

## Case Report

# Primary Intracranial Leiomyosarcoma- Report of a Rare Case with Literature Review

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### Abstract:

A 29 years old man presented with a H/O headache and vomiting for 4 months, blurring of vision and gait ataxia for 2 months. A contrast MRI of brain indicated the lesion arose from the dura over left cerebellar hemisphere. A posterior fossa craniotomy was performed with a provisional diagnosis of meningioma. Histopathology and immunohistochemistry revealed Leiomyosarcoma. Further evaluation including Contrast CT and PET scan failed to identify any primary site. Primary intracranial leiomyosarcoma is very rare and only few cases were previously reported. The prognosis for primary intracranial leiomyosarcoma is poor with the longest reported survival being 32 months.

**Keywords:** Meningioma, Immunohistochemistry, Leiomyosarcoma

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### Introduction:

Most of the intracranial mesenchymal tumors are usually secondary. Majority of intracranial sarcoma arises from dura mater or cerebral blood vessel. However radiographic appearance of sarcoma can mimic meningioma. Thereafter meningioma should be always a differential diagnosis. Once diagnosis has been made, postoperative adjuvant therapy should be considered. Despite all available current treatment options, the prognosis has been poor. We report the clinical course of a patient with primary intracranial leiomyosarcoma with review of the literature.

### Case History:

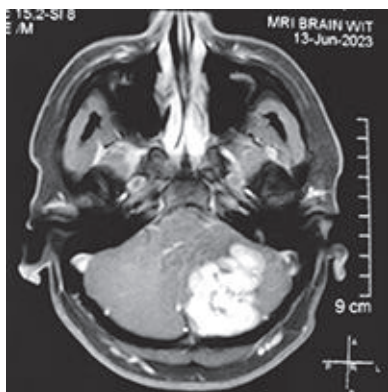
A 29 years old male patient presented with a H/O headache for 4 months which was Insidious in onset, moderate intensity, worse after waking up in the

morning and associated with vomiting. He had also H/O gradually blurring of vision and gait ataxia for 2 months. He had the tendency to fall towards left whenever he went for a walk. There was no symptoms suggestive of involvement of other sites. There was no H/O smoking, I/V drug abuse or sexual promiscuity. On physical examination papilloedema was present, cerebellar signs were positive in left side. All the baseline investigations were normal. Contrast MRI of brain demonstrated irregular shaped heterogenous enhancing mass possibly arising from dura over left cerebellar convexity displacing left cerebellar parenchyma with 4<sup>th</sup> ventricle is compressed and both temporal horns of lateral ventricle are dilated consistent with obstructive hydrocephalus (Figure: 1).

Our radiological diagnosis was meningioma.

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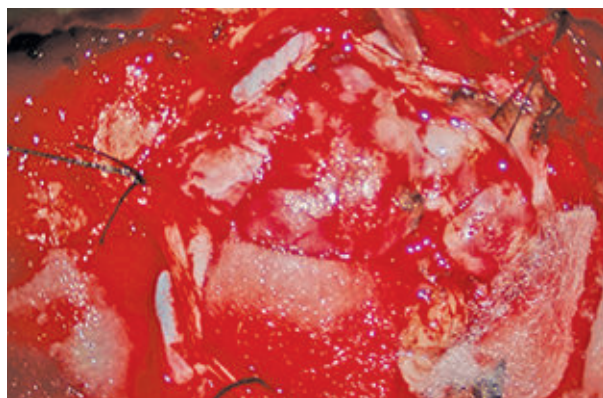


**Fig.-1:** Irregular shaped heterogenous enhancing mass (T1WI+C ,Axial)

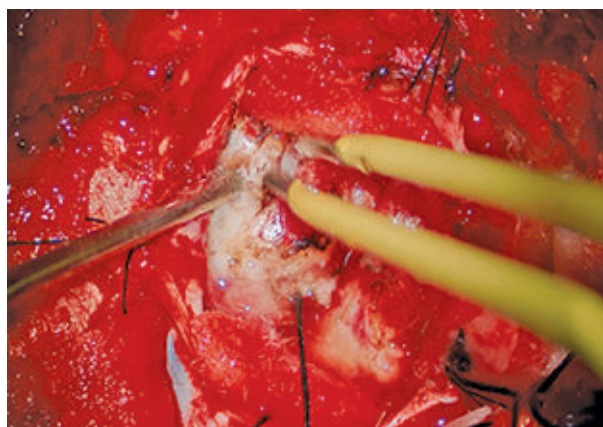
**Surgical description:**

Patient undergone ventriculoperitoneal shunt first. It was done through right Frazier's point.

Then patient was positioned prone. Tumor was approached by left paramedian suboccipital approach. Tumor was attached in dura. Firm in consistency, carefully separated from dura and brain parenchyma (Figure :2,3).



**Fig.-2:** Per-operative appearance of tumor



**Fig.-3:** Separation from dura

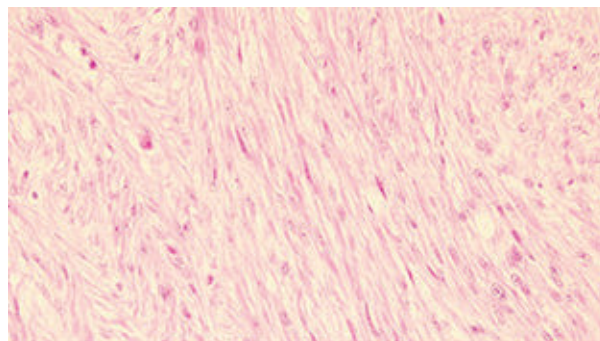
Gross total removal was done by using CUSA. Then cranioplasty was done with titanium mesh.

**Histopathologic description:**

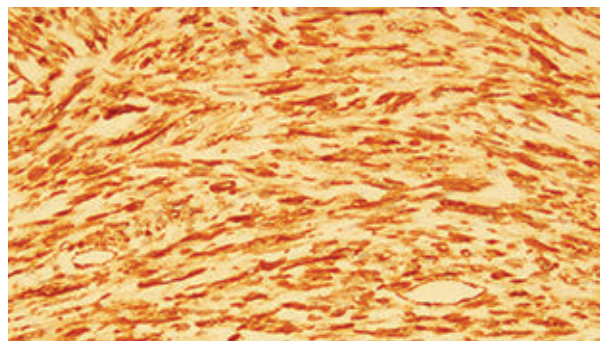
Cellular spindle cell tumor showing prominent nuclear pleomorphism. Mitosis is conspicuous. (Figure:4)

**Immunohistochemistry Analysis:**

On immunohistochemistry - negative for S-100 protein and epithelial membrane antigen and positive for smooth muscle actin, vimentin and caldesmon. (Figure:5)



**Fig.-4:** Histopathological examination revealed malignant spindle cells on H and E staining

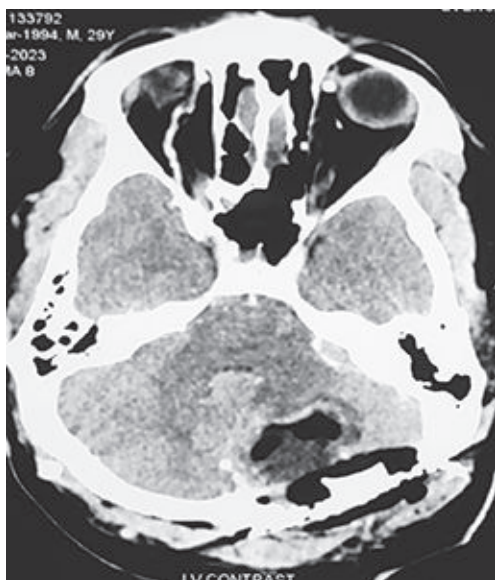


**Fig.- 5:** Immunostain positive for smooth muscle actin and vimentin

**Postoperative analysis:**

Post operative CT scan of brain revealed no residual tumor (Figure:6). The histologic examination demonstrated a malignant spindle cell neoplasm with immunostains positive for smooth muscle actin, Vimentin and Caldesmon and negative for epithelia membrane anigen. Interpretation was- Spindle cell neoplasm consistent with high grade leiomyosarcoma. In search of primary site , staging CT scan of chest, abdomen and pelvis and PET scans were negative.

The patient got post operative radiotherapy. A 1 month follow up, vision and cerebellar functions both were improved.



**Fig.- 6:** Postoperative CT scan of brain showing no residual tumor

#### Discussion:

Soft tissue sarcoma accounts only 1% of all cancers<sup>1</sup>. Majority of intracranial soft tissue sarcomas are secondary, primary intracranial sarcoma is very rare disease<sup>2</sup>. Most of intracranial sarcoma usually originate from dura mater, some from cerebral blood vessels<sup>3</sup>. The origin of this tumor is difficult to determine, but since it originates from the dura mater, it may penetrate the skull bone or invade the parenchyma within the brain<sup>3,4</sup>.

An association of the disease with immunocompromised patients has been noted, particularly in patients infected with AIDS and Epstein-Barr virus<sup>5,6</sup>.

Radiological diagnosis of leiomyosarcoma can be confusing because it looks very similar to meningioma, even preoperatively it looks like meningioma. The diagnosis has to be confirmed by histopathology and immunohistochemistry afterwards.

Our patient's tumor was negative for S-100 protein and epithelial membrane antigen and positive for smooth muscle actin, vimentin and caldesmon.

Once diagnosis of intracranial leiomyosarcoma has been made, It's mandatory to search other sites to establish the diagnosis of primary intracranial leiomyosarcoma. However no primary sites were identified in our patient with staging CT scan and PET scans.

The prognosis for primary intracranial leiomyosarcoma is poor with the longest reported survival being 32 months<sup>7,8</sup>. There are several limitations of adjuvant therapy. The limitations of adjuvant therapy and the aggressiveness of tumor are thought to be limitation of survival.

All limitation should be kept aside and multimodal treatment should be continued.

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