Case Report **Open Access**

Amitabha Chakrabarti*, Manujesh Bandyopadhyay and Biswarup Purkayastha

Malignant perivascular epitheloid cell tumour (PEComa) of the lung – a rare entity

DOI 10.1515/iss-2016-0032
Received October 28, 2016; accepted January 9, 2017; previously published online February 23, 2017

Abstract: Malignant Perivascular Epitheloid Cell Tumour (PEComa) of the lung is very rare, with only six cases reported in literature. This case presented with a large mass originating from right upper lobe of the lung with dilemma in its histopathological diagnosis and management. Postoperative histopathology after a right upper and middle lobectomy describes a tumour with an alveolar/nested pattern of growth and epitheloid morphology with expression of TFE-3 and diagnosed as PEComa. After 6 months the patient had a local recurrence inside the thorax & chest wall.This case qualifies it as a rare type of malignant PEComa with younger age of presentation, aggressive clinical behaviour & malignant histological features along with TFE3 positivity on immunohistochemistry. This case is probably the first of its kind with the largest reported size involving two lobes of the lung.

Keywords: HMB45 negative; lung; PEComa; TFE – 3 positive.

Introduction

Perivascular epithelioid cell (PEC) tumours are a rare group of mesenchymal tumours, with an abundance of periodic acid Schiff (PAS)-positive glycogen. These tumours are mostly seen in the gastrointestinal tract and pelvic organs and are mostly benign [1].

Owing to its rarity, the World Health Organization (WHO) defines a perivascular epithelioid cell (PEComa) tumour as a "mesenchymal tumour composed of histologically and immunohistochemically distinctive perivascular epithelioid cells".

The lung is an uncommon location for PEC tumour, which is otherwise commonly seen in the gastrointestinal tract and pelvic organs. Pulmonary PEC tumours also are mostly benign [2], with malignancy being very rare [3].

The diagnostic challenge arises from the silent and indolent course of this tumour, which is otherwise asymptomatic in most cases, with hemoptysis in only a few of them. Our case stands out in being a primary pulmonary PEComa with malignant characteristics, and adding to its rarity is the fact that the tumour showed a TFE (transcription factor 3) positivity. The complex characteristic and rare nature of the tumour accounted for a dilemma in diagnosis and management.

Case report

A 36-year-old nonsmoker, nonalcoholic, nondiabetic male presented with right-sided chest pain for 2 months and a history of hemoptysis. Chest X-ray revealed right upper zone haziness. The contrast-enhanced computed tomography (CECT) of the thorax showed large encapsulated heterogeneously enhancing mass in right paratracheal region indenting right upper lobe and lobar bronchi. It measured 86×78 mm, with a sharp defined margin and no obvious invasion or enlarged mediastinal nodes. Lung fields were otherwise normal. Further, a positron emission tomography (PET)/CT scan suggested an active primary disease in the large necrotic mass in the right upper lobe of the lung with mediastinal invasion, and diffuse bone marrow uptake metastatic involvement. The fine-needle aspiration cytology (FNAC) was consistent with non-small cell lung cancer (NSCLC). True-cut biopsy of the same lesion suggested TFE-3-positive alveolar soft-part sarcoma.

Presurgical workup indicated leukocytosis to the tune of >50,000/mm^3 with neutrophilic predominance. All other parameters were within normal limits.

Surgery was decided in a multidisciplinary team meeting. Right thoracotomy was done and a solid mass

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Manujesh Bandyopadhyay and Biswarup Purkayastha: Department of CTVS-NH Rabindranath Tagore International Institute of Cardiac Sciences, Kolkata, India

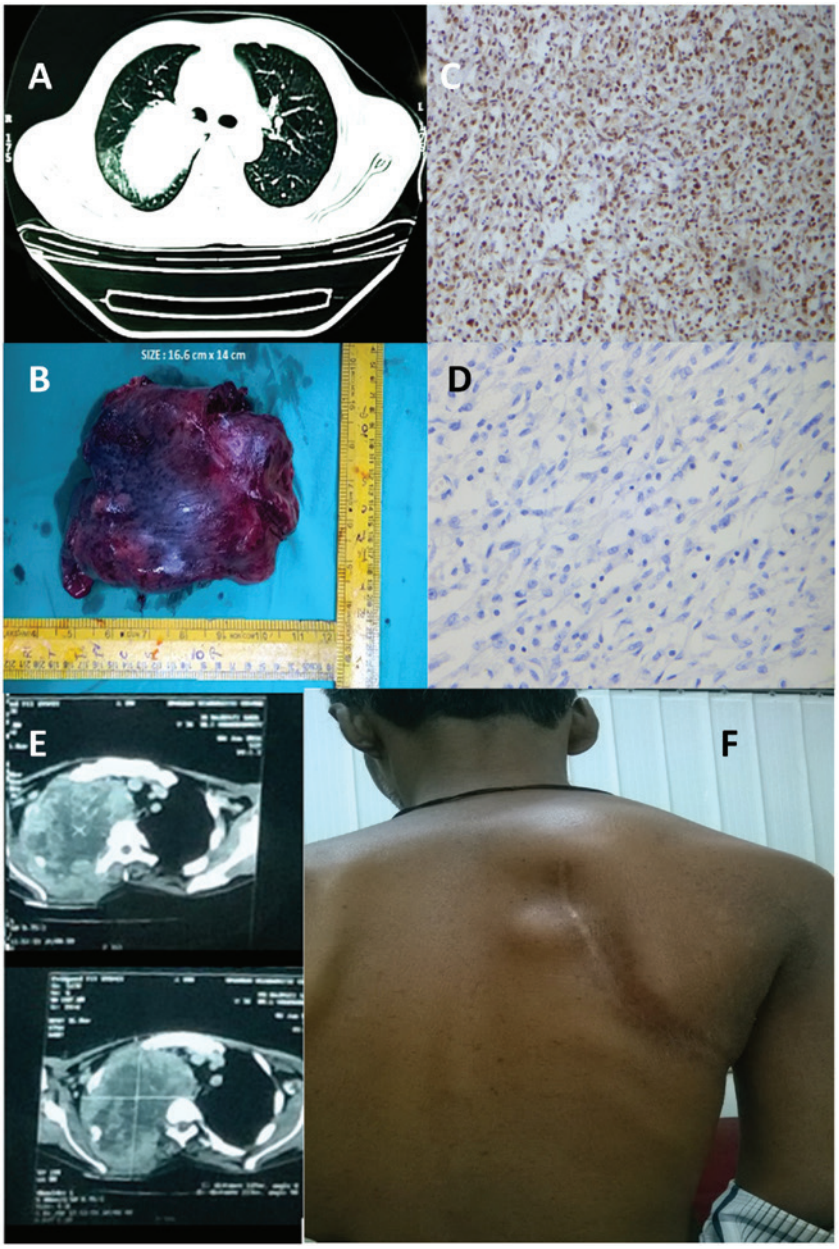

Figure 1: Pictures showing pre operative CT , histology, IHC, operative specimen and post operative recurrence.
(A) CT scan of thorax showing mass in the right hemithorax. (B) Resected mass. (C) TFE3 positivity in immunohistochemistry of resected mass. (D) HMB-45 positivity in immunohistochemistry of resected mass. (E) Slice of CT scan of thorax at 6 months follow-up showing recurrent mass in right hemithorax. (F) Swelling in the back and base of the neck at 6 months follow-up showing patients' back scar from earlier operation.

originating from the right upper lobe extending into the right middle lobe was found. A right upper and middle lobectomy was performed. The postresection mass measured 18×13 cm in size. The postoperative course was uneventful. On first postoperative day, leukocytosis plummeted to 12,000/mm^3.

Tissue was sent for histopathology, and the microscopy describes a tumour with an alveolar/nested pattern of growth and epitheloid morphology. Focally, the tumour cells had spindled morphology and were arranged in fascicles. A major portion was necrotic. The tumour cells expressed TFE-3, desmin (focal), and smooth muscle actin (SMA; focal) and were negative for cytokeratin, epithelial membrane antigen (EMA), CD68, HMB-45, Melan-A, S100 protein, myogenin, and microphthalmia transcription factor (MiTF). It was diagnosed to be a PEComa.

At 6 months postoperation, the patient came back with a heaviness in the right thorax. The CT scan of the thorax showed lung mass extending into the lower part of the neck up to the posterior paravertebral soft tissue with

Table 1: Relevant literature on malignant pecoma of the lung.

Author	Demographics	CT scan findings	Treatment	Immunohistochemistry marker	Remarks
Ye et al. [4]	50/F	4 cm round and well-circumscribed mass in RLL	Right lower lobectomy with mediastinal LN clearance	HMB-45, PNL2, A013	Negative for vimentin, AE1/AE3, and CAM2.5
Parfitt et al. [5]	53/F	5.4 cm mass in RUL with multiple lung nodules, 4.8 cm left adrenal mass, temporal lobe mass	Could not be operated		Brain metastasis after several months
Yan et al. [6]	78/F	3 cm coin lesion on chest radiograph	Surgical resection		No recurrence
Lim et al. [1]	63/M	12 cm well-circumscribed mass in LLL with nodules in both lungs	Left lower lobectomy	S100 and SMA	Appeared to be arising from LLL bronchus with dense pleural attachment; initially VATS attempted but converted to open; not positive for HMB-45
Liang et al. [7]	63/M	6.7 × 9.8 cm large mass with moderate heterogeneous enhancement in the anterior and middle mediastinum and with well-defined margins	Surgical excision of the mass	Vimentin, HMB-45, Melan-A, and Ki67	Pan-cytokeratin, EMA, and S100
Sambo [8]	40/M	7 cm centrally located mass in the left lower lobe of the lung with an endobronchial lesion	Given the location of the tumour and to achieve a negative margin of surgical resection, a left pneumonectomy was performed	Negative for S100, HMB-45, CD31, SMA, calponin, and desmin immunostatin	Final pathology yielded a poorly differentiated malignant epithelioid and spindle cell neoplasm, consistent with malignant PEComa

erosion of upper ribs and metastatic lesion in the right head of the humerus. The patient was sent to a medical oncologist for further management (Figure 1).

Comments and discussion

After an extensive search, we could only find six relevant cases in the literature (Table 1) of malignant PEC tumours of the lung. Unlike mesenchymal tumours that express positivity for HMB-45, our case is rare in the sense that this case had TFE3 positivity and HMB-45 negativity.

PEC is a type of cell present in a constellation of tumours viz. angiomyolipoma (AML), lymphangioleiomyomatosis (LAM), clear-cell sugar tumour (CCST), clear-cell myomelanocytic tumour of falciform ligament, and clear-cell tumour of other anatomical sites.

LAM is a rare tumour, the pulmonary equivalent of AML commonly affecting premenopausal women. It consists of a nodular, often widespread, and bilateral interstitial proliferation of smooth muscle cells positive for HMB-45, actin, and desmin. They cannot alter their immunophenotype like PEComa alternating between muscular and epithelioid components, neither do they express a wide range of melanocytic markers. The spindle-shaped cells are usually arranged around thin-walled, branching vascular channels. LAM is usually sporadic, with occasional association with tuberous sclerosis complex (TSC) [3].

CCST was originally described in the lung [9]. It is a rare and benign neoplasm composed of uniform round-to-polygonal epithelioid cells, with a clear or eosinophilic cytoplasm and well-defined borders. Tumour cells are surrounded by prominent and thin-walled vascular channels. CCST has a nested pattern similar to PEC tumour. It is also observed that adipocytic cells are seen between the nesting in a few cases of CCST [9]. Tumour cells are positive for HMB-45 like most mesenchymal tumours (unlike in our case, which is HMB-45 negative) [10–12]. In the vast majority of cases, it is a sporadic tumour with very occasional association with TSC.

PECs stand out from other mesenchymal tumours because they can modulate their morphology and immunophenotype. They can have more muscular feature with

a stronger positivity for actin compared to HMB-45 or more epithelioid features with stronger positivity for HMB-45 and mild reaction to actin [3]. PEC also expresses melanocytic markers, such as gp100 protein [monoclonal antibody (mAb) HMB-45], Melan-A, tyrosinase, and MiTF, and muscle markers, such as SMA, pan-muscle actin, muscle myosin, and calponin. Thirty percent of PEComas express desmin, which does not guarantee a true smooth muscle origin [13]. Also, it has recently been appreciated that approximately 30% of PEComas express S100 protein, and this too does not necessarily imply a melanocytic origin. It has been established that a subset of PEComas harbours TFE3 gene fusions (as in our case). The presenting features include younger age of presentation, absence of association with TSC, and strong (3+) immunoreactivity for TFE3. They present with malignant histological features and aggressive clinical behavior. Despite significant morphologic and immunohistochemical overlap, PEComas harbouring TFE3 gene fusions are nowadays taken as a distinctive entity [14].

Folpe et al. [13] reported a significant association between tumour size >5 cm, infiltrative growth pattern, high nuclear grade, necrosis and mitotic activity >1/50 HPF, and recurrence and/or metastasis of PEC tumours, with the tumour being malignant. In our case, the size (>5 cm in diameter), large area of necrosis, and recurrence within 6 months qualified it as a malignant PEComa. To our knowledge, an optimal treatment strategy does not exist for such an aggressive PEC tumour, although surgical resection remains the most common modality. Recently, limited studies have reported encouraging results of targeted therapy after an oral administration of mammalian target of rapamycin (mTOR) inhibitors in a metastatic retroperitoneal PEC. No consensus exists to the specific targeted therapy for this extremely rare malignant PEComa of the lung with metastasis, and a further study is required for rational usage.

There is no consensus on an optimal management strategy for such aggressive PEC tumour of the lung. However, surgical resection remains the most common and favoured modality of treatment with variable results. A few recent studies have shown encouraging results with oral mTOR targeted therapy and merit further research.

Author Statement

Funding: Authors state no funding involved. Conflict of interest: Authors state no conflict of interest. Informed consent: Informed consent has been obtained from all individuals included in this study. Ethical approval: The research related to human use complies with all the relevant national regulations and institutional policies and was performed in accordance with the tenets of the Helsinki Declaration and has been approved by the authors' institutional review board or equivalent committee.

Author Contributions

Manujesh Bandyopadhyay: writing of the manuscript; approval of the manuscript. Biswarup Purkayastha: writing of the manuscript; approval of the manuscript.

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Supplemental Material: The article (DOI: 10.1515/iss-2016-0032) offers reviewer assessments as supplementary material.

Reviewer Assessment **Open Access**

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Reviewers' Comments to Original Submission

Reviewer 1: anonymous

Nov 28, 2016

Reviewer Recommendation Term:	Revise with Major Modifications
Overall Reviewer Manuscript Rating:	50

Custom Review Questions	**Response**
Is the subject area appropriate for you?	3
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?	5 - High/Yes
Does the introduction present the problem clearly?	2
Are the results/conclusions justified?	3
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?	4
Is the number of cases adequate?	5 - High/Yes
Are the experimental methods/clinical studies adequate?	3
Is the length appropriate in relation to the content?	4
Does the reader get new insights from the article?	3
Please rate the practical significance.	2
Please rate the accuracy of methods.	3
Please rate the statistical evaluation and quality control.	4
Please rate the appropriateness of the figures and tables.	5 - High/Yes
Please rate the appropriateness of the references.	1 - Low/No
Please evaluate the writing style and use of language.	4
Please judge the overall scientific quality of the manuscript.	2
Are you willing to review the revision of this manuscript?	Yes

Comments to Authors:

The authors present one case of malignant PEComa that relapsed impressively within a very short interval following lobectomy. From the surgical point of view, the manuscript is informative insofar as the existance of this rare entitiy is communicated.

The short description of PEComas in the introduction, however, causes confusion rather than clarity, and should be written in a clear-cut way. Especially lymphangioleiomyatosis and tuberous sclerosis which are mentioned separately (once in the introduction and once in the discussion, respectively) should be put into clear context.

The biological properties of this very tumour are the central message of this article. The PEComa in this patient seems to have displayed a rather unusual immunohistochemical profile that has not been described before. Nevertheless, there are more and more recent publications on that topic than mentioned by the authors.

Reviewer 2: anonymous

Nov 05, 2016

Reviewer Recommendation Term:	Revise with Major Modifications
Overall Reviewer Manuscript Rating:	60

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?	4
Do the keywords clearly reflect the paper's content?	3
Does the introduction present the problem clearly?	3
Are the results/conclusions justified?	3
How comprehensive and up-to-date is the subject matter presented?	4
How adequate is the data presentation?	3
Are units and terminology used correctly?	1 - Low/No
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.	4
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.	1 - Low/No
Please rate the appropriateness of the references.	1 - Low/No
Please evaluate the writing style and use of language.	1 - Low/No
Please judge the overall scientific quality of the manuscript.	3
Are you willing to review the revision of this manuscript?	Yes

Comments to Authors:

Here is an interesting case report that is valuable because of the rare incidence of „Malignant Perivascular Epitheloid Cell Tumor of the Lung". Authors should be commended for writing a manuscript in a language different than their mother tongue. There are significant grammatical (present, past tense in the text ect.) and spelling errors.

The figures are well done and easily understandable exept figure E. Figure E should be just one axial CT slice of good quality showing the recurrent tumor. In addition please define all abbreviations when first used. All authors named in the text and table have to be cited properly according to the style of the journal in the references.

Overall, this is a highly interesting report of a very rare case with PEComa of the lung. Further intensive rework to meet the standards for publication of this journal had to be done

Authors' Response to Reviewer Comments

Dec 07, 2016

1] The short description of PEComas in the introduction, however, causes confusion rather than clarity, and should be written in a clear-cut way. Especially lymphangioleiomyatosis and tuberous sclerosis which are mentioned separately (once in the introduction and once in the discussion, respectively) should be put into clear context.- introduction and discussion changed and elaborated on the specific points
2] The biological properties of this very tumour are the central message of this article. The PEComa in this patient seems to have displayed a rather unusual immunohistochemical profile that has not been described before. Nevertheless, there are more and more recent publications on that topic than mentioned by the authors.- biological characteristics of the tumor and related discussions added in discussion and at least one more recent study added to table.
3] Authors should be commended for writing a manuscript in a language different than their mother tongue. There are significant grammatical (present, past tense in the text ect.) and spelling errors. - grammer and spelling checked with best of our knowledge
4] The figures are well done and easily understandable exept figure E. Figure E should be just one axial CT slice of good quality showing the recurrent tumor. In addition please define all abbreviations when first used. All authors named in the text and table have to be cited properly according to the style of the journal in the references. - figure E corrected and table referencing done

Reviewers' Comments to 1st Revision

Reviewer 1: anonymous

Dec 16, 2016

Reviewer Recommendation Term:	Accept with Minor Revision
Overall Reviewer Manuscript Rating:	80

Custom Review Questions	**Response**
Is the subject area appropriate for you?	3
Does the title clearly reflect the paper's content?	3
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?	4
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?	5 - High/Yes
Are the experimental methods/clinical studies adequate?	4
Is the length appropriate in relation to the content?	5 - High/Yes
Does the reader get new insights from the article?	3
Please rate the practical significance.	3
Please rate the accuracy of methods.	3
Please rate the statistical evaluation and quality control.	5 - High/Yes
Please rate the appropriateness of the figures and tables.	4
Please rate the appropriateness of the references.	5 - High/Yes
Please evaluate the writing style and use of language.	1 - Low/No
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 authors have made efforts to improvements to the manuscript, inserting passages about the different presentations of PEC-Oma. There is just the problem that these statements seem to be hardly connected to the rest of the manuscript. Some editing, some connecting, explaning words would be required.

The help of a native speaker is recommended.

Reviewer 2: anonymous

Dec 12, 2016

Reviewer Recommendation Term:	Accept
Overall Reviewer Manuscript Rating:	N/A

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?	4
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?	4
How adequate is the data presentation?	N/A
Are units and terminology used correctly?	3
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.	2
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.	3
Are you willing to review the revision of this manuscript?	No: accepted for publication

Comments to Authors:

I believe the report of this rare case is worthwhile to read.

Authors' Response to Reviewer Comments

Dec 22, 2016

We have taken the help of a native speaker to make the language more comprehensive and connected and as per the suggestions of reviewers.

Reviewers' Comments to 2nd Revision

Reviewer 1: anonymous

Jan 06, 2017

Reviewer Recommendation Term:	Accept
Overall Reviewer Manuscript Rating:	80

Custom Review Questions	**Response**
Is the subject area appropriate for you?	3
Does the title clearly reflect the paper's content?	5 - High/Yes
Does the abstract clearly reflect the paper's content?	4
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?	4

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?	5 - High/Yes
Is the number of cases adequate?	5 - High/Yes
Are the experimental methods/clinical studies adequate?	3
Is the length appropriate in relation to the content?	4
Does the reader get new insights from the article?	5 - High/Yes
Please rate the practical significance.	4
Please rate the accuracy of methods.	4
Please rate the statistical evaluation and quality control.	3
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.	1 - Low/No
Please judge the overall scientific quality of the manuscript.	3
Are you willing to review the revision of this manuscript?	No: Reviewed the manuscript twice already. Hardly any improvement in writing style.

Comments to Authors:

The description and the patho-histological background have been improved according to the suggestions.

Reviewer 2: anonymous

Jan 07, 2017

Reviewer Recommendation Term:	Accept
Overall Reviewer Manuscript Rating:	N/A

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?	4
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?	3
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?	3
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.	3
Please rate the appropriateness of the references.	3
Please evaluate the writing style and use of language.	2
Please judge the overall scientific quality of the manuscript.	3
Are you willing to review the revision of this manuscript?	No: Article accepted

Comments to Authors:

This case report is after all worth to be published. After major corrections There is a much better reading after major corrections.