in remission for >4 years.

Sarcoid is a systemic disorder of unknown cause
characterized by noncaseating granulomas.15 It
is well known in adults for its protein features includ-
ing abnormal chest radiographs and progressive
multiorgan failure.15 In adults, “the lungs and tho-
racic lymph nodes are almost always involved; most
patients report acute or insidious respiratory prob-
lems.”15 Neuropathy, especially facial nerve palsy, is
the most common neurologic complication among
adults.16-19 Among older children sarcoid is primar-
ily viewed as a pulmonary or ocular disorder,10 with
the triad of skin, joint, and eye disease occurring in
younger children.20-22 In younger children, systemic
sarcoid differs clinically from the adult form of the
disease and can mimic various disorders unique to
childhood.23 Adult and childhood sarcoid also differ
epidemiologically. Children do not appear to show
the female predominance noted in adults24; addi-
tionally, in the United States, they do not seem to
demonstrate the higher prevalence in African Americans
seen in adult populations.15,22 Although our patient
lives in a rural area, it is unknown if American
children as a group show the rural predominance
noted in American adults with this disorder.15,24

In adults, neurosarcoid is estimated to occur in 5%
to 10% of patients with systemic sarcoid, although
this may be an underestimate.25,26 Adult neurosar-
coid characteristically presents with cranial nerve
palsies, most commonly involving the 7th nerve.19,27
A recent review reported that 50% of adults pre-
sented with cranial nerve palsies, 30% with head-
aches, and 10% each with seizures, pituitary dys-
function, sensory deficits, neuropsychological
deficits, and cerebellar signs (patients could have >1
sign on presentation).19,27 In adults, both seizures26,29
c and CNS involvement26 have been considered mark-
ers of poor outcomes.

Our review of the literature finds a very different
pattern for childhood neurosarcoid. As with other
encephalopathies, prepubertal children are likely to
present with seizures. The adult pattern of cranial
nerve involvement appears only after puberty. Evi-
dence of hypothalamic dysfunction is also common
in children, with patients showing growth failure,
diabetes insipidus, and lack of sexual maturation.
<table>
<thead>
<tr>
<th>No.</th>
<th>Age at CNS Onset (y)</th>
<th>Sex</th>
<th>CNS Presentation</th>
<th>Other Clinical Findings</th>
<th>CNS Imaging</th>
<th>Diagnostic Tests</th>
<th>Pathology</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3/12</td>
<td>F</td>
<td>Convulsions</td>
<td>Hepatosplenomegaly and skin rash</td>
<td>N/A</td>
<td>CSF: pleocytosis</td>
<td>Autopsy: sarcoid lesions in falx cerebri and meninges</td>
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<td>2</td>
<td>2</td>
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<td>Short stature</td>
<td>Lymphadenopathy</td>
<td>N/A</td>
<td>Abnormal chest x-ray?</td>
<td>Autopsy: granulomas in multiple tissues including the pituitary</td>
<td>36</td>
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<td>3</td>
<td>6</td>
<td>F</td>
<td>Intermittent confusion, focal seizures, and papilledema</td>
<td>Fever, rash, lymphadenitis, and uveitis</td>
<td>N/A</td>
<td>CSF: pleocytosis, increased pressure, and increased protein</td>
<td>Lymph node biopsy positive</td>
<td>30</td>
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<td>4</td>
<td>7</td>
<td>M</td>
<td>Seizures</td>
<td>Arthritis, rash, and uveitis</td>
<td>MRI: disseminated hyperdense areas</td>
<td>N/A</td>
<td>Skin biopsy positive</td>
<td>20</td>
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<td>5</td>
<td>8</td>
<td>F</td>
<td>Seizure</td>
<td>Wheezing, macular skin lesions, and adenopathy</td>
<td>Ring lesion in cortex on CT</td>
<td>Chest x-ray: disseminated lesions in lungs</td>
<td>Lymph node biopsy positive</td>
<td>9</td>
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<tr>
<td>6</td>
<td>9</td>
<td>F</td>
<td>Polydipsia and polyuria</td>
<td>Shortness of breath and rash</td>
<td>N/A</td>
<td>Chest x-ray: disseminated lesions throughout the lungs</td>
<td>Skin biopsy positive</td>
<td>8</td>
</tr>
<tr>
<td>7</td>
<td>9</td>
<td>F</td>
<td>Headache and seizures</td>
<td>Isolated CNS sarcoid</td>
<td>MRI: increased T2 of caudate and multiple cortical gyri</td>
<td>Brain biopsy positive</td>
<td>This study</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>9</td>
<td>F</td>
<td>Seizures and somnolence</td>
<td>Arthritis, rash, and uveitis</td>
<td>N/A</td>
<td>CSF: elevated protein</td>
<td>Skin biopsy positive</td>
<td>20</td>
</tr>
<tr>
<td>9</td>
<td>9</td>
<td>F</td>
<td>Seizures and motor signs</td>
<td>Arthritis, rash, and uveitis</td>
<td>N/A</td>
<td>N/A</td>
<td>Autopsy: granulomas in brain and multiple other organs</td>
<td>20</td>
</tr>
<tr>
<td>10</td>
<td>11</td>
<td>F</td>
<td>Seizure with slow intellectual and motor deterioration</td>
<td>Isolated CNS sarcoid</td>
<td>N/A</td>
<td>N/A</td>
<td>Autopsy: granulomas in brain and lungs</td>
<td>37</td>
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<tr>
<td>11</td>
<td>11</td>
<td>M</td>
<td>Stiff neck and headache Papilledema and headache</td>
<td>Fever, chills and weight loss</td>
<td>N/A</td>
<td>Kveim test positive</td>
<td>Lymph node and liver biopsies positive</td>
<td>38</td>
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<td>12</td>
<td>12</td>
<td>M</td>
<td>Papilledema and headache</td>
<td>Parotitis</td>
<td>N/A</td>
<td>Chest x-ray: hilar lymphadenopathy</td>
<td>Lymph node biopsies positive</td>
<td>31</td>
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<tr>
<td>13</td>
<td>12</td>
<td>F</td>
<td>Asymptomatic</td>
<td>Lymphadenopathy</td>
<td>MRI: nodular lesions in optic chiasma and right sylvian fissure, below right lenticular nucleus</td>
<td>Chest x-ray: hilar adenopathy</td>
<td>Cervical mass biopsy</td>
<td>10</td>
</tr>
<tr>
<td>14</td>
<td>12</td>
<td>F</td>
<td>Decreased vision (unilateral)</td>
<td>Enlarged lymph nodes, liver, and spleen and conjunctivitis</td>
<td>N/A</td>
<td>Chest x-ray: wide upper mediastinum</td>
<td>Inguinal node positive</td>
<td>39</td>
</tr>
<tr>
<td>15</td>
<td>13</td>
<td>F</td>
<td>Diabetes insipidus and sexual immaturity Superior oblique ocular palsy</td>
<td>Adenopathy</td>
<td>N/A</td>
<td>Chest x-ray: hilar adenopathy</td>
<td>Supraclavicular node positive</td>
<td>39</td>
</tr>
<tr>
<td>16</td>
<td>13</td>
<td>M</td>
<td>Superior oblique ocular palsy</td>
<td>Panuveitis (bilateral)</td>
<td>MRI: multiple CNS masses</td>
<td></td>
<td></td>
<td>11</td>
</tr>
<tr>
<td>17</td>
<td>13</td>
<td>F</td>
<td>Focal seizures and headache</td>
<td>Fever, malaise, vomiting, and weight loss</td>
<td>CT normal</td>
<td>EEG showed left midtemporal seizure focus; CSF: 1 white blood cell, and protein: 49</td>
<td></td>
<td>7</td>
</tr>
<tr>
<td>18</td>
<td>13</td>
<td>F</td>
<td>Multiple cranial nerve signs</td>
<td>Parotiditis</td>
<td>N/A</td>
<td>Chest x-ray: hilar adenopathy</td>
<td>Parotid biopsy positive</td>
<td>25 and 40</td>
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<td>No.</td>
<td>Age at CNS Onset (y)</td>
<td>Sex</td>
<td>CNS Presentation</td>
<td>Other Clinical Findings</td>
<td>CNS Imaging</td>
<td>Diagnostic Tests</td>
<td>Pathology</td>
<td>Reference</td>
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<tr>
<td>19</td>
<td>13</td>
<td>F</td>
<td>Diabetes insipidus</td>
<td>Vitreous hemorrhage</td>
<td>Skull x-ray normal</td>
<td>Chest x-ray: hilar adenopathy; CSF: 8 lymphocytes, protein = 76 mg%</td>
<td>Lymph node biopsy positive</td>
<td>41</td>
</tr>
<tr>
<td>20</td>
<td>13</td>
<td>F</td>
<td>Right-sided facial weakness and right visual loss</td>
<td>Bilateral uveitis, rash, and lymphadenopathy</td>
<td>N/A</td>
<td>Chest x-ray: hilar adenopathy</td>
<td>Lymph node positive</td>
<td>42</td>
</tr>
<tr>
<td>21</td>
<td>15</td>
<td>F</td>
<td>Bilateral facial palsies without parotid swelling</td>
<td>Uveitis</td>
<td>N/A</td>
<td>N/A</td>
<td>Iris and lymph node biopsies positive</td>
<td>43</td>
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<tr>
<td>22</td>
<td>15</td>
<td>M</td>
<td>Polyuria, polydipsia, and short stature</td>
<td>Cervical and axillary adenopathy</td>
<td>N/A</td>
<td>Water-deprivation specific gravity of 1.006</td>
<td>Cervical lymph node positive</td>
<td>44</td>
</tr>
<tr>
<td>23</td>
<td>&lt;16</td>
<td>M</td>
<td>Headache, vomiting, and somnolence</td>
<td>Arthritis, rash, and uveitis</td>
<td>N/A</td>
<td>N/A</td>
<td>Skin biopsy positive</td>
<td>20</td>
</tr>
<tr>
<td>24</td>
<td>16</td>
<td>M</td>
<td>Monocular visual loss</td>
<td>Radionuclide brain scan negative</td>
<td>Radionuclide scan &amp; EEG: left frontal abnormality</td>
<td>Chest x-ray: hilar adenopathy</td>
<td>Optic nerve granuloma</td>
<td>45</td>
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<tr>
<td>25</td>
<td>16</td>
<td>M</td>
<td>Hemiparesis and papilledema</td>
<td>Isolated CNS sarcoid</td>
<td>N/A</td>
<td>Joint mass</td>
<td>Brain biopsy positive</td>
<td>12</td>
</tr>
<tr>
<td>26</td>
<td>16</td>
<td>M</td>
<td>Failure of growth and of sexual maturation</td>
<td>Seizures</td>
<td>N/A</td>
<td>MRE enhancing temporal mass</td>
<td>Joint mass: noncaseating granuloma</td>
<td>41</td>
</tr>
<tr>
<td>27</td>
<td>17</td>
<td>M</td>
<td>Seizures</td>
<td>Seizures</td>
<td>Cough and adenopathy</td>
<td>Normal pneumoencephalogram</td>
<td>“Histologic confirmation”</td>
<td>13</td>
</tr>
<tr>
<td>28</td>
<td>17</td>
<td>M</td>
<td>Seizures</td>
<td>Isolated CNS sarcoid</td>
<td>Mass on Technetium 99m brain scan and angiogram</td>
<td>Kveim test negative</td>
<td>Autopsy: leptomeningeal and grey matter granuloma</td>
<td>46</td>
</tr>
<tr>
<td>29</td>
<td>18</td>
<td>M</td>
<td>Hemiparesis and papilledema</td>
<td>Isolated CNS sarcoid</td>
<td>Mass on Technetium 99m brain scan and angiogram</td>
<td>Kveim test negative</td>
<td>Brain mass: noncaseating granuloma</td>
<td>14</td>
</tr>
</tbody>
</table>

EEG indicates electroencephalography.

* Child had onset of sarcoid at 2 months of age and was reported at 16 years of age. Age at onset of CNS involvement was not given.
Most surprising of all is the large number of children with mass-like lesions on imaging. We found 7 cases not including 2 children with papilledema who were reported in 1937 and in 1969 before the advent of modern imaging. It is also striking that 1 of the 7 children had no neurologic symptoms, and the mass-like lesion was discovered by MRI done “for completeness.” Although adults with seizures and other intracranial lesions respond poorly to therapy and have a higher rate of chronic handicap, this does not seem to be true of our 9-year-old patient or of other children. Previous authors have regarded mass-like lesions as rare. It is unclear if they are wrong and this is an important and previously unrecognized feature of childhood neurosarcoid or if this is an artifact of a survey of reported cases, with unusual findings being more likely to be published.

Sarcoid is characterized by noncaseating granulomas, and all cases that we accepted were biopsy- or autopsy-proven, although biopsy material was often not from the CNS. Other tests can be useful in establishing the diagnosis, but biopsy still provides the strongest evidence for this disease. Elevated ACE levels are not specific for sarcoid but are elevated in the serum of 70% to 80% of patients with systemic disease and in the CSF of ~50% of patients with neurosarcoid. Analysis of CSF may show a mild lymphocytosis (100 cells per μL), mildly elevated protein (to 70 mg%), mildly decreased glucose, and increased immunoglobulins with oligoclonal banding. When available, the Kveim test can be positive in 85% of patients. MRI has become increasingly useful. Findings in neurosarcoid include periventricular high-signal lesions on T2-weighted images, leptomeningeal enhancement, and solitary masses. Many of the reported pediatric cases antedate the availability of MRI, and whether there are different patterns in children than in adults is unclear.

The most appropriate treatment for pediatric neurosarcoid is uncertain. Corticosteroids are widely used and are often effective. For adult patients for whom corticosteroid therapy fails, azathioprine or cyclosporin A have been prescribed.

An important caveat to our analysis is that published case reports may not be representative of the population of patients with a disorder. Furthermore, many of these cases were published by nonneurologists whose interests were in pulmonary, rheumatic, or other manifestations of the disease. The neurologic findings were presented for completeness. Although we omitted those cases where we could not confidently extract a detailed picture of the patients’ findings on presentation, it is always possible that if experienced neurologists rather than nonneurologists had reported these patients, they would have conveyed different findings.

CONCLUSIONS

An analysis of 29 pediatric neurosarcoid cases indicates that compared with adults, children are more likely to have seizures, less likely to have cranial nerve palsies, and perhaps more likely to have a space-occupying lesion. Analyzing these children by age group, preadolescents and adolescents, suggests that pediatric cases evolve to adult patterns of disease, with more frequent cranial neuropathies and less frequent seizures as the patients progress to adulthood.

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