Primary Synovial Sarcoma of the Inner Ear: A Case Report

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**Competing Interests:**
Authors declares no competing interests
Primary Synovial Sarcoma of the Inner Ear: A Case Report

Author(s): Glaoui M, Siham I, Meryem A, Faraj H, Mesmoudi M, Errihani H

Abstract

Ear synovial sarcoma is thus rare, to our knowledge, there has been only one reported case in English medical literature. Here we report a second case of a locally advanced synovial sarcoma affecting the inner ear in a thirty-year-old female managed by chemotherapy and radiotherapy. The objective of the present paper was to describe and discuss the diagnosis and treatment of this rare malignant inner-ear tumor.

Introduction

Malignant tumor of the ear is infrequent, with an incidence of 1 per six million of the population. Whatever the histological type, an inner ear location is exceptional. Diagnosis should not be delayed, so as to allow satisfactory tumor removal without risk of neurologic sequelles.

Case Report

A 30-year-old female presented in the medical oncology department of national institute of oncology with a painful contracture of facial muscles with a six months history of otorrhea associated with intermittent otalgia. Physical examination revealed a budding tumor filling the external auditory canal associated with complete peripheral right facial nerve palsy. Computed tomography scan of the face was performed and confirmed the presence of a 56*37 mass extending to the tympanum and external auditory canal with osteolysis of the mastoid. There was no evidence of metastasis to lung or other sites. A biopsy was performed and the diagnosis of a monophosphatic synovial sarcoma FNCLCC grade II was rendered.

Histological analysis of the tumor revealed a massive infiltration by an elongated cells with spindleshaped nuclei. The reticulin pattern was heavy with prominent vascularity and some dilated vascular channels. There were several foci of distinct cartilaginous differentiation within the tumour. Immunohistochemical study revealed a positive reaction for EMA and PSA 100 but negative for CD34, HMB45, KL1 and desmine. A final diagnosis of monophosphatic synovial sarcoma FNCLCC grade II was made.

Surgical approach wasn’t possible because of the extending of the tumor and the patient underwent three cycles of neo adjuvant chemotherapy consisting of doxorubicin (60mg/m2day1) and ifosfamide (3g/m2 day 1 to 3) regimen.

CT scan after final courses showed a progressive disease with skin ulceration, local bleeding and lung metastasis. A palliative radiotherapy was indicated followed by oral Cyclophosphamide. The patient died after 6 months.

Discussion

Although rare, synovial sarcoma is one of the most common malignant soft-tissue sarcomas in children and adolescents, the head and neck is involved in 6.8% of all synovial sarcomas. Ear location is extremely rare, only one case of a primary synovial sarcoma of the middle ear has been reported by O'keefee et al in 1993. Synovial sarcomas often arise in areas remote from structures containing synovial membrane such as joints or bursae. Mackenzie et al suggested that synovial sarcomas arise from undifferentiated mesenchymal tissue which retains the potential for synovioblastic differentiation. Immunohistochemical studies by Abenoza et al support the theory of mesenchymal origin demonstrating a positive immunoreaction for the epithelial markers, epithelial membrane antigen [EMA] and cytokeratin [CK]. The tumour is classically biphasic, containing both epithelial and spindle cells. The biphasic nature of the tumour is the only diagnostic histological criterion although a monophosphatic variety where either cell type predominates is recognized as in this case. Treatment of ear tumor depends on the degree of extension. Classically, it associates a complete surgical resection of the primary tumor and/or radiochemotherapy. Surgery should follow the rules of cancer surgery rather than the general principles of ear surgery. Removal is incomplete in 50% of cases, accounting for the high rate of local recurrence at 10 months. Prognosis is generally poor but depends on the time to
diagnosis [6].

Conclusion

Inner-ear synovial sarcoma is a rare malignant tumor that requires early diagnosis to allow a complete surgical resection which is the mainstay of treatment. A multidisciplinary approach associating otologist, surgeons, radiologist and oncologist is essential for efficient management.

References

Illustrations

Illustration 1

Axial bone marrow reconstruction on CT scan showing an osteolysis of the temporo mandibular articulation
Illustration 2

Axial parenchymal window on CT scan showing a tissular process sitting in the right temporal fossa
Illustration 3

Axial parenchymal window through the paranasal sinuses showing an increase of the lesion with a mass effect of the nasopharynx and osteolysis of
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Reviews

Review 1

Review Title: Primary Synovial Sarcoma of the Inner Ear: A Case Report

Posted by Dr. Wafaa Kaikani on 23 Jun 2011 05:41:23 PM GMT

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<td>Yes</td>
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<td>No</td>
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<td>Yes</td>
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<td>Can you suggest brief additions or amendments or an introductory statement that will increase the value of this paper for an international audience?</td>
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Rating: 5

Comment:
ligne 15 : the presence of a 56*37 mm mass extending..
ligne 24 : preoperative chemotherapy rather than neoadjuvant chemotherapy
ligne 33: Synovial sarcomas often arises
ligne 41: O'keefee et al in 1993 (3)
ligne 43: Mackenzie et al suggested .. reference ??

Concerning imaging: is MRI better ??

Competing interests: no

Invited by the author to make a review on this article? : No

Experience and credentials in the specific area of science:
experience in diagnosis and treatment of sarcoma.

Some articles published in other area.

Publications in the same or a related area of science: No

How to cite: Kaikani W.Primary Synovial Sarcoma of the Inner Ear: A Case Report[Review of the article 'Primary Synovial Sarcoma of the Inner Ear: A Case Report ' by ].WebmedCentral 1970;2(6):WMCRW00835
Review 2

Review Title: Synovial sarcoma ORL

Posted by Dr. Youssef Bensouda on 18 May 2011 01:03:22 PM GMT

1. Is the subject of the article within the scope of the subject category? Yes
2. Are the interpretations / conclusions sound and justified by the data? Yes
3. Is this a new and original contribution? Yes
4. Does this paper exemplify an awareness of other research on the topic? No
5. Are structure and length satisfactory? Yes
6. Can you suggest brief additions or amendments or an introductory statement that will increase the value of this paper for an international audience? No
7. Can you suggest any reductions in the paper, or deletions of parts? No
8. Is the quality of the diction satisfactory? Yes
9. Are the illustrations and tables necessary and acceptable? Yes
10. Are the references adequate and are they all necessary? Yes
11. Are the keywords and abstract or summary informative? Yes

Rating: 6

Comment:
Nice and original article,
I join the comments made by the other authors

Competing interests: no

Invited by the author to make a review on this article? : Yes

Experience and credentials in the specific area of science:
Oncology

Publications in the same or a related area of science: No

How to cite: Bensouda Y. Synovial sarcoma ORL [Review of the article ‘Primary Synovial Sarcoma of the Inner Ear: A Case Report ‘ by ]; WebmedCentral 1970;2(5):WMCRW00769
Review 3

Review Title: A very rare clinical entity: Sarcoma of the year

Posted by Prof. Esther Una Cidon on 12 Apr 2011 05:31:52 AM GMT

1. Is the subject of the article within the scope of the subject category? Yes
2. Are the interpretations / conclusions sound and justified by the data? Yes
3. Is this a new and original contribution? Yes
4. Does this paper exemplify an awareness of other research on the topic? No
5. Are structure and length satisfactory? Yes
6. Can you suggest brief additions or amendments or an introductory statement that will increase the value of this paper for an international audience? No
7. Can you suggest any reductions in the paper, or deletions of parts? No
8. Is the quality of the diction satisfactory? Yes
9. Are the illustrations and tables necessary and acceptable? Yes
10. Are the references adequate and are they all necessary? Yes
11. Are the keywords and abstract or summary informative? Yes

Rating: 0

Comment:
I think this is a very interesting article. It shows us a very rare clinical case of a primary synovial sarcoma arising of the inner ear. The communication of these kind of rare clinical cases is understandable from the point of view of increasing the doctors’ awareness about the relevance of expedite diagnosis and treatment and also to increase the knowledge of these diseases to make doctors suspect about their existence. Despite of the comments above, I think this article could be somewhat improved. Sometimes it needs a language polishing.

In the introduction the authors should include more data about the synovial sarcomas in general, frequency, most usual locations and therapeutic approaches. Then, authors could focus (as they have done) on the ear tumours and the relevance of early diagnosis and treatment to get the best results.

In the section “Case report”, authors should explain the abbreviation they have used FNCLCC (French Federation of Cancer Centers Sarcoma Group).

I would like to have seen if this case was treated in a multidisciplinary committee and if radiation therapy was considered as neoadjuvant treatment as chemotherapy was.

I miss some pictures about histology.

I would like also to know why this patient had received oral cyclophosphamide as palliative treatment once she had got progressive disease with the combination of ifosfamide and doxorubicin and for how long was she receiving this treatment and also the evolution she presented (stable disease, partial remission or progressive disease). On the other hand the dosis of palliative radiotherapy and the clinical benefits she got from this technique.

I think all these points would improve our knowledge about this rare disease.

In the end the references need to be reviewed (there is a mistake in reference 3).

Esther Una Cidon, MD, PhD, Professor

Invited by the author to make a review on this article? : Yes

Publications in the same or a related area of science: No

How to cite: Una Cidon E. A very rare clinical entity: Sarcoma of the year[Review of the article 'Primary Synovial Sarcoma of the Inner Ear: A Case Report ' by ].WebmedCentral 1970;2(4):WMCRW00664
Review 4

Review Title: Review of a Case Report on Primary Synovial Sarcoma of the Inner Ear

Posted by Dr. Thomas F Heston on 11 Apr 2011 03:56:56 PM GMT

| 1 | Is the subject of the article within the scope of the subject category? | Yes |
| 2 | Are the interpretations / conclusions sound and justified by the data? | Yes |
| 3 | Is this a new and original contribution? | Yes |
| 4 | Does this paper exemplify an awareness of other research on the topic? | Yes |
| 5 | Are structure and length satisfactory? | No |
| 6 | Can you suggest brief additions or amendments or an introductory statement that will increase the value of this paper for an international audience? | Yes |
| 7 | Can you suggest any reductions in the paper, or deletions of parts? | No |
| 8 | Is the quality of the diction satisfactory? | Yes |
| 9 | Are the illustrations and tables necessary and acceptable? | Yes |
| 10 | Are the references adequate and are they all necessary? | Yes |
| 11 | Are the keywords and abstract or summary informative? | Yes |

Rating: 6

Comment:
Thank you for this interesting case report.

I was only able to see the CT image before treatment, not afterwards. Also, do you have an MRI? Do you have a PET/CT image? In your description of the CT, please give units of measurement.

Some of the abbreviations you used were unfamiliar to me. Could you please give the full description instead of just abbreviations?

Competing interests: no

Invited by the author to make a review on this article? : No

Experience and credentials in the specific area of science: Molecular imaging physician.

Publications in the same or a related area of science: No


Webmedcentral > Case Report
Review 5

Review Title: Primary Synovial Sarcoma of the Inner Ear: A Case Report

Posted by Prof. Eman I El-Abd on 27 Mar 2011 09:02:22 AM GMT

1. Is the subject of the article within the scope of the subject category? Yes
2. Are the interpretations / conclusions sound and justified by the data? Yes
3. Is this a new and original contribution? Yes
4. Does this paper exemplify an awareness of other research on the topic? Yes
5. Are structure and length satisfactory? Yes
6. Can you suggest brief additions or amendments or an introductory statement that will increase the value of this paper for an international audience? Yes
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9. Are the illustrations and tables necessary and acceptable? Yes
10. Are the references adequate and are they all necessary? Yes
11. Are the keywords and abstract or summary informative? Yes

Rating: 5

Comment:
I suggest:
- refer to the grading system that was used
- add the illustrations for the IHC for EMA, PSA100, CD34, HMB45, KL1, and desmine with the proper positive and negative controls
- add the scoring system for these markers and illustrate why they have been used (e.g. desmine as myoepithelial marker, KL1 as epithelial marker .....etc)
- Explain why diagnosis was not confirmed by molecular markers
- Follow consistent method in writing references

Competing interests: No

Invited by the author to make a review on this article? : No

Experience and credentials in the specific area of science:
- oncology

Publications in the same or a related area of science: Yes

How to cite: El-Abd E. Primary Synovial Sarcoma of the Inner Ear: A Case Report [Review of the article 'Primary Synovial Sarcoma of the Inner Ear: A Case Report ' by ]. WebmedCentral 1970;2(3):WMCRW00629
Review 6

Review Title: Primary Synovial sarcoma of the inner ear?!

Posted by Dr. Narendra Hulikal on 26 Mar 2011 05:57:08 AM GMT

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Rating: 4

Comment:
I wish authors should elaborate on certain issues and also calrify few of them. Can they clarify the exact site of origin of the tumor as it appears clinically as well as on CT films to be involving external and middle ear, mastoid and temperomandibular area and not inner ear. There are case reports of the tumor arising from temperomandibular region (atleast 2). Authors should have provided illustration of microscopic findings. It is difficult to diagnose these tumors in small specimens even by experienced sarcoma pathologist. At present time diagnosis of monphasic synovial sarcoma needs demonstration of X;18 translocation which is characteristic of these tumors. Extensive articles on synovial sarcoma of head and neck and orbit can be found.

Competing interests: none

Invited by the author to make a review on this article? : No

Experience and credentials in the specific area of science: surgical oncologist in an academic institute

Publications in the same or a related area of science: No

How to cite: Hulikal N.Primary Synovial sarcoma of the inner ear?! [Review of the article ‘Primary Synovial Sarcoma of the Inner Ear: A Case Report ’ by ].WebmedCentral 1970;2(3):WMCRW00627
Review 7

Review Title: Primary Synovial Sarcoma of the Inner Ear: A Case Report

Posted by Dr. Juan S Yakisich on 23 Mar 2011 09:25:23 PM GMT

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Rating: 5

Comment:
This manuscript reports a rare case of a primary case of synovial sarcoma localized in the middle ear. The manuscript may need the addition of the histological analysis. Minor corrections are also needed (e.g. consistency in the references).

Competing interests: None

Invited by the author to make a review on this article? : No

Experience and credentials in the specific area of science:
Cancer.

Publications in the same or a related area of science: No

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