Clinical Case Report

Medicine®

OPEN

Alveolar soft part sarcoma with multiple brain and lung metastases in pregnancy

A case report and literature review

Yubo Wang, MD[a], Jiayue Cui, PhD[b], Xu Yan, MD[c], Rihua Jin, MD[a], Xinyu Hong, MD[a,*]

Abstract

Rational: Alveolar soft part sarcomas (ASPSs) with multiple brain metastases in pregnancy is a rare entity.

Patient concerns: We report our experience with a 19-year-old pregnant woman who presented with intermittent headaches and vomiting at 38 weeks gestation.

Diagnoses: The patient was initially diagnosed as brain metastasis according to computed tomography and magnetic resonance imaging (MRI) imaging.

Interventions: Cesarean section and craniotomy (complete resection of both brain metastatic lesions) was performed sequentially.

Outcomes: A healthy baby girl was delivered safely and no neonatal malformations were found. Histological analysis confirmed the diagnosis of ASPS. Follow-up MRI performed 10 months after surgery revealed no residual tumor or signs of recurrence.

Lessons: We report a case of ASPS with multiple brain and lung metastases in a pregnant woman. We recommend timely MRI examination for diagnosis and have discussed the approach to the treatment of pregnant women with brain metastasis.

Abbreviations: ASPS = alveolar soft part sarcoma, MRI = magnetic resonance imaging.

Keywords: alveolar soft part sarcoma, brain metastases, brain neoplasms, pregnant women

1. Introduction

Alveolar soft part sarcomas (ASPSs) are highly vascular, rare malignant soft tissue tumors that typically occur in the extremities of adolescents and young adults. These account for approximately 1% of all soft tissue sarcomas. ASPS was first described by US scientists in 1952.[1] ASPS with multiple brain metastases in pregnancy is a rare entity. To the best of our knowledge, ASPS with multiple brain metastases in a pregnant woman has not been previously reported. Here, we report a patient with such a lesion and review the related literature.

Editor: N A.

YW and JC contributed equally to this work.

Informed Patient Consent: The patient has consented to submission of this case report to the journal.

The authors have no funding and conflicts of interest to disclose.

[a] Department of Neurosurgery, First Hospital of Jilin University, [b] Department of Histology and Embryology, College of Basic Medical Sciences, Jilin University, [c] Department of Pathology, First Hospital of Jilin University, Changchun, Jilin, P.R. China.

[*] Correspondence: Xinyu Hong, Department of Neurosurgery, First Hospital of Jilin University, 71, Xinmin Street, Changchun 130021, Jilin, P.R. China (e-mail: drhongxinyu@163.com).

Medicine (2017) 96:46(e8790)

Received: 23 October 2017 / Accepted: 27 October 2017

http://dx.doi.org/10.1097/MD.0000000000008790

2. Case report

A 19-year-old pregnant woman (38 weeks gestation) was admitted to the Department of Maternity with a history of intermittent severe headache and vomiting since 20 days. Magnetic resonance imaging (MRI) revealed 2 brain masses, 1 each in the left occipital lobe and the left cerebellar hemisphere, accompanied by severe peritumoral edema. As compared to the gray matter, the lesions appeared slightly hypointense on T1- and slightly hyperintense on T2-weighted sequences; the lesions showed homogeneous enhancement following administration of a contrast agent (Fig. 1). Lung computed tomography was suggestive of multiple metastases. A caesarean section was performed under general anesthesia on the 3rd day, and a healthy baby girl was delivered safely. No neonatal malformations were found. The patient was transferred to the Department of Neurosurgery on the 4th day. Emergency craniotomy was performed due to the increased intracranial pressure. Gross total resection of both tumors was performed separately. Macroscopically, the tumors were purple in color and were of firm consistency. Histologically, large, round tumor cells were found to be arranged in nests separated by delicate fibrovascular stroma. Immunohistochemical staining revealed that the lesions were positive for lysozyme, CD10, TFE3, and ki-67, but negative for glial fibrillary acidic protein, s-100, and epithelial membrane antigen (Fig. 2). The pathological features were strongly suggestive of ASPS with multiple brain metastases. Postoperatively, intracranial pressure was markedly alleviated. However, the patient refused positron emission tomography–computed tomography scanning and further radiotherapy, chemotherapy for "personal reasons." She was discharged a few days later, postoperative period was uneventful. Follow-up MRI performed 10 months after surgery

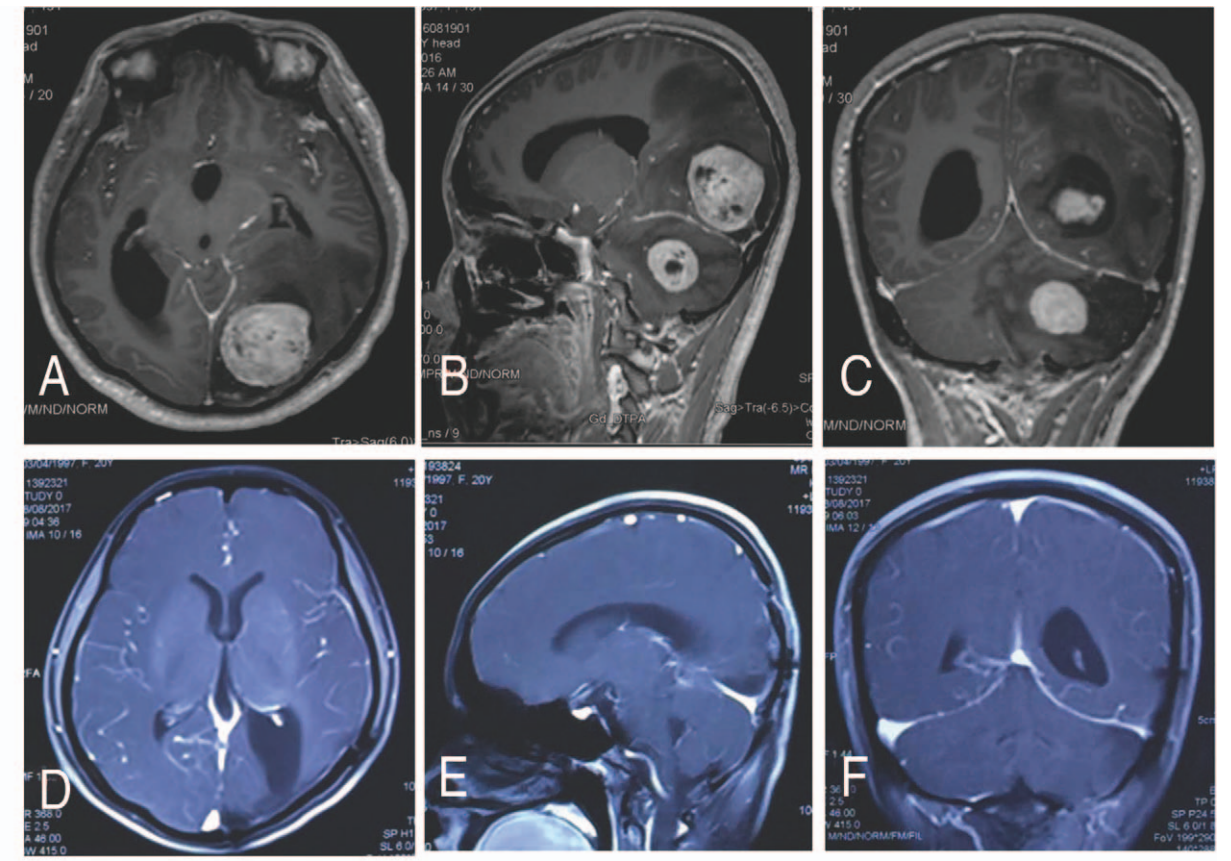

Figure 1. Preoperative and postoperative brain magnetic resonance imaging (MRI). Preoperative contrast-enhanced MRI (Gd-diethylenetriamine pentaacetic acid) showed 2 enhanced lesions, located at left occipital lobe and the left cerebellar hemisphere (A, axial view; B, sagittal view; and C, coronal view). MRI examination at 10 months after the operation showed no residual tumor or signs of recurrence (D, axial view; E, sagittal view; and F, coronal view).

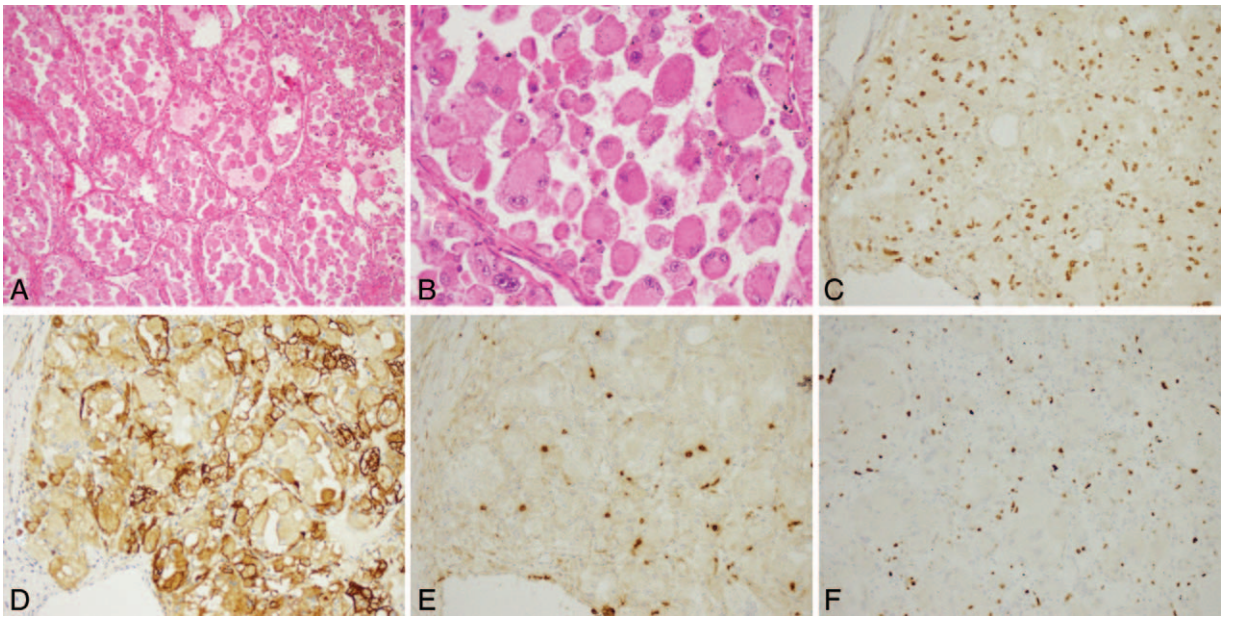

Figure 2. Microscopic characteristics of the resected tumor tissue. Hematoxylin-eosin staining at low (A, ×10) and high magnification (B, ×40). Immunohistochemical examination revealed positive staining for TFE3 (C, ×20), CD10 (D, ×20), lysozyme (D, ×40), and ki-67 (F, ×40).

revealed no residual tumor or signs of recurrence of the brain tumor (Fig. 1).

3. Discussion

Most ASPSs occur in adolescents and young adults in the age-group of 15 to 35 years. Some series show a female predilection especially during the 1st 2 decades of life.[2] ASPS most commonly affects the soft tissues of the pelvic cavity and lower limbs; however, the head and neck are common sites of lesion in children. Some patients have metastatic disease at the time of diagnosis. Lungs, bone, central nervous system, and liver are the most common sites of metastasis.[3] These typically exhibit high signal intensity on T1- to T2-weighted MRI images and show marked contrast enhancement.[2,3]

The prognosis is typically poor. Age at diagnosis, size of the tumor, and the presence of metastatic disease at presentation are the most important prognostic factors. Despite availability of chemotherapy, gene therapy, and other therapeutic modalities, radical resection is considered the treatment of choice.[2,3] A characteristic nonreciprocal translocation in the *ASPSCR1* gene on chromosome 17 and in *TFE3* gene on the X chromosome has been described in these patients. The *ASPSCR1-TFE3* fusion gene is postulated to be involved in the pathogenesis of this tumor.[4] It is hypothesized that the female predominance is due to the presence of an extra X chromosome and to the fusion gene not being subject to X-inactivation.[5]

The first published report of a brain tumor diagnosed in a pregnant woman was authored by Bernard in 1898.[6] Advances in the treatment of brain tumors have improved the prospects of successful pregnancy in women with brain tumors.[7] Management of these patients requires a multidisciplinary approach with involvement of physicians from different specialties.[6] A review of literature revealed that reports of brain metastasis in pregnant women are exceedingly rare. To our knowledge, this is the 1st report of ASPS with multiple brain and lung metastases in a pregnant woman.

The diagnosis of brain tumor in the present case was established very late because of the pregnancy. The patient may have had a relatively long history of headache, but which was likely ignored and attributed to the pregnancy. A timely MRI examination of the brain may have facilitated an early diagnosis and averted the development of a life-threatening situation. Cesarean section and craniotomy were successfully performed, and both tumors were successfully resected simultaneously. We believe that complete resection should be recommended in such cases. In the present case, the patient was satisfied with the treatment outcomes. Because the prognosis of multiple metastatic ASPS was poor, she refused chemoradiotherapy in order to be able to spend more time with the newborn.

In our opinion, neurosurgical resection is the cornerstone of treatment of pregnant women with increased intracranial pressure caused by brain metastasis. If the diagnosis is confirmed during 1st trimester, termination of pregnancy is recommended. After complete resection of brain tumors, combined treatment including chemotherapy, radiotherapy, and/or surgical resection of primary extracranial tumors should be performed in time. If the condition is diagnosed in 2nd trimester, neurosurgical resection and chemotherapy are suggested under the premise of fetal safety (during the 2nd and 3rd trimesters surgery is considered safe[8]). Combined therapy should subsequently be performed after delivery. If diagnosed in the 3rd trimester, cesarean section or natural delivery should be the preferred approach. Neurosurgical resection and combined therapy are suggested sequentially.

4. Conclusion

Cases of ASPS with multiple brain metastases are not well-documented in contemporary literature. We report a case of ASPS with multiple brain and lung metastases in a pregnant woman, which, probably, has not been reported before. We recommend timely MRI examination for diagnosis and have discussed the approach to treatment of pregnant women with brain metastasis.

References

[1] Christopherson WM, Foote FJ, Stewart FW. Alveolar soft-part sarcomas; structurally characteristic tumors of uncertain histogenesis. Cancer 1952;5:100–11.
[2] Zarrin-Khameh N, Kaye KS. Alveolar soft part sarcoma. Arch Pathol Lab Med 2007;131:488–91.
[3] Jaber OI, Kirby PA. Alveolar soft part sarcoma. Arch Pathol Lab Med 2015;139:1459–62.
[4] Ladanyi M, Lui MY, Antonescu CR, et al. The der(17)t(X;17)(p11;q25) of human alveolar soft part sarcoma fuses the TFE3 transcription factor gene to ASPL, a novel gene at 17q25. Oncogene 2001;20:48–57.
[5] Bu X, Bernstein L. A proposed explanation for female predominance in alveolar soft part sarcoma. Noninactivation of X; autosome translocation fusion gene? Cancer 2005;103:1245–53.
[6] Bonfield CM, Engh JA. Pregnancy and brain tumors. Neurol Clin 2012;30:937–46.
[7] Yust-Katz S, de Groot JF, Liu D, et al. Pregnancy and glial brain tumors. Neuro Oncol 2014;16:1289–94.
[8] Verheecke M, Halaska MJ, Lok CA, et al. Primary brain tumours, meningiomas and brain metastases in pregnancy: report on 27 cases and review of literature. Eur J Cancer 2014;50:1462–71.