Multicentric epithelioid hemangioendothelioma: An unusual case report

Liqa Al Mulla, Jawad Al Khalaf, Ayesha Ahmed, Areej Al Nemer, Yasser El-Ghoniemy, Tarek M El-Sharkawy

ABSTRACT

Epithelioid hemangioendothelioma (EHE) is a rare malignant tumor of vascular origin. Most of the times it affects liver, lung and bones, although this kind of tumor may involve the head and neck area, breast, lymph nodes, mediastinum, brain and meninges, spine, skin, abdomen and many other sites. We report a case of a 59-year-old Jordanian man, who was found to have multiple lesions, in the liver, anterior abdominal wall and the lung. Due to the short duration between the presentation, and as most common reported EHE metastatic cases occur in the bone, we raise up the possibility of primary EHE multi-centric lesions from the beginning rather than metastasis.
Multicentric epithelioid hemangioendothelioma: An unusual case report

Liqa Al Mulla, Jawad Al Khalaf, Ayesha Ahmed, Areej Al Nemer, Yasser El-Ghoniemy, Tarek M El-Sharkawy

ABSTRACT

Epithelioid hemangioendothelioma (EHE) is a rare malignant tumor of vascular origin. Most of the times it affects liver, lung and bones, although this kind of tumor may involve the head and neck area, breast, lymph nodes, mediastinum, brain and meninges, spine, skin, abdomen and many other sites. We report a case of a 59-year-old Jordanian man, who was found to have multiple lesions, in the liver, anterior abdominal wall and the lung. Due to the short duration between the presentation, and as most common reported EHE metastatic cases occur in the bone, we raise up the possibility of primary EHE multi-centric lesions from the beginning rather than metastasis.

Keywords: Epithelioid, Hemangioendothelioma, Multicentric

INTRODUCTION

Epithelioid hemangioendothelioma (EHE) is an uncommon low-grade malignant tumor of vascular origin that may develop in the soft tissue, lung, bone, brain, liver, and small intestine. However, the recent (2002) World Health Organization (WHO) classification does not strictly define these lesions as having intermediate behavior, but instead describes them as lesions that fall into the category of locally aggressive tumors and those with metastatic potential [1].

CASE REPORT

A 59-year-old Jordanian male who sought medical advice complaining of chronic fatigue, abdominal discomfort and weight loss. Abdominal examination revealed a huge nodular liver and moderate splenomegaly. The positive lab findings were leukocytosis, thrombocytosis, low Hg level with low MCV and MCH, high ESR, elevated alkaline phosphatase and GGTP. Computed tomography scan of chest and abdomen showed multiple bilateral hypodense nodules in the lungs, liver and spleen. He had been followed-up by cardiothoracic and oncology departments for many years. One month back, he presented to the surgical clinic with an anterior abdominal wall hyperpigmented lesion. Physical examination showed a dark grey firm nodule on anterior abdominal wall hypopigmented lesion. Histopathological findings revealed a dermal neoplasm formed of nests and cords of epithelioid and spindle cells embedded in fibro collagenous stroma (Figure 1A). The cells have abundant cytoplasm with occasional vacuoles and lumena, some of them have red blood cells (Figure 1B).
There is low mitotic activity and mild to moderate pleomorphism. Some of the nuclei are grooved. Scattered eosinophils are present within tumor cells and in stroma.

Immunohistochemical stains show positivity for Cluster of differentiation (CD31) (Figure 2) CD34, factor VIII and focal positivity for S100 protein in neoplastic cells and negativity for cytokeratin (CK), carcinoembryonic antigen (CEA) and tumor protein 63 (p63).

A diagnosis of epithelioid hemangioendothelioma was made. One month later, when the liver biopsy was taken from an outside hospital, the slides were received and showed the same histopathological and immunohistochemical findings. In the same month, the right lung nodule was discovered, with similar findings histopathologically and immunohistochemically, consistent with diagnosis of epithelioid hemangioendothelioma. The patient was doing well after receiving imatinib that was discontinued later on because he was intolerant to it as he developed gastric upset. On October 2016, he developed severe jaundice, tense ascites, melena and due to respiratory failure he passed away (Figure 3).

DISCUSSION

This case of a 59-year-old male with epithelioid hemangioendothelioma presented with multiple lesions found sequentially in anterior abdominal wall, liver and lung.

Most of reported cases of EHE in literature have single organ involvement. However, EHE can arise from many organs, including lungs, liver, bone, and soft tissue, simultaneously or sequentially. When this occurs, it may be difficult to determine if the tumor is multicentric from the beginning or if there is a primary lesion with metastases to the other organ tissue.

Hua Zhang et al. reported a case of a 20-year-old male, who presented with a right knee pain for eight months and diagnosed as EHE [2]. Lucas Rios Torres et al. reported a case of a 28-year-old female presented with a hypoechogenic hepatic nodule incidentally found at routine ultrasonography (US), discovered to be an EHE after hematoxylin and eosin and immune histochemical staining [3]. Muna M. Dahabreh et al. in 2011 reported a case of a 12-year-old with epithelioid hemangioendothelioma presented with simultaneously found multiple lesions in the lungs, trachea, liver and abdominal rectal muscle [4].

Jinghong et al. reported one case of a 20-year-old female with pulmonary epithelioid hemangioendothelioma accompanied by bilateral multiple calcified nodules in lung [5]. In 2010, Madhusudhan et al. reported a case of an 11-year-old boy with hemoptysis who was diagnosed with EHE simultaneously involving lung and liver [6]. Kalra et al. reported a case of a 70-year-old female with coexistent hepatic and pulmonary epithelioid hemangioendothelioma [7]. Al-Shraim et al. reported a case of a 51-year-old man with primary pleural epithelioid hemangioendothelioma with metastases to the skin [8]. Adher et al. in 2005 reported a case of a child with syncopal episodes who was found to have generalized multifocal EHE lesions in bones, lung, kidney and liver [9]. Besides, Kasteren et al. reported a single case of EHE which was misdiagnosed initially as lung histiocytosis but was later found to have multi-organ involvement at autopsy [10] (Table 1).
Our case was initially presented with anterior abdominal wall hyperpigmented nodule, where skin biopsy was obtained and was confirmed to be EHE lesion. In less than one month, a lung nodule was discovered incidentally, and a tissue biopsy taken, in the same time where we received an outside slides of a liver biopsy, all consistent with the diagnosis of EHE.

**CONCLUSION**

Epithelioid hemangioendothelioma (EHE) is a rare tumor of vascular origin, where patients present with multi-nodular lesions involving more than one organ. Due to the short duration between the presentation, and as most common reported EHE metastatic cases occur...
in the bone, we raise up the possibility of primary EHE multi-centric lesions from the beginning rather than metastasis.

Author Contributions
Liqa Al Mulla – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Jawad Al Khalaf – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Ayesha Ahmed – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Areej Al Nemer – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Yasser El-Ghoniemy – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Tarek M El-Sharkawy – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

Copyright
© 2017 Liqa Al Mulla et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.

REFERENCES
About Edorium Journals
Edorium Journals is a publisher of high-quality, open access, international scholarly journals covering subjects in basic sciences and clinical specialties and subspecialties.

Invitation for article submission
We sincerely invite you to submit your valuable research for publication to Edorium Journals.

But why should you publish with Edorium Journals?
In less than 10 words - we give you what no one does.

Vision of being the best
We have the vision of making our journals the best and the most authoritative journals in their respective specialties. We are working towards this goal every day of every week of every month of every year.

Exceptional services
We care for you, your work and your time. Our efficient, personalized and courteous services are a testimony to this.

Editorial Review
All manuscripts submitted to Edorium Journals undergo pre-processing review, first editorial review, peer review, second editorial review and finally third editorial review.

Peer Review
All manuscripts submitted to Edorium Journals undergo anonymous, double-blind, external peer review.

Early View version
Early View version of your manuscript will be published in the journal within 72 hours of final acceptance.

Manuscript status
From submission to publication of your article you will get regular updates (minimum six times) about status of your manuscripts directly in your email.

Our Commitment

Six weeks
You will get first decision on your manuscript within six weeks (42 days) of submission. If we fail to honor this by even one day, we will publish your manuscript free of charge.*

Four weeks
After we receive page proofs, your manuscript will be published in the journal within four weeks (31 days). If we fail to honor this by even one day, we will publish your manuscript free of charge and refund you the full article publication charges you paid for your manuscript.*

Favored Author program
One email is all it takes to become our favored author. You will not only get fee waivers but also get information and insights about scholarly publishing.

Institutional Membership program
Join our Institutional Memberships program and help scholars from your institute make their research accessible to all and save thousands of dollars in fees make their research accessible to all.

Our presence
We have some of the best designed publication formats. Our websites are very user friendly and enable you to do your work very easily with no hassle.

Something more...
We request you to have a look at our website to know more about us and our services.

* Terms and condition apply. Please see Edorium Journals website for more information.