

Case Report

Primary Intracranial Leiomyosarcoma

Babak Alijani MD¹, Shahrokh Yousefzade MD¹, Ali Aramnia MD¹, Alireza Mesbah MD²**Abstract**

Primary intracranial leiomyosarcomas are rare tumors that arise from the mesenchymal cells of the dura mater or cerebral blood vessels. Here we report the case of an extra axial leiomyosarcoma in the right parieto-occipital region of a 19-year-old male who had normal clinical and laboratory findings. Diagnostic imaging showed bony destruction, dural involvement and no parenchymal invasion. No primary site was found after metastasis work up. Specific serology tests were negative. The patient underwent a craniotomy, total tumor resection, duraplasty and skull reconstruction. He received radiotherapy and after 18 months of follow-up, no clinical and radiological signs of recurrence have been found.

Keywords: Craniotomy, leiomyosarcoma, skull

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Introduction

Primary intracranial leiomyosarcomas are the rare tumors that originated from the mesenchymal cells of the dura mater or cerebral blood vessels. There is few report of secondary metastasis to the scalp, calvarium and dura mater.

In the previous literature, primary involvement of skull is really rare. In this article we describe a case with primary involvement of skull which has been treated successfully.¹ Primary intracranial leiomyosarcoma in an HIV-infected patient.

Case Report

A 19-year-old male presented with a four month history of a parieto-occipital bump which had increased in size. He had no neurological complaints and there was no history of smoking, IV drug abuse or sexual promiscuity.

Physical examination demonstrated a palpable 5 cm firm, non-tender, mobile mass over the right parieto-occipital area. Other physical and neurological evaluations were normal as were all routine laboratory studies.

A brain CT scan demonstrated a lytic lesion which had destroyed the skull and had extracranial expansion. Brain MRI showed a right parieto-occipital well-defined extra axial heterogeneous mass, with a hypointense soft tissue signal intensity that had remarkable enhancement after Gadolinium (GD) injection. Dural enhancement was obvious and there was no parenchymal invasion noted (Figure 1).

Staging CT scan of the chest, abdomen and pelvis and radionuclide scan were negative for other sites of involvement. Viral serology was negative for HIV, HCV, HBV and EBV.

The patient underwent a wide craniotomy in the parieto-occipital region to achieve radical excision of the tumor. Scalp was

grossly tumor-free. After the craniotomy we observed a well-defined mass that had destroyed the skull and invaded the dura, with visible borders and no parenchymal involvement. The tumor was completely resected with 2 cm margins of dura and skull. Duraplasty with a pericranial patch was performed and the skull was reconstructed with titanium mesh.

The surgical specimen consisted of a piece of discoid bony tissue along with pieces of creamish-gray soft tissue that showed a cream solid cut surface with a soft consistency. Sections of malignant neoplastic tissue were composed of highly pleomorphic, mitotically active neoplastic cells with a diffuse, fascicular growth pattern that invaded the adjacent bony trabeculae. Neoplastic cells had the following immunohistochemistry profile, which was positive for vimentin, SMA, desmin, and EMA. Cells were negative for cytokeratin, LCA, CD34, NSE, and S100. Findings (Immunohistochemistry and Hematoxylin eosinophil: IHC&HE) were consistent with a diagnosis of leiomyosarcoma (Figures 2 and 3).

Following surgery, the patient underwent radiation therapy. A follow up MRI at 12 months showed no evidence of recurrent tumor and after 18 months, the patient has remained well with no observed signs and symptoms of recurrence (Figure 4).

Discussion

Sarcomas are tumors that arise from mesenchymal tissue. The nervous system has multiple tissues of mesenchymal origin and thus may serve as an origin for sarcomas. In the brain, these tumors are most often in continuity with the dura. However, because of the mesenchymal origin of blood vessels and tela choroidea, intraparenchymal sarcomas can be seen.²

In adults, leiomyosarcoma usually occurs as a result of metastatic spread from primary sites such as the gastrointestinal tract, uterus and subcutaneous tissue.³ Most reported cases of leiomyosarcoma are metastases from distant sites.⁴ Primary intracranial leiomyosarcoma represents approximately 0.1% of all intracranial tumors³ and only 14 cases of primary intracranial leiomyosarcoma have been reported in the literature.⁴

The cause of intracranial sarcomas is often unknown. Sarcoma develops in some patients after radiotherapy or chemotherapy treatment for a brain tumor.² The association of these tumors with

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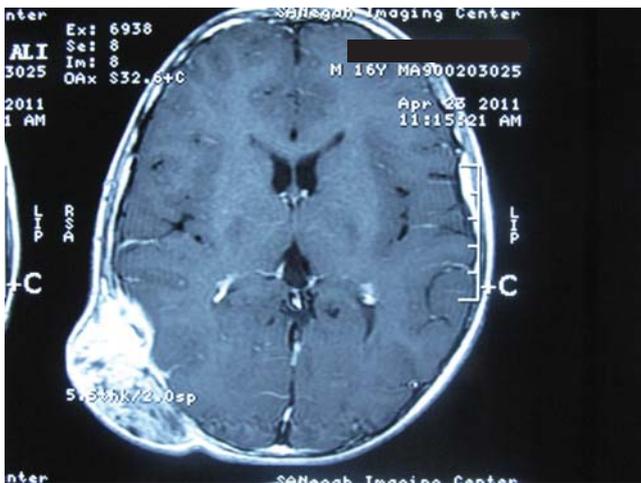


Figure 1. Axial T1 MRI after Gadolinium injection (GD) injection.

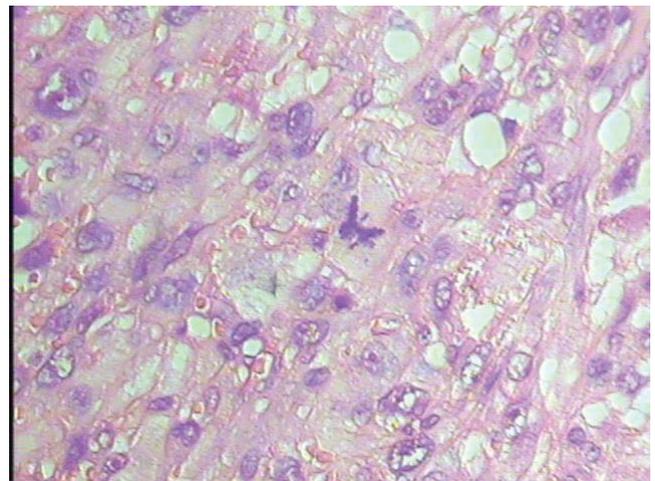


Figure 2. Oathologic feature of tumor after resection (H&E,100x)

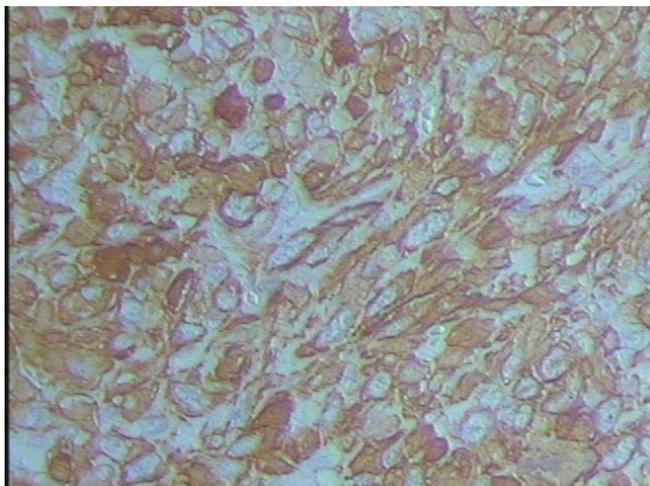


Figure 3. For definite diagnosis we have used immunohistochemistry (IHC Staining – SMA). Complete information please.

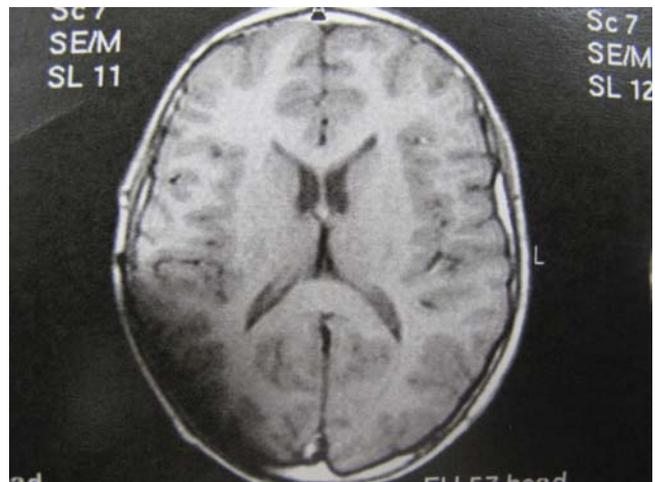


Figure 4. Post-surgical MRI at 12-month follow up.

EBV infection and AIDS is well documented in the literature.¹ In less than half of the cases of primary intracranial leiomyosarcoma reported in the literature, some forms of immune suppression have been identified. Thus, leiomyosarcoma has now been recognized as an AIDS-related tumor. It appears that co-infection with EBV may be necessary for the development of such lesions.³

However, leiomyosarcoma tends to hematogenous spread to the lung prior to the appearance of brain metastasis; and if it happens, the metastasis usually involves brain parenchyma more than skull.⁴

MRI is the main imaging study for these lesions. Occasionally angiography to define vessel involvement or CT to evaluate bony destruction may be valuable. A thorough search for a primary sarcoma outside the CNS is recommended.² The diagnosis of leiomyosarcoma is confirmed by ultrastructural features of smooth muscle cells and immunohistochemistry.⁴

The prognosis for primary intracranial leiomyosarcoma is poor⁵ Torosian et al. have observed that patients with leiomyosarcoma are more likely to develop metastases than those with other histological types of tumors.⁵ Intrauterine leiomyosarcoma has a much better prognosis than deep-seated soft-tissue leiomyosarcoma.⁶

Treatment generally involves an attempt at radical surgical excision.² Survival is probably limited by the difficulty in obtaining adequate surgical margins and an adequate radiation therapy

dose to the intracranial location. Intracranial and meningeal tumor spreads may also limit the benefits of systemic adjuvant chemotherapy. Despite these limitations, treatment should probably include aggressive multimodality therapy.¹

In this case of a primary intracranial-extra axial leiomyosarcoma with bone destruction, dural involvement and no parenchymal invasion, total surgical resection the tumor seemed to be the most important part of his treatment.

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