Clinical Case Report

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Primary intracranial leiomyosarcoma in an immunocompetent patient

Case report with emphasis on imaging features

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Abstract

Rationale: Primary intracranial leiomyosarcoma (LMS) is an extremely rare tumor in the central nervous system (CNS), and usually seen in immunocompromised individuals. Only a few cases of primary intracranial LMS have been documented in the literature and no study focused on their MRI findings. We reported a case of primary intracranial leiomyosarcoma in a immunocompetent patient and review its imaging features.

Patient concerns: A 20-year-old female was admitted to our hospital, complaining with nausea, weight loss and progressive headache in recent 2 years.

Diagnosis: The magnetic resonance imaging scan of the brain revealed a large well-defined extra-cerebral mass adherent to left temporal meninges. The mass was iso-intense on T1-weighted images (T1WI), lightly iso to hypointense on T2-weighted images (T2WI) and enhanced uniformly with contrast medium. The preoperative diagnosis is meningioma.

Interventions: A left craniotomy was performed for the complete resection of the mass.

Outcomes: The lesion was diagnosed via surgical histopathology and immunochemistry as leiomyosarcoma. No evidence of recurrence and complications were found in the following 13 months.

Lessions: Primary intracranial leiomyosarcoma which has some special MR imaging features should be considered in the differential diagnosis of intracranial tumor.

Abbreviations: CNS = central nervous system, EBV = Epstein–Barr virus, HIV = human immunodeficiency virus, LMS = leiomyosarcoma, MRI = magnetic resonance images.

Keywords: brain neoplasms, immunocompetent, leiomyosarcoma, magnetic resonance images

1. Introduction

Primary intracranial leiomyosarcoma (LMS) occurs rarely in the central nervous system and is usually associated with radiation exposure,[1,2] immunocompromised states,[3] and infections with human immunodeficiency virus (HIV)[4–11] and Epstein–Barr virus (EBV).[6,8,10,12] Primary intracranial LMS in immunocompetent patient is exceedingly rare, with only a few cases documented in the literature[13–18] and no study focused on their MRI findings. This study reported a case of primary intracranial leiomyosarcoma with its MRI findings in an immunocompetent patient, and presented a brief review of MR imaging features of this disease.

2. Case report

A 20-year-old female was admitted to our hospital, complaining with nausea, weight loss, and progressive headache in recent 2 years. The patient had no history of smoking, intravenous drug abuse, or sexual promiscuity. The physical examination revealed mild weakness of left upper and lower extremities. Sensory changes were not noted. The blood examination findings were normal. Viral serology for HIV was also negative. The magnetic resonance imaging scan of the brain revealed a $6.8 \times 7.5 \times 6.8$ cm well-defined extra-cerebral mass adherent to left temporal meninges. The mass was iso-intense to the gray matter on T1-weighted images (T1WI) (Fig. 1A) and lightly iso to hypointense on T2-weighted images (T2WI) (Fig. 1B). The mass enhanced uniformly with contrast medium, and compressed the temporal lobe (Fig. 1C). With a preoperative diagnosis of meningioma, a left craniotomy was performed for the complete resection of the

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The patient was informed that the data from her case would be submitted for publication and provided consent.

The authors have no conflicts of interest to disclose.

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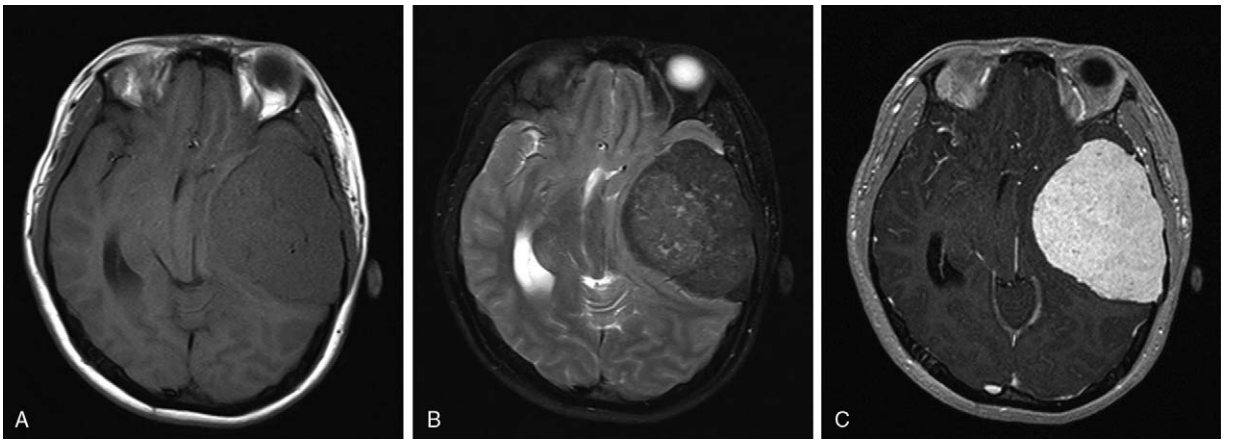

Figure 1. T1-weighted axial images of MRI (A) showed a large, well-defined, and iso-intense mass adherent to left temporal meninges. T2-weighted axial images (B) showed the tumor was lightly iso to hypointense and has no peritumoral edema. The contrast enhanced MRI revealed a markedly homogeneous enhancement and the temporal lobe was compressed (C). MRI = magnetic resonance images

mass. The lesion was fleshy, solid, and exhibited extensive vasculature. The mass lesion was found adherent to the dura and had a clear plane separating it from the brain.

Microscopic examination revealed that the tumor consisted of spindle-shaped cells with presence of mitotic figures and nuclear atypia (Fig. 2A). Immunohistochemical staining was positive for smooth muscle actin (SMA) (Fig. 2B), desmin, and H-caldesmon but negative for human melanoma black-45 (HMB-45), myogenin, S-100, epithelial membrane antigen (EMA), CD34 and carcinoembryonic antigen (CEA). The Ki-67 rate was 5–10%. Overall, the histopathological examinations confirmed a diagnosis of leiomyosarcoma. As most intracranial leiomyosarcomas represent metastatic disease, an intensive search for extracranial primary sites including full body CT, bone scan, and serum tumor markers were performed and no positive findings were found. Thereby a diagnosis of primary intracranial LMS was established. Two months after the surgery, the patient underwent concurrent chemoradiotherapy. The patient has no reoccurrence of primary intracranial leiomyosarcoma in the following 13 months.

3. Discussion

Intracranial leiomyosarcoma is a rare tumor in CNS. The majority of intracrannial leiomyosarcomas are metastasized from gastrointestinal tract or female reproductive system. In contrast, primary intracranial LMS is much rarer. Previous studies reported that less than 1% of brain biopsies (or 3 out of 25,000 brain tumors) are positive for LMS.[19] Primary intracranial LMS can occur in both adult and pediatric populations with no sex predilection.[3,6,8,9,18,19] No specific symptoms were associated with these cases of intracranial LMS, and clinical presentations varied depending on the size and location of the tumor. The most common symptom was headache as the present case.

The cellular origin and tumorgenesis of primary intracranial LMS remains unclear. The tumor was deemed probably arising from the mesenchymal cells of the dura matter or the cerebral blood vessel epithelium.[20] Most cases suggested the tumor might arise from the dura,[10,11,16,21,22] which was supported by our present case showing dural-based masses. Besides, there were still few cases reporting intra-axial lesions suggested the tumor may

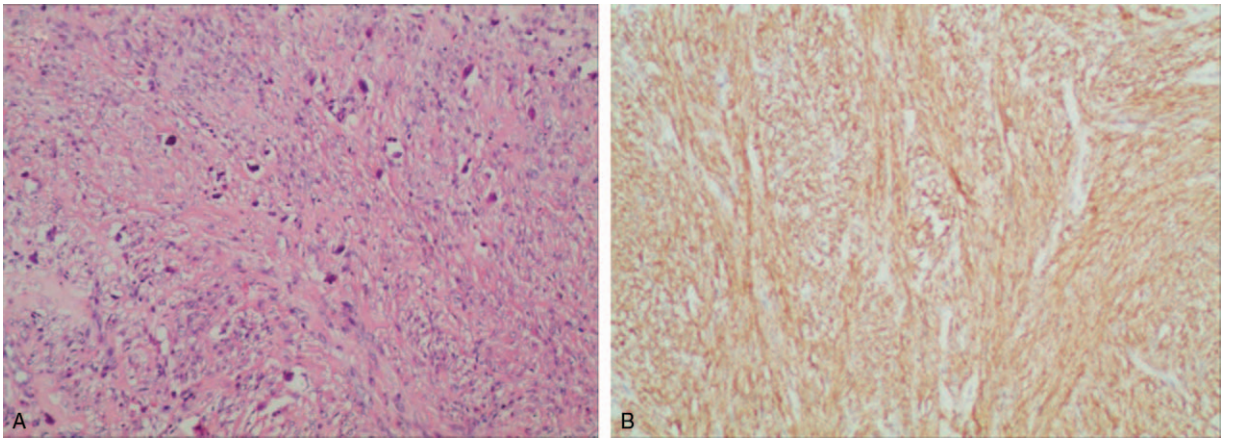

Figure 2. The histology, H&E staining of the tumor showed characteristic spindle cell and moderate unclear polymorphism (A). The tumor cells expressed smooth muscle actin (SMA) (B) (original magnification, ×100). SMA = smooth muscle actin.

arise from the cerebral blood vessel epithelium.[9,23] Although primary intracranial LMS has been reported in both immunocompetent and immunocompromised patients, it is more likely to develop in immunocompromised patients, especially in patients with HIV and Epstein–Barr virus infections. Therefore immunodeficiency plays an important role in the pathogenesis of primary intracranial LMS. In our case, the patient was negative for HIV serology, and had no significant medical or family history that may indicate the possibility of immunodeficiency. The test for EBV was not measured because the patient refused. There is no special risk factor for this lesion in our patient and the pathogenesis needs further study.

As the main imaging study for these lesions, MRI can excellently demonstrate the internal architecture and extent of tumors. We reviewed the literature of MR imaging of primary intracranial LMSs and found that primary intracranial LMS has some specified MR imaging features: most cases of primary intracranial LMSs showed single lesion, only one study reported multiple lesions.[24] Primary intracranial LMS can present as either extra-cerebral or intra-cerebral tumors. The imaging features of extra-cerebral and intra-cerebral lesions are different. Most extra-cerebral lesions including our case which appear as circumscribed dural-based tumors usually arise in the dura, sellar and parasellar region, cerebellopontine angle, and cavernous sinuses.[4,6,10–12,15,16,18,21] These lesions typically showed homogeneous hypointense or isointense to the gray matter on T1WI, lightly iso to hypointense or hyperointense on T2WI, and equally homogeneous enhancement with or without dural tail after administration of gadolinium, resembling meningioma.[4,10,11,16,21] The lesions can also infrequently involve the skull.[25] On the other hand, primary intracranial LMS presenting as intra-cerebral mass were also reported.[8,9,19,23,26,27] Such lesions usually appeared as irregular masses which presented heterogeneous intense and heterogeneous enhanced pattern.[8,9,26,27] Mild-to-marked surrounding edema and mass effect can also be seen in these cases. Although the presence of inhomogeneous intense and inhomogeneous enhanced pattern were occasionally described in extra-cerebral cases,[7,14] it is not a dominant feature of primary intracranial extra-cerebral LMS.

Primary intracranial LMS should be differentiated from other brain tumors because of different clinical management. Since primary intracranial LMS may show as both extra-cerebral and intra-cerebral tumors, the main differential diagnosis for extra-cerebral lesions should include meningioma and schwannoma, and for intra-cerebral LMSs should include high-grade glioma and lymphoma. Meningioma commonly appears as benign and slow-growing tumor, and is not associated with immunodeficiency. Schwannoma usually shows cystic component and often locates in cerebellopontine angle. High-grade glioma usually demonstrates hypointense on T2WI with hemorrhage and necrosis components. Lymphoma often shows diffuse growth pattern with mild mass effect. Therefore, typical MRI features may be useful in distinguishing primary intracranial LMS from others. However, due to the rarity, it is difficult to diagnosis before the operation, frequently being misdiagnosed.

There is no established treatment regimen for primary intracranial LMS. Treatment of LMS depends on the site and extent of the tumor present and needs multidisciplinary cooperation.[25] The treatment mainly involves maximal surgical resection followed by radiotherapy or/and chemotherapy to control local recurrence. The clinical outcome of primary intracranial leiomyosarcoma is relatively poor. Limited literatures reported that most patients survived 6 months to 2 years.[14] Up to the submission of this article, our patient has survived 13 months after diagnosis.

There are some limitations in our case that should to be addressed. First, Epstein-Barr virus infection has been demonstrated in primary intracranial leiomyosarcoma.[6,8–10] However, the test for EBV was not measured in our case because we didn't expect the diagnosis of primary intracranial leiomyosarcoma before operation and the patient refused the examination after operation. Second, as there currently is no standard treatment regimen for primary intracranial LMS, the treatments including total excision and concurrent chemoradiotherapy in our case were made according to the patient's age, extent of the tumor present and pathological findings. Fortunately, to date the outcome of our patient is still good.

4. Conclusion

Primary intracranial leiomyosarcoma has some special MR imaging features. It should be considered in the differential diagnosis of intracranial tumor. Although the final diagnosis of primary intracranial leiomyosarcoma requires histopathology and immunohistochemistry analysis, MRI features may provide the first clue in identifying this rare disease and deepen our understanding.

Author contributions

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Investigation: Running Liu.
Project administration: Guohui Xu, Zhuzhong Cheng.
Writing – original draft: Xiuli Li, Jing Ren, Xiao Jiang, Guohui Xu, Peng Zhou, Zhuzhong Cheng.

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