

## Reviewer Assessment

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# Localized eosinophilic fasciitis (Shulman's disease) as a differential diagnosis of nerve compression syndrome

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## Reviewers' Comments to Original Submission

### Reviewer 1: Holger Bannasch

Jan 09, 2017

#### Reviewer Recommendation Term:

Accept with Minor Revision

#### Overall Reviewer Manuscript Rating:

65

#### Custom Review Questions

Custom Review Questions	Response
Is the subject area appropriate for you?	4
Does the title clearly reflect the paper's content?	4
Does the abstract clearly reflect the paper's content?	5 - High/Yes
Do the keywords clearly reflect the paper's content?	4
Does the introduction present the problem clearly?	4
Are the results/conclusions justified?	4
How comprehensive and up-to-date is the subject matter presented?	3
How adequate is the data presentation?	4
Are units and terminology used correctly?	5 - High/Yes
Is the number of cases adequate?	4
Are the experimental methods/clinical studies adequate?	4
Is the length appropriate in relation to the content?	4
Does the reader get new insights from the article?	4
Please rate the practical significance.	2
Please rate the accuracy of methods.	3
Please rate the statistical evaluation and quality control.	N/A
Please rate the appropriateness of the figures and tables.	3
Please rate the appropriateness of the references.	4
Please evaluate the writing style and use of language.	5 - High/Yes
Please judge the overall scientific quality of the manuscript.	3
Are you willing to review the revision of this manuscript?	Yes

**Comments to Authors:**

This is an interesting case report on a rare disease as a differential diagnosis of a nerve compression syndrome.  
 The article is written comprehensive and concise.  
 The quality of figures 2A and 2B is low, therefore one of them can be omitted.

I have a question regarding the MRI interpretation: Can the radiologist retrospectively comment on the totally different diagnosis?

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**Reviewer 2: Thomas Kremer**

Jan 16, 2017

**Reviewer Recommendation Term:**  
**Overall Reviewer Manuscript Rating:**

Accept with Minor Revision  
 N/A

**Custom Review Questions****Response**

Is the subject area appropriate for you?	5 - High/Yes
Does the title clearly reflect the paper's content?	5 - High/Yes
Does the abstract clearly reflect the paper's content?	5 - High/Yes
Do the keywords clearly reflect the paper's content?	5 - High/Yes
Does the introduction present the problem clearly?	4
Are the results/conclusions justified?	5 - High/Yes
How comprehensive and up-to-date is the subject matter presented?	4
How adequate is the data presentation?	N/A
Are units and terminology used correctly?	N/A
Is the number of cases adequate?	N/A
Are the experimental methods/clinical studies adequate?	N/A
Is the length appropriate in relation to the content?	4
Does the reader get new insights from the article?	4
Please rate the practical significance.	3
Please rate the accuracy of methods.	N/A
Please rate the statistical evaluation and quality control.	N/A
Please rate the appropriateness of the figures and tables.	4
Please rate the appropriateness of the references.	4
Please evaluate the writing style and use of language.	2
Please judge the overall scientific quality of the manuscript.	4
Are you willing to review the revision of this manuscript?	Yes

**Comments to Authors:**

The authors describe an interesting case of a rare disease and provide a very good overview on differential diagnoses. Overall the manuscript merits publication in ISS. However, a few issues should be addressed.

General: Overall, the manuscript is written well. However, some of the paragraphs should be edited by a native English Speaker to improve the English-

Abstract: As I understand, the authors did not perform any surgical procedure in Addition to the Biopsy and the patients# symptoms resolved spontaneously over time. This should be made more clear.

Case Report: The preoperative Evaluation is well presented in paragraphs 1 and 2. Likewise, the surgical procedure (biopsy) is well presented. However, I do not really understand why the authors did not perform a „Tumor“ debulking in a second procedure. Why did the authors not open the cubital tunnel, secondarily? The authors should describe the different Treatment Option in more Detail and justify their choice. „Further surgical neurolysis or tumor debulking would not have been of a benefit either,...“ is not enough. What else did the authors do (e.g. antiinflammatory drugs [NSAIDs], physical therapy ....). Overall, the authors provide good Information on differential diagnoses but should also provide more Information on different Treatment Options.

Conclusion: I agree with the authors' conclusion.

Literature: Only a few citations are provided by the authors. However, Literature selection and relevance is appropriate and related to the rarity of the disease.

Figures: The figures and legends are appropriate. However, the Definition of figures 2A and B should be improved.

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# Authors' Response to Reviewer Comments

Jan 23, 2017

## REVIEWER #1:

Thank you very much for appreciating the value of our case report elucidating the appearance of a rare disease and its differential diagnosis.

1. "The quality of figures 2A and 2B is low, therefore one of them can be omitted."

We thank the reviewer for his comment. In the revised version of the manuscript the quality of the given figure is increased and reduced to only on image.

2. "Can the radiologist retrospectively comment on the totally different diagnosis?"

We have discussed this case with different radiologists which have all be uncertain and were unable to allow a clear unambiguous diagnosis by first instance. However, they rated the case most likely as a tumorous process. After the description of the diagnosis with the current knowledge the radiologists did not changed their initial statement, however, weakened it that this kind of process might also be of inflammatory origin. None of the contacted radiologists had ever seen such a rare disease at this particular part of the body.

## REVIEWER #2:

The authors very much appreciate that the manuscript transported the interest in this rare case and its differential diagnosis. That is the exact intention of this case report.

1. "...some of the paragraphs should be edited by a native English Speaker to improve the English "

The revised version of the manuscript was proof-read by a native speaker to address this issue in general. Please see all corrections given by orange font and underlining throughout the manuscript.

2. "Abstract: As I understand, the authors did not perform any surgical procedure in Addition to the Biopsy and the patients# symptoms resolved spontaneously over time. This should be made more clear."

The reviewer is completely right. There was no additional surgical procedure performed other than the biopsy. We changed this by adding a new paragraph which reads now as follows:

Consequentially, the presented patient would not have benefitted from further surgical neuolysis or tumor debulking since the eosinophilic fasciitis is an inflammatory and systemic disease. The patient's symptoms decreased spontaneously, after four week of post-surgical treatment including NSAIDs.

3. "Case report: However, I do not really understand, why the authors did not perform a "Tumor" debulking in a second procedure. Why did the authors not open the cubital tunnel, secondarily?"

Since the pathology revealed the diagnosis of a localized eosinophilic fasciitis there was no indication for a second surgical procedure. The reason is that this disease is treated solely by immunomodulation /-suppression. Therefore, a second surgical procedure wouldn't have been of any benefit. Most probably it would have led to an extensive scaring with a small chance of relief of the symptoms. In addition, a dissection of the cubital tunnel was and is not needed in the author's opinion because the "pseudo"-tumor was located above the cubital tunnel and was most likely the reason for the patient's clinical symptoms.

The new paragraph in the manuscript reads now as follows:

Therefore, the evaluation of treatment options showed the following:

In our case the pathology of the nerve's entrapment revealed the diagnosis of an eosinophilic fasciitis. Since the eosinophilic fasciitis is an inflammatory and usually systemic disease the general treatment consists of immune suppression with common drugs, especially corticosteroids in compliance with the actual literature [3]. Any further surgical procedure like tumor debulking or neurolysis was considered to be unrewarding in this special situation. Furthermore, in the author's opinion an additional surgical event would have the possible side effect of extensive scaring and could cause an even more severe entrapment of the nerve. However, in the patient's good state of health with no systemic affection by the disease there was currently no indication for a systemic therapy with corticosteroids. After a period of four weeks of intensive follow-ups combined with a NSAID-treatment (diclofenac and later ibuprofen in reducing doses) the patient reported of strongly attenuating neurologic symptoms over time.

## 4. "Case report: The authors should describe the different treatment option in more detail and justify their choice."

The authors agree with the reviewer's critique and therefore the paragraph about the treatment was edited and should now provide more information about the different treatment options in this case and why we have chosen the described one in the manuscript. This is now added in the revised version of the manuscript, marked by orange font and underlining and reads as follows:

Therefore, the evaluation of treatment options showed the following:

In our case the pathology of the nerve's entrapment revealed the diagnosis of an eosinophilic fasciitis. Since the eosinophilic fasciitis is an inflammatory and usually systemic disease the general treatment consists of immune suppression with common drugs, especially corticosteroids in compliance with the actual literature [3]. Any further surgical procedure like tumor debulking or neurolysis was considered to be unrewarding in this special situation. Furthermore, in the author's opinion an additional surgical event would have the possible side effect of extensive scarring and could cause an even more severe entrapment of the nerve. However, in the patient's good state of health with no systemic affection by the disease there was no currently indication for a systemic therapy with corticosteroids. After a period of four weeks of intensive follow-ups combined with a NSAID-treatment (diclofenac and later ibuprofen in reducing doses) the patient reported of strongly attenuating neurologic symptoms over time.

## 5. "Literature: Only a few citations are provided by the authors. However, Literature selection and relevance is appropriate and related to the rarity of the disease."

The reviewer points out an important fact when research is performed about rare diseases: There is only a rare number of reports in literature to the one described in the here presented case report as well. Therefore, the authors tried to highlight the background and initial description of the disease. Unfortunately, there are no big epidemiology reports about the eosinophilic fasciitis at all. The most valuable reports are larger case series. Regarding the circumstances of a localized appearance – like in our case – there are only a few reports in the literature at all, which have been cited throughout the manuscript. We hope that we could still fulfill the reviewer's reasonable claim for references regarding our report.

## 6. "Figures: The figures and legends are appropriate. However, the Definition of figures 2A and B should be improved."

We thank the reviewer for his comment. In the revised version of the manuscript the quality of the given figure is increased and reduced to only one image.

## Reviewers' Comments to Revision

### Reviewer 1: Holger Bannasch

Jan 30, 2017

<b>Reviewer Recommendation Term:</b>	Accept
<b>Overall Reviewer Manuscript Rating:</b>	70
<b>Custom Review Questions</b>	<b>Response</b>
Is the subject area appropriate for you?	4
Does the title clearly reflect the paper's content?	4
Does the abstract clearly reflect the paper's content?	4
Do the keywords clearly reflect the paper's content?	3
Does the introduction present the problem clearly?	3
Are the results/conclusions justified?	4
How comprehensive and up-to-date is the subject matter presented?	4
How adequate is the data presentation?	4
Are units and terminology used correctly?	4
Is the number of cases adequate?	N/A
Are the experimental methods/clinical studies adequate?	N/A
Is the length appropriate in relation to the content?	4

Does the reader get new insights from the article?	4
Please rate the practical significance.	3
Please rate the accuracy of methods.	2
Please rate the statistical evaluation and quality control.	N/A
Please rate the appropriateness of the figures and tables.	3
Please rate the appropriateness of the references.	4
Please evaluate the writing style and use of language.	4
Please judge the overall scientific quality of the manuscript.	3
Are you willing to review the revision of this manuscript?	Yes

**Comments to Authors:**

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**Reviewer 2: Thomas Kremer**

Feb 17, 2017

<b>Reviewer Recommendation Term:</b>	Accept
<b>Overall Reviewer Manuscript Rating:</b>	80

**Custom Review Questions**

Is the subject area appropriate for you?	<b>Response</b> 5 - High/Yes
Does the title clearly reflect the paper's content?	5 - High/Yes
Does the abstract clearly reflect the paper's content?	5 - High/Yes
Do the keywords clearly reflect the paper's content?	5 - High/Yes
Does the introduction present the problem clearly?	5 - High/Yes
Are the results/conclusions justified?	5 - High/Yes
How comprehensive and up-to-date is the subject matter presented?	5 - High/Yes
How adequate is the data presentation?	5 - High/Yes
Are units and terminology used correctly?	N/A
Is the number of cases adequate?	N/A
Are the experimental methods/clinical studies adequate?	N/A
Is the length appropriate in relation to the content?	5 - High/Yes
Does the reader get new insights from the article?	5 - High/Yes
Please rate the practical significance.	3
Please rate the accuracy of methods.	N/A
Please rate the statistical evaluation and quality control.	N/A
Please rate the appropriateness of the figures and tables.	5 - High/Yes
Please rate the appropriateness of the references.	5 - High/Yes
Please evaluate the writing style and use of language.	5 - High/Yes
Please judge the overall scientific quality of the manuscript.	3
Are you willing to review the revision of this manuscript?	Yes

**Comments to Authors:**

The revised manuscript is now appropriate for publication!