2015 Persian Gulf Criteria for Early Diagnosis of Relapsing Polychondritis

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Abstract
Relapsing polychondritis (RPC) is a systemic autoimmune disease with unknown etiology but genetic background, involving chondral structures and tissues consisting collagens, especially types II, IX and XI. This disorder is presenting as auricular chondritis (bilateral or unilateral), nasal chondritis, main airway chondritis, oligo/polyarthritis, eye inflammation and cochlear/vestibular dysfunction along with many other organs involvement, including heart, kidney, skin, gastrointestinal tract and etc. Nowadays, McAdam’s criteria are used for the diagnosis of RPC. The author wants to show that it may have its own defects for early diagnosis of RPC and in some cases it doesn’t have accuracy for diagnosis. So the author decided to introduce 2015 Persian Gulf Criteria for early/accurate diagnosis of RPC.

Keywords
Relapsing Polychondritis, McAdam’s Criteria, Persian Gulf Criteria

1. Introduction
Relapsing polychondritis (RPC) is a systemic autoimmune disease with unknown etiology but genetic background, involving chondral structures and tissues consisting collagens, especially types II, IX and XI. This disorder is presenting as auricular chondritis (bilateral or unilateral), nasal chondritis, main airway chondritis, oligo/polyarthritis, eye inflammation and cochlear/vestibular dysfunction along with many other organs involvement, including heart, kidney, skin, gastrointestinal tract and etc (1,2).

2. Main Body
There is no specific test for the diagnosis of this disease. The diagnosis of RPC is made by clinical/laboratory judgment of an expert rheumatologist. We know that McAdam’s Criteria (3) has been used for the diagnosis of RPC for many years. McAdam’s Criteria includes six items:

- Large airway chondritis (Laryng/o tracheo/bronchomalacia)
- Cochlear/Vestibular dysfunction (Tinnitus, Vertigo and Sensory-neural hearing loss)

With at least three out of six above items, the diagnosis of RPC will be established. Many years of clinical practice in Rheumatology, has given the author (Iraj Salehi-Abari) the opportunities to visit some cases that with clinical/laboratory judgment were RPC, but by using McAdam’s criteria, they could not be classified as that.

For example, a 50-year old woman with unilateral auricular chondritis, nasal chondritis and polyarthritis was not a case of RPC according to McAdam’s criteria whereas after enough evaluation and rulling out the other diagnosis, she was a confirmed case of RPC.

In another case of RPC who was a 48-year old man with two episodes of bilateral auricular chondritis and saddle nose deformity along with normal nasal mucosa, McAdam’s criteria cannot confirm the diagnosis of RPC.

In opposite, some cases have been visited that with clinical/ laboratory judgment were not RPC but by using McAdam’s criteria, the diagnosis of RPC could be established for them. For example, a 30-year old man with Uveitis, Oligoarthritis of both knees and sensory-neural hearing loss that fulfilled three out of six criteria of...
McAdam’s for RPC, but due to the past history of recurrent oral aphthosis and positive pathergy test, he was a definite case of Behcet’s disease.

Another case was a 42-year old woman with saddle nose deformity, tinnitus, vertigo, sensory-neural hearing loss and polyarthritis of knees and ankles that fulfilled three out of six criteria of McAdam’s for RPC but due to the history of bloody-purulent nasal discharge, nasal ulcer and bilateral otitis media, she was a definite case of granulomatosis with polyangiitis (Wegener’s).

All cases of RPC that have been visited by author in these many years of practice, had at least one of the below items:
- Auricular chondritis
- Nasal chondritis
- Main airway chondritis
- Bilateral auricular chondritis, confirmed the diagnosis of RPC by the author- Iraj Salehi Abari- that is presented here in polyangiitis (Wegener’s).

3. Conclusion

All cases of RPC that have been studied in the literature by author, had at least one of above items too. The author has had cases of RPC in clinical practice that by recurrent pattern of chondritis in them, the diagnosis of RPC had been established upon clinical judgment many weeks before fulfilling McAdam’s criteria. The author has had three cases of RPC that by doing biopsy and pathologic evaluation in them, the diagnosis of RPC had been established a few months before fulfilling McAdam’s criteria. Also the author has seen one case of RPC that by trial therapy with 1 mg/kg/day of Prednisolone in her, dramatic response of bilateral auricular chondritis, confirmed the diagnosis of RPC by clinical judgment when it didn’t fulfill McAdam’s criteria.

Anyway the author thinks that the McAdam’s criteria is a good instrument for detecting RPC in many advanced/established cases of RPC but it is not suitable for early diagnosis of RPC in the initial presentation of the disease. After 16 years of practice in Rheumatology and seeing some cases of RPC along with spending a lot of time looking for and studying the items of RPC in literature, a new diagnostic criteria for early diagnosis of RPC has been made by the author- Iraj Salehi Abari- that is presented here in table A:

By this letter, the author wants to introduce 2015 Persian Gulf Criteria for early diagnosis of RPC to the world of Rheumatology. The cases of RPC that have been seen by the author, are not enough in number to deliver an original article and he thinks it is better for this new diagnostic criteria of RPC to be checked by other Rheumatologists in their confirmed groups of RPC cases by clinical/laboratory judgment and report their results.

Finally, by this way, Iraj Salehi-Abari as the creator of Iran Criteria for early diagnosis of Rheumatoid arthritis, Ankylosing Spondylitis, Granulomatosis with polyangiitis, 2015 ACR/SLICC revised criteria for diagnosis of Systemic lupus erythematosus and Chondromalacia Patella (4,5,6,7,8), would like to ask the ACR (American College of Rheumatology) and/or EULAR (European League Against Rheumatism) members and all of other Rheumatologists in the world to evaluate this new criteria for the diagnosis of RPC and McAdam’s criteria separately in the initial presentation of cases of RPC diagnosed by clinical/laboratory judgment.

By this way, the world of Rheumatology can answer this question whether or not this new criteria is better than McAdam’s criteria for diagnosis of RPC? I think it is better, what’s your opinion?

Table A. 2015 Persian Gulf criteria for early diagnosis of Relapsing Polychondritis

<table>
<thead>
<tr>
<th>Domain I</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Auricular chondritis</td>
<td></td>
</tr>
<tr>
<td>Unilateral</td>
<td>1.p</td>
</tr>
<tr>
<td>Bilateral</td>
<td>2.p</td>
</tr>
<tr>
<td>Nasal chondritis</td>
<td>1.p</td>
</tr>
<tr>
<td>Main airway chondritis</td>
<td>1.p</td>
</tr>
<tr>
<td>Domain II</td>
<td></td>
</tr>
<tr>
<td>Oligo/Polyarthritis</td>
<td>1.p</td>
</tr>
<tr>
<td>Eye inflammation</td>
<td>1.p</td>
</tr>
<tr>
<td>Cochlear/Vestibular dysfunction</td>
<td>1.p</td>
</tr>
<tr>
<td>Relapsing feature</td>
<td>1.p</td>
</tr>
<tr>
<td>Dramatic improvement with corticosteroid</td>
<td>1.p</td>
</tr>
<tr>
<td>Pathologic findings</td>
<td>Up to 2.p</td>
</tr>
<tr>
<td>- Lymphocytic infiltration</td>
<td>1.p</td>
</tr>
<tr>
<td>- IgG and Complement deposition</td>
<td>1.p</td>
</tr>
</tbody>
</table>

a. Any items cannot be explained by other etiologies better than Relapsing Polychondritis. If there are at least three points out of eleven, including at least one point from domain I, the diagnosis of RPC will be established.

b. It is not along with regional lymphadenopathies and it spares ear lobe.

c. It presents as severe nasal pain/tenderness and swelling in initial presentation and with persistency or recurrency as saddle nose deformity.

d. It presents as hoarseness, stridor, dyspnea on exertion and wheezing/rhonchi in clinical picture and tracheal/bronchial narrowing in imaging including plain CXR or HRCT scan of lung during inspiration and expiration (9,10)

e. It is non-erosive and seronegative (No positive RF, No Positive anti-CCP).

f. Conjunctivitis and/or Epi/Scleritis and/or Keratitis and/or Uveitis.(11)

g. Tinnitus and/or vertigo and/or sensory-neural hearing loss(12)

h. There are at least two episodes of the attack of chondritis.

i. Significant improvement of chondritis and other items with 1 mg/kg/day of prednisolone within one week.

j. There is lymphocytic infiltration as dominant cells along with other types of WBC in light microscopy and/or IgG plus Complement deposition in Immunofluorescent microscopy of dermal-chondral or perichondral tissue.
References


