

## Primary Extradural Meningioma of Posterior Fossa Associated with Acquired Chiari Malformation: A Short Review

### Abstract

Generally, meningioma is considered intracranial lesion occurring in the intradural compartment. However, meningioma can also occur and usually confined in the extradural compartment called as primary extradural meningioma (PEM). PEM represents a special subgroup of meningioma constitute about 1% of all meningioma. PEM arises outside the subdural compartment and usually contains neither connection underlying subdural structures nor extends into with subdural compartment. It is commonly located in the paranasal sinus, middle ear, rarely in the intradiploic spaces of calvarial bone such as temporal, frontal, and parietal bone and orbit but extremely uncommonly in the occipital and sphenoid bones. Authors did detailed Pubmed search for posterior fossa, occipital bone extradural, ectopic intraosseous meningioma which yielded only four publications in the form of isolated case report analyzing only five case of PEM. Authors report a rare case extradural meningioma in a 40-year-old male, who presented with progressive headache and gait imbalance. Magnetic resonance imaging study of brain revealed the presence of PEM of posterior fossa associated with acquired Chiari malformation. The patient was managed successfully surgically with excision of meningioma and release of associated acquired tonsillar descent was carried out. Authors are analyzing total of five cases including four cases from published literature and one our current case. PEM of the posterior fossa tends to have equal predilection in male and female (3:3), with a mean age of 48 years (range 25–64 years). All cases were surgically and underwent gross total surgical excision. The clinical features, imagings, and management of this rare entity along with the pertinent literature are briefly discussed.

**Keywords:** Extradural, intradiploic meningioma, management

### Introduction

Meningiomas are one of the most common intracranial tumors, originating from arachnoid cap cells and most commonly located in the subdural compartment. Primary extradural meningioma (PEM) denotes a different subset of meningioma arising outside the subdural compartment as the primary tumors and without containing any connection or extension into with underlying subdural compartment or contained structures.<sup>[1-3]</sup> Primary extradural intraosseous meningioma of the calvarium is extremely rare entity.<sup>[4-7]</sup> In the past, PEM was also termed as intraosseous, diploic, cutaneous, ectopic, or extra calvarial meningioma.<sup>[2,3,7,8]</sup> In 1904, Winkler first described a meningioma originating from extradural location.<sup>[8]</sup> Till date, only four cases of PEM occurring in occipital intradiploic is reported.<sup>[4,6,7,9]</sup> [Table 1] Authors present 40-year-old male, who had primary extradural posterior fossa

meningioma, was surgically managed successfully.

### Case Report

A 40-year-old male presented to our neurosurgical services with the complaints of progressive worsening headache for 1-year associated with occasional vomiting and insidious onset progressive difficulty in walking for 3 months. He had no associated history of diminution of vision, double vision, or hearing impairment. The physical examination at admission revealed stable vitals were, he was conscious, oriented, visual acuity was 6/12 in both eyes and the presence of bilateral papilledema on fundi examination. Gross cerebellar signs were present on both side and the rest of the neurological examinations were essentially normal limit. Hematological and biochemical tests were normal range.

Noncontrast computerized tomography (CT) scan, head revealed presence of a hyperdense

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mass lesion, measuring 5.4 cm × 5.9 cm × 5 cm, located within the extradural compartment and chiefly attached to the occipital bone. The lesion was well circumscribed causing dural displacement and also producing mass effect leading to antero-medial displacement of the cerebellar hemisphere with marked compression of the fourth ventricle producing obstructive hydrocephalus [Figure 1]. Bone window CT scan revealed hyperostosis of occipital bone adjoining the mass lesion. The lesion showed homogeneously intense contrast enhancement on contrast CT scan. Magnetic resonance imaging (MRI) of brain, T1-weighted image showed presence of iso-hypointense heterogeneous well circumscribed mass lesion extradural compartment attached to the occipital bone, typically demarcated with dural outline [Figure 2], also showing heterogeneous hypointense signal on T2 weighted image with causing displacement of cerebellar hemisphere with kinking of fourth ventricle causing moderate obstructive hydrocephalus and acquired Chiari malformation. On gadolinium enhanced MRI study showed intense enhancement [Figure 3].

He was planned for surgical management. He was taken-up for microsurgical excision of mass lesion under general anesthesia. A midline suboccipital incision was given and after raising scalp flap and a midline suboccipital craniectomy along with removal of foramen magnum rim was carried out. Further, posterior arch of the C1 vertebrae was removed to make to expose access the tumor and release of tonsillar descent due to acquired Chiari. The lesion was well circumscribed and well confined in the extradural compartment and completely extra-axial. It was causing destruction of the outer cortex of the occipital bone with a small breach and also causing expansion and erosion of the inner table. Tumor was soft, suckable, and highly vascular. Gross total microsurgical excision along with involved bone was carried out. After excision of tumor, dura was opened and subpial resections of herniated tonsils were carried out and lax duraplasty also carried out utilizing the pericranium graft.

Postoperative course was uneventful. He had complete remission of headache with improvement in the gait. Postoperative CT scans head showed complete excision of meningioma with subsidence of obstructive hydrocephalus with opening of fourth ventricle [Figure 4]. The histopathology was suggestive of meningotheial meningioma, who Grade 1, MIB labeling index was 2%. At the last follow-up at 6 months following surgical treatment, he was doing well with complete relief of headache and minimal gait disturbance still persisting.

## Discussion

The meningioma is considered as primary intradural lesion, whereas PEM variety represents uncommon subgroup of meningioma, which can originate in the paranasal sinus,

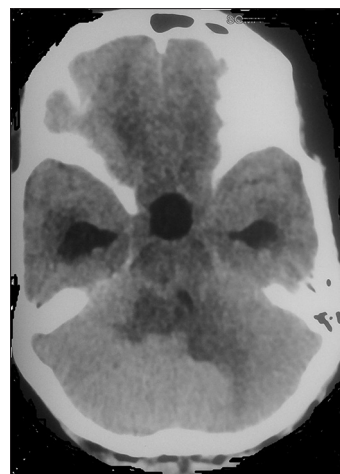


Figure 1: Computed tomogram, head showing hyperdense lesion in the occipital bone region and causing hyperostosis of internal occipital protuberance

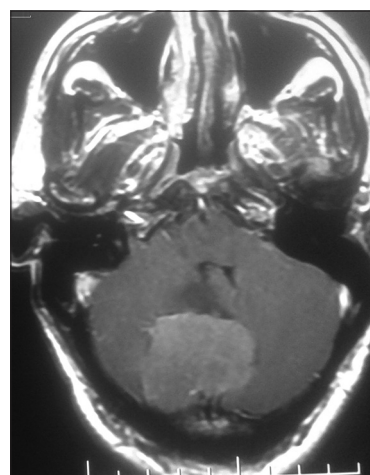


Figure 2: Magnetic resonance imaging brain, axial section T1-weighted image, showing presence of extra-durally located meningioma causing mass effect on cerebellar hemisphere with kinking of fourth ventricle

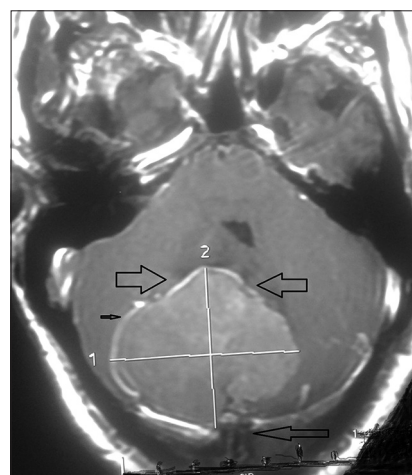


Figure 3: Contrast enhanced magnetic resonance imaging brain, axial section image showing typically displacement of dura around mass lesion (marked with arrow head) and causing mass distortion of fourth ventricle

orbit, neck, salivary gland, calvaria, and along perineural sheath of cranial nerves.<sup>[1,2,7,9-13]</sup>

PEM is postulated to arise from arachnoid cell rests captured at inappropriate sites during embryonic developmental phase of fetal life or rarely acquired implantation. According to one postulate entrapment of arachnoid cells within the bone is responsible for PEM occurrence.<sup>[2]</sup> Other acquired postulates are abnormal cranial moldings during child birth and acquired cranial trauma. Even the arachnoid cells distributed along the course of blood vessels and cranial nerves passage while traversing through the skull-base foramen can also cause entrapment of the meningocytes or arachnoid cells in the skull-base bone.<sup>[2,8,11]</sup>

Crawford *et al.* put forward a diagnostic criteria for characterization of intraosseous meningioma as the histopathology of the lesion should be consistent with the meningioma, the lesion should be located in intraosseous or extradural compartment and the adjoining dural layer of meninges, arachnoid membrane and brain parenchyma should not be involved or infiltrated.<sup>[1]</sup>

PEM can be further sub-classified according to Lang and colleagues into three types; first group comprising purely extracalvarial compartment localized PEM, second type is complete localization within the calvarium and the final third type representing calvarial meningioma associated with extracalvarial extension.<sup>[11]</sup> Our case represented third type of intradiploic meningioma.



Figure 4: Postoperative computed tomogram of head showing complete excision of meningioma

Liu *et al.* observed PEM of head tends is more common in male 1:0.8–1.4, whereas for intradural meningioma is 1:2 intradural.<sup>[3]</sup> However posterior fossa meningioma has equal incidence in both male and females was 1:1.<sup>[3]</sup>

PEM predominantly occur after sixth decade, however, mean age of posterior fossa meningioma occurrence is 48 years, range being 25–64 years.<sup>[3]</sup> Clinically, patient present with nonspecific feature of raised intracranial pressure or feature of obstructive hydrocephalus. Our case had headache with gross cerebellar features.<sup>[3,10,11]</sup>

X-ray can delineate the location of PEM and pressure effect produced by mass on the adjoining bone.<sup>[2,3,5-9,11]</sup> It shows generally osteoplastic or hyperostosis reaction in the form of speckled or granular calcification. The osteolytic variety is rare, causing expansion and thinning of the outer or dinner tables of the skull, however mixed types also have been reported.

CT scan shows gross outline of lesion, bony changes produced by mass lesion and on contrast administration, it may show enhancement. MRI scan of brain is modality of choice, showing better soft tissue delineation, extension of meningioma, encasement of vessel and nerves and also extent and size of extension into adjacent foramina, intradural spread, hydrocephalus, associated acquired Chiari malformation.<sup>[2,5]</sup>

Surgery is usually considered as standard modality of treatment. In the present case, we achieved complete microsurgical excision of the lesion with removal of involved bone with excellent neurological recovery and also subsidence of obstructive hydrocephalus.<sup>[2,9,11]</sup>

Yamazaki *et al.* reported a rare intraosseous meningioma associated with intradural extension; neuro-imaging revealed an extradural mass lesion located in the right posterior fossa and a small daughter lesion extending inside the dura. Intraoperatively, the lesion was found to be lying just beneath the thinned outer table within the extradural space, whereas the daughter lesion transgressing the dura. However, there was no attachment to the dura was noted. Histopathology was compatible with meningotheliomatous meningioma.<sup>[4]</sup>

Hayhurst *et al.* reported a PEM in a 25-year-old male, located in the occipital bone. He presented with a painless scalp nodule in the occipital region, CT scan revealed bony defect

Table 1: Summary of previously published posterior fossa extradural meningioma

Authors/references	Publication year	Age (year)/sex	Location	Type	Surgical impression	Outcome
Yuge <i>et al.</i> <sup>[7]</sup>	1991	64/female	Posterior fossa	IIB	Total excision	Good
Changhong <i>et al.</i> <sup>[9]</sup>	1997	54/female	Occipital	IIB	Excision	-
Changhong <i>et al.</i> <sup>[9]</sup>	1997	42/female	Occipital	IIB	Excision	-
Yamazaki <i>et al.</i> <sup>[4]</sup>	2001	62/male	Posterior fossa	IIB	Gross total excision	Good at 4 months follow-up
Hayhurst <i>et al.</i> <sup>[6]</sup>	2004	25/male	Occipital	IIB	Gross total excision	Good at 1 year
Current case	2015	40/male	Occipital	IIIB	Gross total excision	Good

in the occipital bone, located at the level of torcula with soft tissue lesion communicating with dura through bony defect. The histopathology of the resected specimen was suggestive of meningioma. The patient had no recurrence till the last follow-up at 1-year after the surgery.<sup>[6]</sup>

Yuge *et al.* reported a – 64-year-old female with posterior fossa intraosseous meningioma, who presented with headache, CT scan head revealed homogeneously enhancing mass. During surgery, the tumor was partially attached to dura. She underwent successfully surgical excision; histopathology was fibroblastic meningioma.<sup>[7]</sup>

Changhong *et al.* reviewed the angiographic and CT scan findings in ten cases of primary intraosseous meningioma and two cases out of them had primary posterior fossa PEM.<sup>[9]</sup>

Authors reported a 40-year-old male, who had transitional meningioma in the intradiploic space of orbital roof and underwent surgical excision with reconstruction of orbital roof and he had no recurrence or exophthalmos during follow-up period.<sup>[10]</sup> Authors reported another case of PEM, located in the he orbital roof in a 16-year-old girl, who presented with progressive proptosis, with normal vision and she was managed successfully surgically.<sup>[13]</sup> Goyal *et al.* reported a case of parasagittal meningioma in which, the overlying skull was extremely hyperostotic and appeared like a mountain.<sup>[14]</sup>

## Conclusion

PEMs are extremely uncommon lesion and usually do not cause dural infiltration, so complete surgical excision is preferred surgical option. Current study reminds the possibility of such meningioma should also be considered by neurosurgeon while dealing with calvarial based lesion causing atypical skull changes evident in radiology.

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## Conflicts of interest

There are no conflicts of interest.

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