Recurrent Meningitis Associated With Complete Curarino Triad in an Adult
—Case Report—

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Abstract

A 58-year-old woman presented with Curarino triad manifesting as recurrent meningitis. Curarino triad is a combination of a presacral mass, a congenital sacral bony abnormality, and an anorectal malformation, which is caused by dorsal-ventral patterning defects during embryonic development. She had a history of treatment for anal stenosis in her childhood. Radiographic examinations demonstrated the characteristic findings of Curarino triad and a complicated mass lesion. The diagnosis was recurrent meningitis related to the anterior sacral meningocele. Neck ligation of the meningocele was performed via a posterior transsacral approach after treatment with antibiotics. At surgery, an epidermoid cyst was observed inside the meningocele. The cyst content was aspirated. She suffered no further episodes of meningitis. The meningitis was probably part of the clinical course of Curarino triad. Radiography of the sacrum and magnetic resonance imaging are recommended for patients with meningitis of unknown origin. The early diagnosis and treatment of this condition are important.

Key words: adult case, Curarino triad, epidermoid cyst, recurrent meningitis

Introduction

The triad of a presacral mass, a sacral bony abnormality, and an anorectal malformation was first described in 1926.12) This triad was recognized as a syndrome, and embryogenesis postulated as the cause in 1981.4) The presacral mass is most frequently a teratoma (18% of cases) or an anterior sacral meningocele (69%), whereas neurenteric cysts or dermoid (3%) and epidermoid cysts (1%) are unusual.10,11,14–17,19,20) The mass is associated with a hemisacral or scimitar sacral defect, but segmentation anomalies are less common. The anorectal malformation is usually either anal stenosis or atresia.

Currarino triad is caused by malformation of the caudal notocord which leads to aberrant secondary neurulation and an incomplete separation of endodermal and ectodermal layer in the developing embryo.3–5,8,16,18) The triad may also occur in the majority of patients as an autosomal dominant trait and associated with mutations in the homeobox gene $HLXB9$.1,9,13)

Currarino triad has been described in over 200 cases since 1926, of which at least 50% were familial. The primary pediatric symptom is early infant constipation, which occurs in 80.4% of cases before 12 years of age, and is identified in 91% of cases before 30 years of age.16) We treated a 58-year-old woman with Curarino triad manifesting as recurrent meningitis, and describe the clinical characteristics of this adult case.

Case Report

A 58-year-old woman had suffered recurrent back pain and feverishness for 10 years. She was referred to our hospital for increasingly frequent episodes of these symptoms. Her past medical history was notable for anorectal malformation (details unknown), and she had previously undergone a two-stage operation at 6 years of age.

On admission, physical and neurological examinations found no abnormalities except for mild vesicorectal dysfunction. She became feverish again after admission to our hospital. The diagnosis was...
bacterial meningitis based on the remarkable number of polymorphonuclear leukocytes in the cerebrospinal fluid (CSF). However, CSF culture identified no organism.

Radiography demonstrated a bony deformity of the sacrococcygeal region in the shape of a scimitar (Fig. 1). Myelography and postmyelography computed tomography demonstrated an anterior sacral meningocele which communicated with the thecal sac (Fig. 2). Magnetic resonance (MR) imaging demonstrated the presacral cyst containing CSF and a cystic tumor (Fig. 3). However, the barium enema test did not show a fistula between the meningocele and rectum.

Surgery was performed through the posterior transsacral approach after treatment with antibiotics. A sacral laminectomy was carried out via a midline lumbosacral incision. The dura mater was opened and a small opening was found in the anterior aspect of the dural sac. The cystic tumor mass was observed inside the meningocele, and pus was found inside this tumor. Culture of the pus identified coryneform bacteria. The presence of pus inside the tumor suggested that the recurrent meningitis might be related to the anterior sacral meningocele and the epidermoid cyst. The epidermoid cyst was tightly adhered to surrounding tissues and poorly circumscribed, so removal was not possible. After aspirating the pus, the operative field was cleaned and disinfected using antibiotic solution. Then, the fluid of the anterior sacral meningocele was aspirated and neck ligation was performed.

The histological diagnosis was epidermoid cyst, based on the cyst wall consisting of keratinizing, stratified squamous epithelia, and infiltration of inflammatory cells inside the cyst (Fig. 4). MR imaging performed 1 month after surgery demonstrated the reduced residual cyst. The patient
Fig. 4 Photomicrograph showing that the cyst wall consists of keratinizing, stratified squamous epithelium accompanied by infiltration of inflammatory cells. Hematoxylin and eosin stain, original magnification $\times 100$.

Table 1 Clinical characteristics of pediatric and adult cases

<table>
<thead>
<tr>
<th></th>
<th>Pediatric cases</th>
<th>Adult cases</th>
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</thead>
<tbody>
<tr>
<td>Female-to-male ratio</td>
<td>2:1</td>
<td>6:1</td>
</tr>
<tr>
<td>Related symptoms</td>
<td></td>
<td></td>
</tr>
<tr>
<td>constipation</td>
<td>67%</td>
<td>37%</td>
</tr>
<tr>
<td>urination difficulty</td>
<td>9%</td>
<td>20%</td>
</tr>
<tr>
<td>pain</td>
<td>1%</td>
<td>36%</td>
</tr>
<tr>
<td>dysmenorrhea</td>
<td>0%</td>
<td>7%</td>
</tr>
<tr>
<td>Associated abnormalities</td>
<td></td>
<td></td>
</tr>
<tr>
<td>teratoma</td>
<td>24%</td>
<td>2%</td>
</tr>
<tr>
<td>tethered cord</td>
<td>17%</td>
<td>0%</td>
</tr>
<tr>
<td>fistula</td>
<td>13%</td>
<td>3%</td>
</tr>
<tr>
<td>dermoid cyst</td>
<td>5%</td>
<td>4%</td>
</tr>
<tr>
<td>genital anomaly</td>
<td>5%</td>
<td>13%</td>
</tr>
<tr>
<td>epidermoid cyst</td>
<td>1%</td>
<td>4%</td>
</tr>
</tbody>
</table>

From refs. 11, 14–17, 19, and 20).

made a satisfactory recovery, and she has remained free from meningitis postoperatively.

Discussion

The female-to-male ratio in adult cases is 6:1 in contrast to the ratio in pediatric cases of 2:1 (Table 1).11,14–17,19,20 The most frequent symptom is constipation (67%) followed by urination difficulty (9%) in pediatric cases, but various symptoms seen in adult cases. The frequency of constipation associated with anorectal malformation is lower in adult cases. In our case, the presenting symptoms were back pain and feverishness. The difference in symptoms between the two groups suggests that anorectal stenosis in pediatric cases may be more severe than in adult cases. In addition, teratoma, tethered cord, and fistula are very rare in adult cases, whereas genital anomaly is more frequent in adult cases. However, the radiographical findings are not remarkably different between the two groups.

Meningitis is one of the most severe complications in Currarino triad, and occurred in 37 cases (including our case), or about 13% of all cases.2,4,6,7,14,19,20 The mortality due to meningitis in Currarino triad surprisingly high at 56%. The etiology was iatrogenic (65%), anal or rectal fistula communicating with the spinal canal (16%), or idiopathic (16%). Previously, aspiration of the meningocele was performed via the rectum.20 Therefore, many patients contracted meningitis and died. The cause of meningitis is now thought to be the fistula communicating between the spinal canal and the rectum or skin.4,7 However, we should also note that some cases are idiopathic meningitis and include three cases of epidermoid cysts containing pus.10,11,19 The mechanism of infection remains uncertain. In our case, no neurenteric or skin fistula was detected. However, a tiny rupture in the meningocele or epidermoid cyst may have occurred in the past. As a result, the patient contracted meningitis.

Currarino triad is becoming more frequently recognized. Recurrent meningitis may occur if this syndrome is left untreated. Since meningitis associated with this syndrome causes severe complications and high mortality, we recommend that early treatment should be undertaken as soon as an anterior sacral meningocele is found. Moreover, radiography of the sacrum and MR imaging are recommended for patients with recurrent meningitis of unknown origin. In our case, further episodes of meningitis were prevented by neck ligation of the meningocele.

References


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