

# Primary Intraosseous Meningioma: Bifrontal Skull Mass

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**Abstract:** Primary intraosseous meningioma is a rare tumor arising commonly from the calvarial bone. Here, we report a case of primary intraosseous meningioma which presented as a Bifrontal skull mass in a 30-year-old woman who presented with a progressive forehead enlargement and headache of 3 years duration. Brain CT shows hyperostosis of the frontal bone and microscopy shows intertrabecular syncytial proliferation of oval to spindle cells at the area forming whorl. Even though it is rare, PIM is the most common subtype of extradural meningioma.

**Keywords:** bifrontal, PIM, calvarium, meningioma

## Introduction

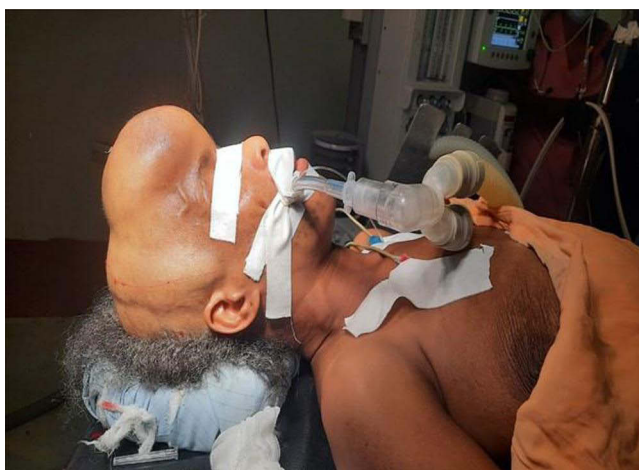
Meningioma is classified into three: primary neuraxial meningioma the commonest type arising within neuraxis, primary extra neuraxial meningioma arising outside of neuraxis, and secondary meningioma which metastasized from elsewhere.<sup>17</sup> A primary intraosseous meningioma is a form of primary extradural meningioma that arises from the bone. It represents the most common form of primary extradural meningioma though it constitutes around 2% of meningiomas overall. Tumors originating from the calvaria usually present with swelling, nonspecific pain, and loss of vision depending on the site of their origin.<sup>2-4</sup> Due to the diverse radiologic features, distinguishing them from their mimickers usually requires additional histopathologic workup.<sup>3-5</sup> The lack of a clear and standardized definition made primary intraosseous meningioma a topic of confusion. According to Arana et al, it is best defined as a meningioma with the largest component contained within the Calvarium with no or only minimal dural involvement.<sup>6</sup> Till the year of 2015, only 100 primary intraosseous meningioma cases were reported.<sup>7</sup>

## Case Report

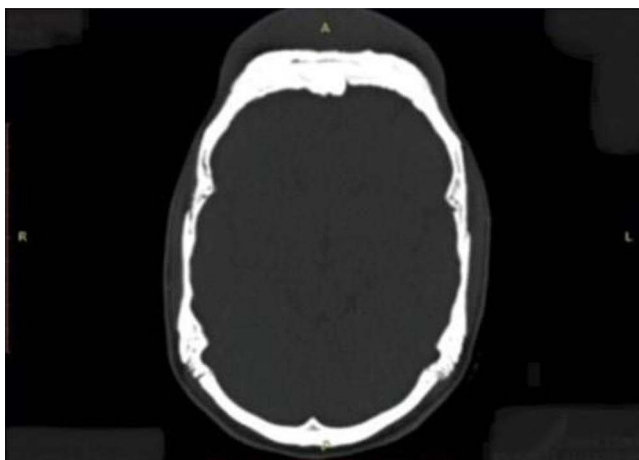
A 30-year-old female patient presented with a global throbbing headache and progressive forehead swelling of 4 years duration. She has a heavy sensation upon opening her both eyes. She has no history of head trauma and neurologic symptoms. On physical examinations, she has a 15x15cm hard, non-tender, hard Bifrontal mass with grossly intact cranial nerves (Figure 1).

CT scan shows hyperostosis of the frontal bone with a well-defined outline measuring 2.2 cm at its maximum thickness. The diploic space is obliterated and replaced by a less dense matrix (ground glass matrix). Both the inner and outer tables of the frontal bone are involved, and the lesion extended to almost all frontal bone being thick at the center and decreasing bilaterally in a symmetric fashion. Overlying forehead soft tissue thickness is also increased paralleling the bone lesion (Figures 2-5).

Complete excision of the tumor with a clear surgical margin and cranioplasty was done. The specimen was sent for histopathologic evaluation.



**Figure 1** 15x15cm hard bifrontal skull mass.



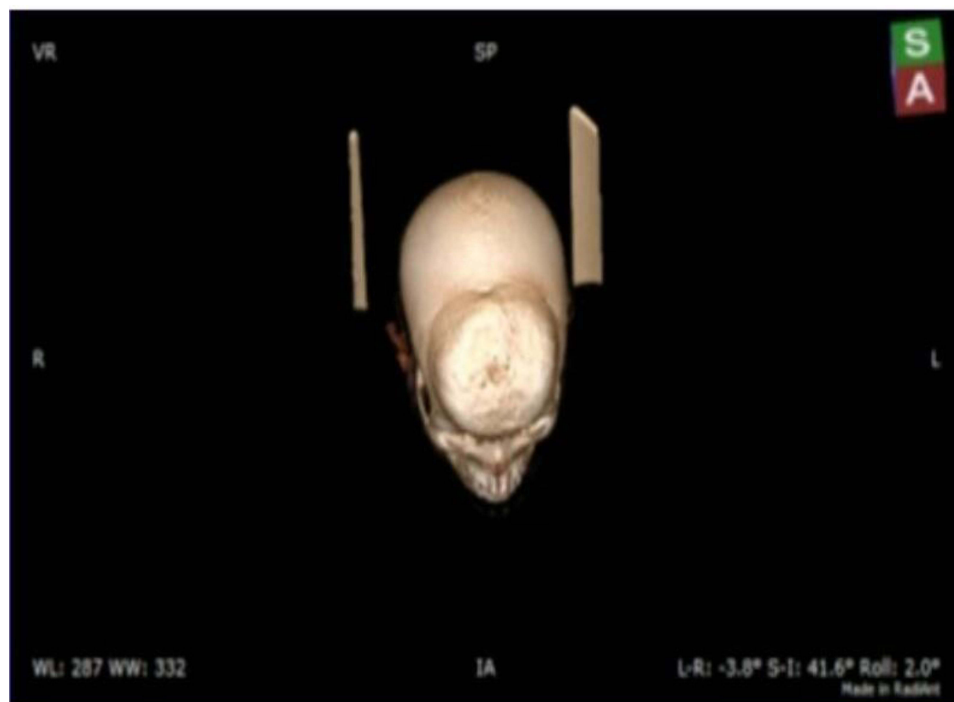
**Figure 2** CT scan (the bone window) shows hyperostosis of the frontal bone with a well-defined outline measuring 2.2 cm at its maximum thickness.



**Figure 3** CT scan of the head showing the thickening of the overlying forehead soft tissue and increased paralleling bone lesions.



**Figure 4** Left lateral view of the CT scan of the head with 3D reconstruction showing the bifrontal mass.

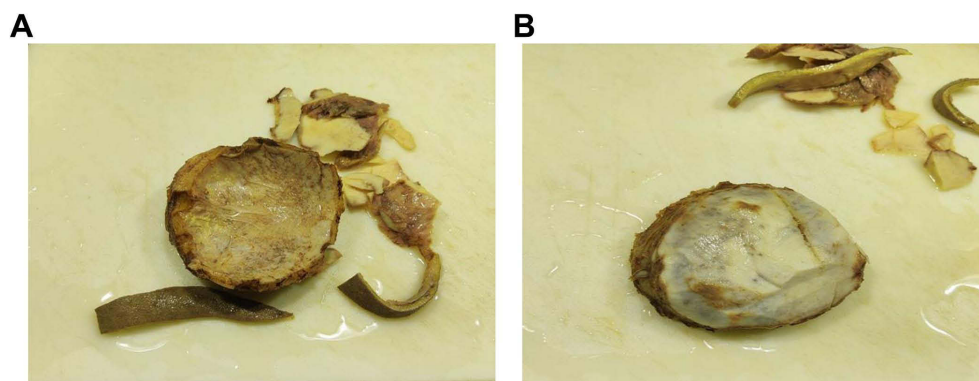


**Figure 5** Superior view of the CT scan of the head with 3D reconstruction showing the bifrontal mass.

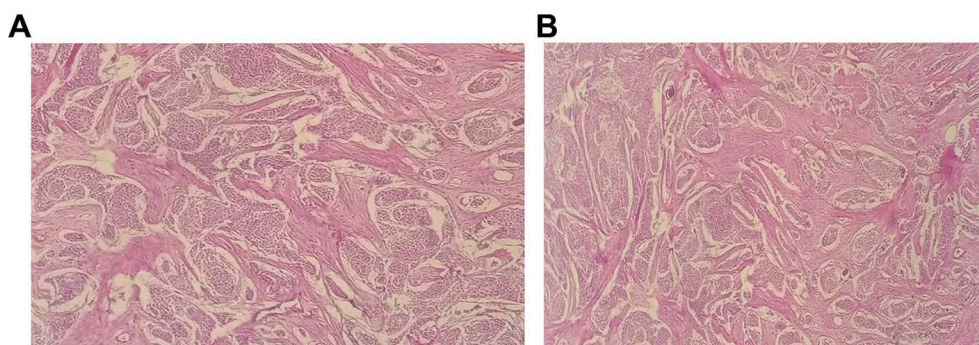
Grossly, 13 x 12cm slightly irregular Bifrontal calvarial bone with 1.5–3 cm thickness and two separate irregular skin tissues larger measuring 13 x 8cm, smaller measuring 10x2 cm were submitted (Figure 6A and B).

Histopathologic evaluation of fixed and decalcified tissue showed thickened lamellar and woven bone cuffing whorl and syncytium of oval to spindle meningotheial cells confined to the dura and bony components with no soft tissue invasion or extension (Figures 7 and 8).

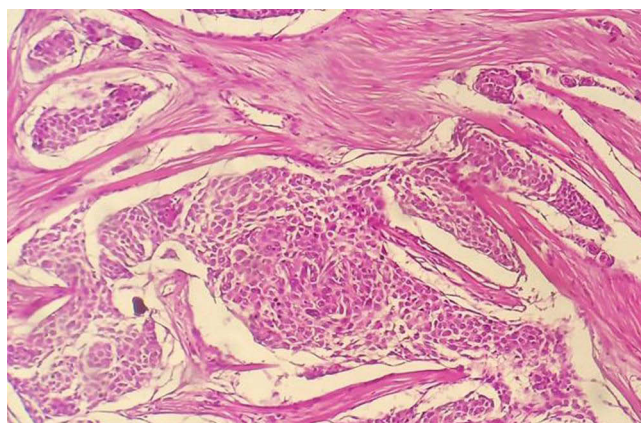
Immunohistochemistry also shows nuclear pattern positivity for progesterone (Figure 9A and B).



**Figure 6 (A and B)** Gross pictures showing, a 13 x 12cm slightly irregular Bifrontal calvarial bone with 1.5–3 cm thickness and two separate irregular skin tissues larger measuring 13 x 8cm, smaller measuring 10x2 cm.



**Figure 7 (A and B)** Microscopic picture on 10/HPF showing thickened lamellar and woven bone cuffing whorl and syncytium of oval to spindle meningeal cells.

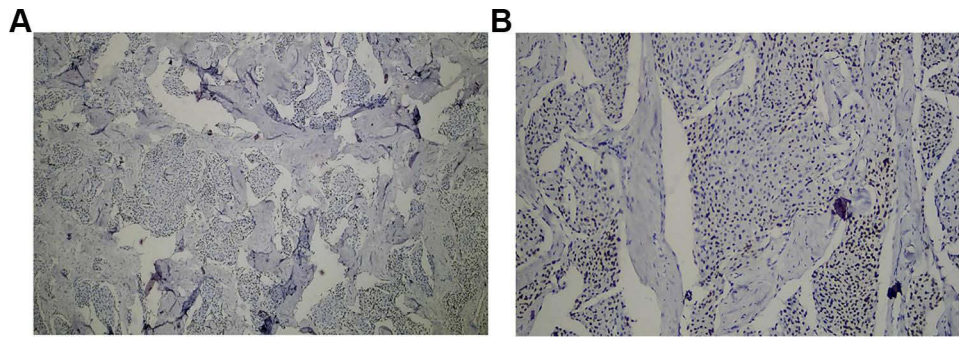


**Figure 8** Microscopic picture on 40/HPF showing whorls and syncytium of oval to spindle meningeal cells.

## Discussion

Meningioma is the most common benign primary intracranial tumor comprising a fifth of all tumors of the same category.<sup>1</sup> They are slowly growing Dura-based tumors and typically they originate from arachnoid cap cells with a high chance of cure.<sup>19</sup> Sometimes it can also arise from the scalp, orbit, paranasal sinus, neck, and skin.<sup>8</sup> Ectopic or extradural primary meningioma accounts for about 2% of all meningiomas.<sup>9</sup> Calvaria is involved in 68% of primary extradural meningioma with frontotemporal and orbital regions being the most involved.<sup>10,11</sup> It has bimodal peak incidence with one peak in the second decade and a second peak during the fifth through seventh decades of life with slight female





**Figure 9 (A and B)** Immunostaining for progesterone shows a nuclear pattern of staining.

preponderance. The tumors are nearly uniformly solitary, although a small number of case reports have presented patients with two separate lesions.

Primary intraosseous meningioma may exhibit osteoblastic, osteolytic, or mixed patterns of growth with the osteoblastic pattern being the most common accounting for around 58% of all cases.<sup>11–13</sup> Based on the location of proliferation, primary extradural tumors are classified as: purely extra calvarial (Type I), purely calvarial tumors (Type II), and calvarial tumors with extra calvarial extension (Type III). Type II and III are further sub-classified as C or B subtypes based on their anatomical location.<sup>10,14</sup>

Histopathologically, there are 15 WHO-recognized morphologic variants though more emphasis is given to the tumor grade regardless of the subtype. There are 3 grades according to the WHO with WHO grade 1 being under the benign category and comprising 2/3rd of the subtypes, and WHO grade 2 and grade 3 are characterized by recurrence and potential for metastasis, respectively.<sup>18</sup>

Like conventional meningioma, PIOM can have any of the histologic subtypes and is positive for EMA, PR, SSTR2a, and vimentin. In one study, progesterone is found to be positive in 87% of low-grade meningiomas as in our case.<sup>9,22</sup>

Management options include complete excision with a wide free margin with subsequent cranioplasty or decompression of tumor near vital structures that is difficult to resect, and adjuvant chemotherapy or radiation can also be given in incomplete resection. In our case, complete excision with a wide free margin with subsequent cranioplasty was done. The patient is stable, and no problem was reported in the last 4 months.

Prognosis is generally good for completely excised lesions with free margin whereas, atypical primary intraosseous meningioma carries around 21% risk of recurrence.

Osteoma, osteoblastoma, fibrous dysplasia, and Paget disease of the bone can radiographically mimic PIOM with osteoblastic growth pattern, whereas, plasma cell tumors, Langerhans cell histiocytosis, lymphomas, and metastatic skull tumors can mimic osteolytic counterparts.<sup>15,16</sup> Detailed clinical evaluation, imaging, and histopathologic evaluation are the mainstay method to distinguish one from the other. Moreover, new instrumental exams and tools should be investigated to possibly improve the detection and management of PIOMs.<sup>20,21</sup>

## Conclusion

Primary intraosseous meningioma is a rare tumor, and it should be taken into consideration in a patient with calvarial mass. Imaging and histopathology are the main means of definitive diagnosis.

## Data Sharing Statement

The data used to support the findings of this study will be available from the corresponding author upon reasonable request.

## Ethics Approval and Consent to Participate

Informed consent was obtained from the patient for the publication of this case report and accompanying images. The study is also approved by the institutional board review of Hawassa University College of medicine and health science.

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## Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis, and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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## Disclosure

All authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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