Falcine Meningioma with Massive Calvarial Hyperostosis

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Abstract
Meningiomas are neoplasms of meningeal origin. Hyperostosis is a peculiar but common sign of meningoima. Though common in skull base meningiomas, massive calvarial hyperostosis is rare in the vertex, with only few reported cases. We report a rare case of massive calvarial hyperostosis due to falcine meningioma, illustrated with multimodality imaging.

Keywords: CT Scan, Hyperostosis, Meningioma, MRI, X-ray

Introduction
Meningiomas represent most common intracranial extraaxial neoplasms. Hyperostosis is a common associated sign of meningioma, most commonly associated with meningioma en plaque and primary intraosseus meningioma. Overall incidence of hyperostosis in meningioma is about 4-5%; but it might be as high as 13-49% in en plaque variety.¹ The cause of hyperostosis in meningioma is not clear; some authors believe it a manifestation of osseous tumor invasion while others believe it a reactionary change.² Imaging plays an important role in the diagnosis of primary neoplasm as well as the reactionary changes. Massive hyperostosis is a challenge during surgery, for the accepted treatment is resection of hyperostotic bone resulting in a large bony defect. It is therefore important to delineate the exact extent of hyperostosis with preoperative imaging. We report the radiological perspectives of a case of massive calvarial hyperostosis due to underlying meningioma; first of its kind in Nepal.

Case Report
A 45 years old female patient was referred for imaging of the swelling over the skull vault of several years duration. There was associated pain over the swelling for last one and half years. On examination, there was bony hard swelling over the vertex extending into both parietal regions. The overlying soft tissues of scalp were intact. There were no focal neurological deficits.

Plain X-ray of the skull (Figure 1) showed dense hyperostosis of the vertex involving both parietal bones. Both the inner and outer tables of skull were involved with extension of the hyperostosis beyond the outer table. Rest of the vault and base of the skull appeared normal apart from the prominent vascularmarkings.

Figure 1: X-ray skull AP and lateral views showing massive hyperostosis in the vertex.

Plain CT scan of the head (Figure 2) showed an isodense extraaxial falcine mass with biparasagittal extension. Marked thickening of the vault was noted overlying the mass. The maximum vertical thickness was 3.3 cm; width was 9.1 cm and anteroposterior extent was 8.8 cm. No calcifications were noted within the mass. Mild vasogenic edema was noted in the adjacent parietal lobes.
Infarction were noted however. MRI revealed a falxine extraaxial mass centred in the midline high parietal convexity. The mass showed biparasagittal extension, with slightly larger tumor volume in the right. Relative isointensity to cortex was noted in T1 and T2 weighted images (Figure 3). Intense homogeneous enhancement was noted after IV gadolinium. Thickening of the adjacent dura with “dural tail” was also noted. The posterior aspect of the superior sagittal sinus was completely encased by the falxine mass. The calvarial hyperostosis appeared as large area of thickened vault with low signal intensity. MR venography showed non-visualization of the posterosuperior sagittal sinus consistent with the tumor encasement (Figure 4). No parenchymal signs of venous infarction were noted however.

Figure 2: CT Scan of head (clockwise); axial brain window, coronal, axial and sagittal bone windows, respectively. Brain window shows isodense falxine mass with biparasagittal extension. Marked thickening of the parietal bones is noted in bone windows.

Figure 4: Clockwise; axial T1 contrast, sagittal T1 contrast, MIP image from TOF MR venography and coronal T1 contrast MR images. Intense enhancement of the falxine mass is noted with biparasagittal extension and calvarial hyperostosis. The posterior half of the superior sagittal sinus is not seen in MIP images implying tumor encasement.

All of the imaging features were consistent with falxine meningioma with massive calvarial hyperostosis.

The tumor and overlying bone were subsequently resected. Histologic features were consistent with meningothelial meningioma (Figure 5).

Figure 3: Axial T1 and T2 weighted MR images showing relatively isointense falxine mass with biparasagittal extension. Note the massive overlying hyperostosis seen as areas of profound hypointensity.

Figure 5: Photomicrographs of the resected specimen (magnified 10 and 20 times, respectively) show tumor consisting of neoplastic meningothelial cells arranged in whorls and synctitial pattern surrounded by thin collagenous septa. Tumor cells show minimal pleomorphism and have ill-defined cell border with oval uniform small nuclei. Mitotic figures, nuclear atypia and necrosis are not seen.
Meningioma is one of the common intracranial tumors, averaging about 15% of all symptomatic and 1/3rd of all incidental intracranial neoplasms. They generally are benign neoplasms (>80% are WHO grade I tumors), however, atypical meningiomas classified as WHO grade II constitute 5 – 15%, and anaplastic or malignant meningiomas classified as WHO grade III constitute 1-3% of meningiomas. They are extra-axial tumors arising from the dura. Most common locations are cerebral convexity, parasagittal and sphenoid wing regions.

Although plain X-ray is useful in demonstrating the degree of calvarial hyperostosis, it gives little information about the ongoing intracranial pathological process.

CT scan is widely available, rapid and can be used in patients with MRI contraindications (metallic implants, pacemakers). It is the best modality for evaluating the intratumoral psammomatosus calcifications and overlying calvarial changes (hyperostosis, destruction, pneumosinus dilatans). Typical meningiomas appear isodense (or slightly hyperdense) to brain parenchyma and show avid enhancement after IV contrast. The degree of bony hyperostosis is accurately identified and the density of the thickened bone can be quantitatively measured, providing the information about relative “hardness” of the bone.

MRI, however, remains the imaging modality of choice due to its high soft tissue contrast. Most meningiomas appear as extra-axial mass with signal similar to brain cortex on spin echo T1 and T2 weighted images, marked and homogeneous enhancement after IV contrast, with or without enhancing dural tail reflecting neoplastic dural infiltration or reactive vascularity or both. Areas of calcifications or intratumoral vessels both appear as intratumoral signal voids. The hyperostosis of overlying bone also appears as thickened area of vault with signal loss. MRI venography/hyperinvasively delineates major intracranial venous sinuses and their relation to the tumor. Association of hyperostosis with meningioma is well known, seen in about 4-5% of all meningiomas and upto 60% on primary intracranial meningiomas. They are also more commonly associated with en plaque types of meningioma.

Only few cases of massive calvarial hyperostosis have been reported. The thickening appeared to be diffuse in these reported cases. Our case showed massive but localized calvarial thickening.

The cause of hyperostosis in meningioma remains controversial. Postulated hypotheses include reactionary changes in response to irritation of the bone by the tumor, production of bone by tumor itself, direct tumoral invasion of the bone etc. But most accepted cause is the tumor invasion of the adjacent bone. Pieper et al showed that hyperostosis when present in association with meningiomas, represents a direct tumor invasion of the bone. Though tumor cells are noted in overlying bone on histopathology, it is still not clear that tumor invasion is the cause or the result of bony changes.

The importance of hyperostosis in treatment of meningioma lies in increased incidence of tumor recurrence. Studies have described the relation of clinical success of meningioma surgery to the extent of resection. So, infiltrated bone should also be removed during the surgery of meningioma in order to prevent recurrence. Preoperative imaging delineates the exact extent of bony hyperostosis and tumoral extent.

References


