Original Article

Reflex sympathetic dystrophy in pregnancy: nine cases and a review of the literature


*Maternity Ward, Poesy Hospital Centre, Poesy, France
*Rheumatology and Physical Medicine and Rehabilitation Service, Victor Dupuy Hospital Center, Argenteuil, France

Received 26 November 1998, accepted 10 February 1999

Abstract

Objective: To better understand the diagnosis of reflex sympathetic dystrophy of the lower extremities in pregnant women. Subject: Disease analysis using a retrospective series of nine cases and a review of the literature (57 patients and 159 sites of reflex sympathetic dystrophy). Results: This disorder should be considered in any painful pelvic girdle syndrome or lower extremity pain. The hip is involved in 88% of cases. Symptoms develop in the third trimester of pregnancy, between the 28th and the 34th weeks. Magnetic resonance imaging (MRI) provides an early, accurate, and very specific diagnosis, although standard radiography continues to be the first-line diagnostic tool. Fracture occurs in 19% of patients. The etiology and pathophysiology remain unclear, although pregnancy itself appears to play a significant role in this disease. Although locoregional mechanical factors partly explain reflex sympathetic dystrophy, hypertriglyceridemia appears to be a risk factor. This disorder develops independently, but the conclusion of pregnancy appears to be necessary for cure. Reflex sympathetic dystrophy does not appear to affect the course of the pregnancy. Indications for cesarean delivery remain obstetrical and should be discussed when a fracture is involved. Simple therapeutic management using gentle physical therapy provides rapid and complete recovery in 2–3 months. Conclusion: Reflex sympathetic dystrophy during pregnancy remains poorly understood and underestimated. Only joints of the upper limbs are involved. MRI appears to be the best diagnostic tool. Pathogenesis remains unclear. Fractures are not rare. Treatment should be non-aggressive. © 1999 Elsevier Science Ireland Ltd. All rights reserved.

Keywords: Pregnancy; Reflex sympathetic dystrophy; Rheumatology; Sudeck’s syndrome

1. Introduction

Pelvic or lower extremity pain during pregnancy poses certain diagnostic difficulties. Although pregnancy can alter the course of some inflammatory disorders, it can also lead to the emergence of mechanical joint diseases, such as reflex sympathetic dystrophy of the lower extremities. Reflex sympathetic dystrophy (RSD) is a type of arthropathy combining a painful syndrome with locoregional trophic disturbances. This poorly understood disorder, which is frequently deceptive and sometimes clinically misleading in the pregnant woman, was first described in 1959 by Curtiss and Kincaid [1]. They first described three cases of transient demineralization of the hip during pregnancy. Lequesne [2], in 1968, described the same pathology and called it transient osteoporosis or neurotrophic rheumatism. Later, other terms were used: migratory osteolysis, reflex sympathetic dystrophy syndrome, algoneurodystrophy, and Sudeck’s atrophy or syndrome [3–5]. Currently these very similar clinical disorders are grouped as RSD in the American English medical literature. So far only case reports have been published in the literature. Therefore we were prompted to analyse a
series of nine cases of RSD: (a) the characteristics of the patients, (b) the relation between the evolution of the pregnancy and the occurrence of RSD and (c) the outcome of the pregnancy.

2. Materials and methods

The clinical cases were compiled retrospectively over a period of 3 years, from May 1994 to May 1997, from four French services: the Rheumatology Service at the Argetne Hospital Center, and the maternity wards at Pontenais Regional Hospital Center, Lariboisier University Hospital Center in Paris, and Poissy Hospital Center. Only cases with certain diagnosis (based on clinical examination, paraclinical diagnostic tools and evolution after treatment and delivery) were involved in this series. During this time, about 8000 deliveries were recorded in the three maternity wards.

3. Results

Nine cases of RSD occurring during pregnancy were found (Table 1). The average age of the patients was 36±3.5 years (range, 30–43). Only two patients were primiparas. The average interval from conception to the appearance of symptoms was 28.1±2.2 weeks (range, 25–32). There were eight singleton pregnancies and one spontaneous triplet pregnancy. The only abnormal sign observed before the onset of the RSD was excessive weight gain, without gestational diabetes, for six of the patients (weight gain superior to 12 kg). In two other cases the total weight gain was only 3 kg, but there was significant initial obesity. In one case the weight gain was not recorded.

Any of the joints of the lower extremity may be involved. The hip was most frequently involved: in seven patients and in 11 of 21 involved joints (52%). Bilateral involvement was observed in four cases. There was no apparent predominant side: seven cases occurred on the right and six on the left. A joint group may become involved, at the onset or later on. Thus, in one case, the involvement of one hip even occurred postpartum.

The diagnosis of RSD was made at the onset; in only one patient. In six cases the initial diagnosis was sciatic-type pain, and in two cases, phlebitis pain.

Clinically, the mechanical symptomatology appeared gradually, and the clinical signs were nonspecific. Functional impairment, sometimes total, was commonly found. Limping was observed in eight cases. Pain was always present, often poorly localized, and of variable severity. In only two cases did we find symptomatology characteristic of an inflammatory phase. In these cases we noted a 'clonisomie' syndrome (that is, defined by weakness of the lower limb in the supine position contrasting with none or only slight weakness upon standing, a normal neurological examination and full passive mobility of the hip). When a reduction in joint mobility was present, it was mild, in contrast with the apparent severity of the impairment.

The diagnosis was made using standard radiography in eight cases, sometimes after several weeks' delay. In most cases the X-rays were initially normal but subsequently revealed areas of patchy bone demineralization in the involved joint. In the hip, the femoral head sometimes appeared 'ghostly'. The joint space remained intact in all cases (Fig. 1). The radiological findings did not always correspond to the clinical signs. Magnetic resonance imaging (MRI) was performed in four cases and revealed a characteristic image each time, thus providing an early diagnosis. There was low signal intensity on T1-weighted images and high signal intensity on T2-weighted images in the affected joints (Figs. 2 and 3).

Treatment was symptomatic until delivery, combining non-weight-bearing with rest and non-narcotic analgesics. Physical therapy was gradually undertaken and was carried out only to the point of pain. After delivery, six patients began a calcitonin-based treatment (Calsyn® synthesis Calcitonin SPECIA® Laboratory Paris France 100 IU/day for 8 days, then 100 IU/1 3 times a week for 3 weeks). Independent of the treatment undertaken, clinical recovery was rapidly obtained, with an average time to recovery of

Table 1
Details of the nine cases of RSD

<table>
<thead>
<tr>
<th>Age</th>
<th>P</th>
<th>Date</th>
<th>Localisation</th>
<th>Side</th>
<th>Recovery</th>
<th>Delivery term</th>
<th>Weight (PERC)</th>
</tr>
</thead>
<tbody>
<tr>
<td>35</td>
<td>4</td>
<td>26</td>
<td>Hip + knee</td>
<td>R</td>
<td>5</td>
<td>V, 38,5 WA</td>
<td>3450 (80)</td>
</tr>
<tr>
<td>39</td>
<td>1</td>
<td>30</td>
<td>Hip + knee + ankle</td>
<td>R</td>
<td>7</td>
<td>CCEASAR, 38 WA</td>
<td>3960 (95)</td>
</tr>
<tr>
<td>43</td>
<td>4</td>
<td>26 1/2</td>
<td>Hip + knee</td>
<td>R + L/R</td>
<td>*</td>
<td>V, 59 WA</td>
<td>3600 (85)</td>
</tr>
<tr>
<td>34</td>
<td>1</td>
<td>29</td>
<td>Hip + knee</td>
<td>R + L/L</td>
<td>*</td>
<td>V, 35 WA</td>
<td>3400 (90)</td>
</tr>
<tr>
<td>36</td>
<td>2</td>
<td>28</td>
<td>Inferior limb</td>
<td>L</td>
<td>24</td>
<td>V, 37 WA</td>
<td>2550 (25)</td>
</tr>
<tr>
<td>38</td>
<td>8</td>
<td>26/PP</td>
<td>Ankle + hip</td>
<td>L/R</td>
<td>3</td>
<td>V, 40 WA</td>
<td>4020 (50)</td>
</tr>
<tr>
<td>35</td>
<td>6</td>
<td>32</td>
<td>Hip + knee</td>
<td>R</td>
<td>4</td>
<td>CCEASAR, 34,5 WA</td>
<td>2340 (50)</td>
</tr>
<tr>
<td>35</td>
<td>4</td>
<td>25</td>
<td>Ankle</td>
<td>L</td>
<td>6</td>
<td>V, 38 WA</td>
<td>3620 (85)</td>
</tr>
<tr>
<td>38</td>
<td>5</td>
<td>30</td>
<td>Hip</td>
<td>R + L</td>
<td>10</td>
<td>V, 38 WA</td>
<td>3620 (85)</td>
</tr>
</tbody>
</table>

*P, parity; date, date of sign apparition in weeks of amenorrhoea; R, right; L, left; recovery, in weeks; PP, post-partum; *, unknown; V, vaginal delivery; CCEASAR, cesarean section; WA, weeks of amenorrhoea; weight, in grams.
Fig. 1. Standard radiograph: homogeneous demineralization of the right femoral head, with joint space intact.

Fig. 2. MRI of the hips, T1-weighted images (coronal sections): left hip, fuzzy contour of homogeneous low signal intensity in the femoral neck; right hip, discrete area of low signal intensity in the inferior femoral head.
LETTERS TO THE EDITOR

Reflex Sympathetic Syndrome and Peripheral Dystonia

I have read with interest about peripheral trauma inducing dystonia, in a study of Movement Disorders (January 2001). Some aspects of these reports surprised me. In 1999, related to the return of subclinical movement disorders, reported in the treatment of Parkinson’s disease, a letter written by Battle stated, “...I notice that the references for both papers were similar yet concluded with opposite views...” The same applies to this controversy, where agreements in the references (quote numbers 1, 6, 7, 8, 13, 16, 22, 45, 46, 52, and 57) by Jankovic, which correspond to quote numbers 2, 96, 42, 45, 41, 56, 29, 27, 48, 50, 54, and 92 by Weinberg, respectively.

I would like to comment on the suggestion that dystonia, in the context of the reflex sympathetic dystrophy (RSD), does exist (it is now called complex regional pain syndrome [CRPS]). To begin with, it may not be appropriate to assume without doubt the existence of this condition in the patient, as RSD and causalgia, respectively. Ambiguous, because it has been said that “...patients diagnosed with RSD are not a homogeneous population, they may have any number of possible disorders generating what we see as the actual pain...”

One article that frequently mentions dystonia and RSD is the work of Schwartzman and colleagues (ref. 18). Reading this work in detail, we found some strange elements; for example, this work begins “Reflex sympathetic dystrophy (RSD) is a condition of severe burning pain, swelling, and vasomotor, sudomotor, dystrophic, and atrophic changes in the affected body parts” (ref. 12). The accompanying clinical description of dystonia, weakness, tremor, and involuntary movements, and spasms have been described (ref. 3–6). Finally, Schwartzman and colleagues criticized the work of Schonacker and coworkers, which stated: “We studied four patients with dystonia, action-induced involuntary postures of the hand could be considered focal dystonia. All four patients had electrophysiological findings consistent with peripheral axon loss lesions (proximal nerve syndrome, radial nerve palsy, lower back pain, plexus lesion and neural nerve lesion) but inadequate description of peripheral nerves forms a small but important group of causes for secondary dystonia...”

Schwartzman and colleagues pointed out, “...The most prevalent motor complaints, common to some degree in all of these patients, were weakness, spasm, and the inability or difficulty in initiating movements of the affected extremity. The most frequent description of the abnormalities was, ‘My mind tells me to move, but it won’t’...” which is in fact quite similar to features that characterize cases of psychogenic dystonia as mentioned previously by Gálvez-Jiménez and Lang. If we critically review the body of evidence, part of the evidence supports the favorable position Jankovic defends and some elements that at least generate a reasonable doubt. This in, however, some incongruence between material reported in some articles and the interpretation provided in the controversy. Jankovic mentioned the work of Turcios and Ochoa in support of his viewpoint, but these authors stated, “...Surprisingly, no cases of CRPS I but only cases of CRPS type II displayed abnormal movements. In addition to an absence of evidence of structural nerve, spinal cord, or intracranial damage, all CRPS I patients with abnormal movements typically exhibited gastrointestinal (anorectal) and skin. In some cases, malingering was documented by slow growth. This study highlights abnormal movements in CRPS I as constituting a significant clinical challenge that differentiates CRPS I from CRPS II. They are consistently of somatoform or malingered origin, signaling an underlying psychoneurological disorder responsible for the entire CRPS profile.” In the same fashion, Jankovic suggests Bhacca and colleagues (ref. 11 by Jankovic). In this work, the authors presented 6 women and 2 men who developed causalgia and dyskinesia. The authors concluded, however, “...The syndrome was typically sustained producing a fixed dystonic posture, in contrast to the mobile spasms characteristic of idiopathic rigidity...”

Further, Jankovic ends his article, “...A relief of the movement disorders with psychotherapy, powerful suggestion, placebo, or physical therapy strongly argues against a neurological etiology because complete and permanent remissions are rare in most organic disorders” and quotes (ref. 67) the work of Crumlish and associates, however, the work cited did not attack the point made by Jankovic. That study had the objective to “...investigate psychiatric and neurological morbidity, diagnostic stability, and indicators of psychopathology in patients previously identified as having medically unexplained motor symptoms”, and concluded, “...Unlike Sander’s study of 1965, a low incidence of physical or psychiatric diagnoses which explained these patients’ symptoms or disability was found. However, a high level of psychiatric comorbidity existed.”

I conclude by mentioning the article of van Hulst and coworkers (ref. 20) regarding treatment with intracranial baclofen in patients with dystonia in the context of RSD. These authors obtained a spectacular response to this treatment (6 women received baclofen in a median dose of 489 µg daily; 3 had a complete resolution of their movement disorder, and the other 3 improved substantially). These results contrast with more modest ones obtained by other groups of patients with primary and secondary dystonia. The striking and unusual response of the RSD patients needs to be interpreted with caution. As emphasized by Jankovic and Pahn, “...Relief of dystonia with psychotherapy, powerful suggestion, placebo, or physical therapy virtually excludes a neurological...”
LETTERS TO THE EDITOR

1213

Reply: Reflex Sympathetic Syndrome and Peripheral Dystonia

I appreciate the interest of Dr. Serrano-Dueñas in my article published in 2001 in the Controversy section of Movement Disorders. Although the article mentioned reflex sympathetic dystrophy syndrome (RSD; or complex regional pain syndrome, CRPS), this was not the primary focus, as it seems to be in Dr. Serrano-Dueñas' letter. Dr. Serrano-Dueñas quotes extensively from the article by Schwartzman and colleagues.1 Serrano-Dueñas and associates,2 Verdugo and Ochoa,3 and Bhattacharya and coworkers,4 but it is not clear whether he agrees or disagrees with the statements in those articles. I am not even sure whether he agrees with published reports5–7 on the response to intrathecal baclofen, also observed in some of our patients with peripherally induced dystonia.8 Judging from his last sentence quoting from the chapter by Jankovic and Fahn,9 I assume that he considers patients with RSD/CRPS to be psychogenic. Although this notion may be true for some patients with this poorly defined syndrome, I do not believe that it applies uniformly to all patients with the features of RSD/CRPS.

Joseph Jankovic, MD

Parkinson's Disease Center and Movement Disorders Clinic
Bayor College of Medicine
Houston, Texas

References


DOI: 10.1002/mds.12082
Published online 12 August 2003 in Wiley InterScience (www.interscience.wiley.com).

Movement Disorders, Vol. 16, No. 10, 2003