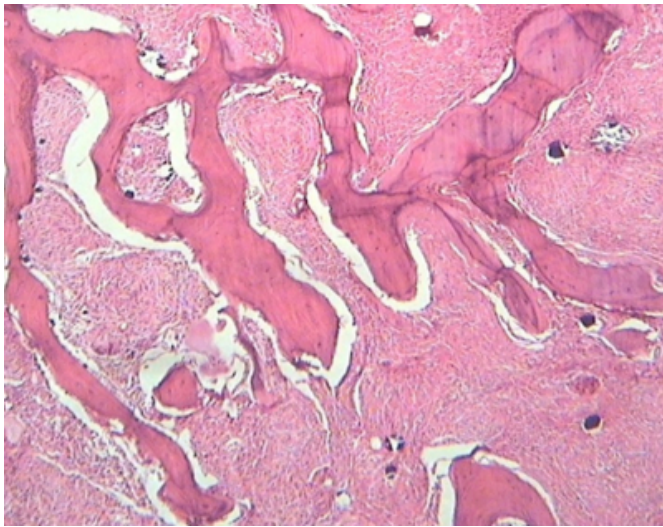




permeating in between the existing bony trabeculae of calvarial bone (Fig 2). Occasional cells showed intranuclear pseudoinclusions. There was no mitotic activity or foci of necrosis. There were large number of psammoma bodies. (Fig 3).

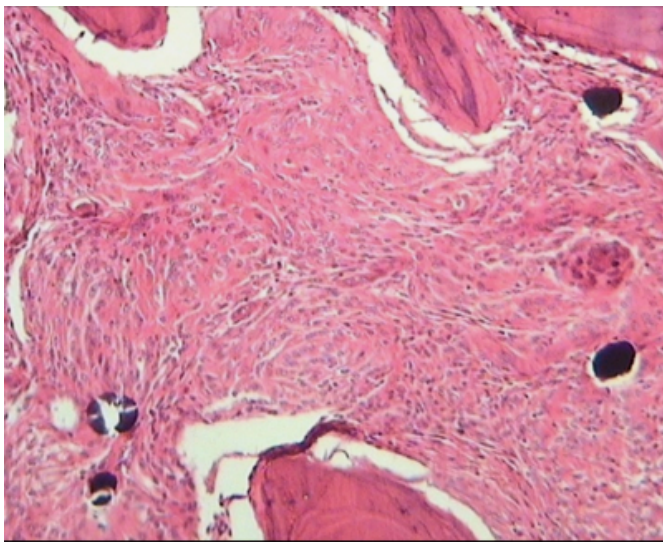
### Figure 2

Figure 2: Photomicrograph of calvarial meningioma permeating in between the bony trabeculae. (H&E, x20)



### Figure 3

Figure 3: Higher magnification showing Psammoma bodies



## DISCUSSION

Extracranial meningiomas are rare; and account for 1%–2% of all meningiomas<sup>12</sup>. The meningiomas arising in locations outside the dural compartment have been called ectopic, extradural (epidural), calvarial, cutaneous, extracranial, extraneuraxial, or intraosseous meningiomas. To avoid the confusion in nomenclature, Lang et al<sup>3</sup> has proposed a

single term, “primary extradural meningioma” for such lesions. This term highlights the origin of these tumors as being separate from the dural coverings of any part of the brain or spinal cord and further differentiates these meningiomas from “primary intradural meningiomas,” which may have secondary extracranial extensions and/or may have metastasized<sup>3</sup>. Although some authors have emphasized that this group of meningiomas should have no connection to the dura mater or any other intracranial structures<sup>45</sup>, other reports include tumors with intracranial growth<sup>678</sup>

Primary extradural meningiomas are classified as purely extracalvarial (type I), purely calvarial (type II), or calvarial with extracalvarial extension (type III). According to the site of location of the tumor, Lang et al<sup>(3)</sup> further subdivided type II and type III lesions into convexity (C) or skull base (B) forms.

Many different hypotheses exist regarding the origin of primary extradural and calvarial meningioma. They are thought to arise from ectopic meningiocytes or arachnoid cap cells trapped in the cranial sutures during moulding of the head at birth.<sup>9</sup> Misplacement and entrapment of meningotheelial cells into suture or fracture lines as a result of trauma has also been speculated as the probable cause of calvarial meningioma.<sup>10</sup> Involvement of multiple sutures is also reported.<sup>11</sup> However, only 8% of the calvarial meningiomas are in relationship with a cranial suture. Cutaneous meningiomas could be congenital in origin where they can arise from arachnoid cell rests located in the skin as a result of defective closure of the neural tube wherein the meningeal tissue is ‘pinched’ off into the surface.<sup>12</sup> They are also thought to arise from multipotent mesenchymal cells as a reaction to an unidentified stimulus.<sup>13</sup> This may be relevant in our case since the meningioma was diffuse and involved the frontoparietal bone widely and extended into the overlying soft tissue. It had no special connection within the cranial sutures or the foramina of the cranial nerves.

According to the literature, 68% of the primary extradural meningiomas involved the calvaria.<sup>3</sup> Frontoparietal and orbital regions are the most common locations for intraosseous meningiomas. They occur with approximately the same frequency in each sex<sup>3</sup>. Though primary extradural meningiomas occur predominantly during later decades of life, they also have a second peak incidence in younger patients (especially during the second decade). Patients with calvarial intraosseous meningiomas typically present with

slow-growing scalp swelling that may or may not be painful. They do not show any neurologic symptoms or signs, unless the lesion extends through the inner table and compresses intracranial structures. These lesions may be asymptomatic and detected incidentally.<sup>13</sup> However Calvarial meningiomas are known to be associated with intracranial hypertension.<sup>11</sup> The marked dural thickening overlying and adjacent to the tumour as well as the hyperostotic bone is attributed to cause intracranial hypertension. Dural sinus occlusion can also be an important cause of the raised intracranial tension.

The incidence of hyperostosis with meningioma has been reported to be about 4.5%.<sup>14</sup> Since Brissaud and Lerebaulet,<sup>15</sup> described this association in 1903, several theories have been postulated regarding its cause. The bony thickening is associated with the presence, in the medullary spaces, of clumps of tumour cells.<sup>16</sup> The cause, management and prognosis of bony hyperostosis remains controversial

Biologically, calvarial meningiomas have been observed to be benign and slow-growing. On the other hand, calvarial meningiomas are more prone to develop malignant changes (11%) compared with intracranial meningiomas (2%)<sup>(3,13)</sup>. Meningiomas presenting with scalp swelling, osteolytic skull lesions and extracranial soft tissue masses are more aggressive in nature.<sup>17</sup>

Surgical resection is the treatment of choice. Although radiotherapy is advocated, it is usually not recommended unless there is evidence of rapid progression of the disease. In cases of diffuse involvement of the calvaria, a wide surgical resection is advisable whenever possible followed by a cranial reconstruction. Radiation therapy is advocated for rapidly progressive disease.

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