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A Case of Intracranial Meningioma with Contiguous Extracranial Extension

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Abstract: Intracranial WHO grade 1 meningioma with contiguous extra cranial extension is a rare occurrence, with very few reported case studies on the same. Here we present an incidentally noted intracranial meningioma with extra cranial extension in a 67 years old female suffering from left sided hemiplegia. Unlike brain parenchymal invasion, which signifies poor prognosis; patients with contiguous extra cranial extension have good prognosis. **Keywords:** Grade 1 Meningioma, Intracranial meningioma, Extracranial meningioma, Extracranial extension.

INTRODUCTION

Meningiomas are benign central nervous system neoplasms arising from the meningothelial cells. The incidence of meningioma in India is not well documented. However, meningioma constitutes about 30% of all the primary intracranial neoplasms[1, 2]. Most meningiomas are well circumscribed and curable by surgery alone[3]. Although uncommon, atypical and malignant meningiomas constitute 5.8% to 10% of all cases of meningiomas. These cases require multimodal therapy to prevent recurrence [3, 4]. Various histologic characteristics are used to define the aggressiveness of meningiomas. Brain or spinal cord parenchymal invasion signifies poor prognosis in meningiomas despite a benign histology. But, bone and soft tissue infiltration are seen in benign meningiomas and do not change the grade of the neoplasm[5]. Invasion of skull or vertebral bone by an intra-cranial meningioma is common and is associated with hyperostosis of bone matrix, well appreciated on radiologic evaluation[6]. However, extra cranial extension of intracranial meningiomas is not common, with only few reported case studies on the same[7-9]. In the present article we describe a case of WHO grade 1 meningioma with both intracranial and extra cranial component.

CASE REPORT

A 67 years old female patient presented to the casualty with right hemiplegia and aphasia of acute onset. There was no history of vomiting or seizures at the time of presentation. No history of similar complaints in the past was provided. At the time of presentation she was conscious and co-operative. On local examination a firm, non-tender swelling was noted on the left temple, measuring 4 x 3cms in size and fixed to the underlying skull and muscle. The skin over the

swelling was healthy. Examination of the cranial nerves and fundus were within normal limits. All other general and systemic examination was within normal limits. An MRI scan of the head was done which showed a sub acute white matter infarct in the parietal region of left hemisphere. Also seen in the right hemisphere is a space occupying intracranial extra-axial lesion buckling the right parieto-temporal brain parenchyma, expanding the intra-diploic region and extending into extra cranial soft tissue, suggestive of a malignant lesion (Figure 1).

Based on the clinical and radiological findings, patient was advised to undergo right frontotemporoparietal craniotomy and excision of tumor along with the involved bone. During surgery the tumor was easily separable from the brain parenchyma, infiltrating through the temporal bone and involving the temporalis muscle. The tumor along with the involved bone and soft tissue were excised and submitted for histopathological evaluation.

On gross examination, a grey white granular lobulated lesion was noted attached to the dura and infiltrating through the underlying bone, and into the soft tissue on the external surface of the bone (Figure 2 & 3). Multiple blocks were taken from the mass, embedded in paraffin and the subsequent sections obtained were stained with haematoxylin and eosin. On microscopic examination the tumor was composed of neoplastic syncytial cell nests with focal whorling pattern consistent with meningiothelial meningioma (Figure 4). Numerous bizarre cells with pleomorphic vesicular nuclei and focal hyper-cellular cell nests were also seen. However, mitotic activity was sparse (<4 mitosis/10 HPF). The meningiothelial cell nests were noted infiltrating through the cortex and into the marrow spaces (Figure 5). Sections from the attached skeletal muscle showed tumor infiltrating into the muscle.

Postoperatively, the patient was managed conservatively for the sub acute left parietal infarct.

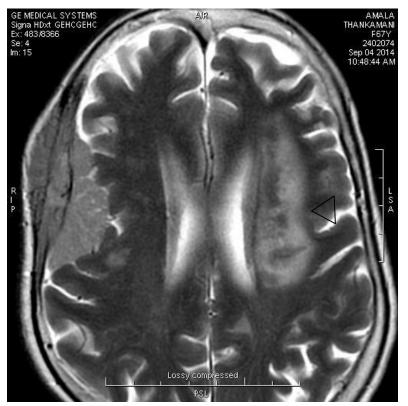


Fig-1: MRI SCAN - In the right hemisphere a space occupying intracranial extra-axial lesion buckling the right parieto-temporal brain parenchyma, expanding the intra-diploic region and extending into extra cranial soft tissue (*arrow*). In the left hemisphere a sub acute white matter infarct noted in the parietal region (*arrow-head*).



Fig-2: GROSS APPEARANCE - A lobulated mass (arrow) attached to the dura (arrowhead) noted.

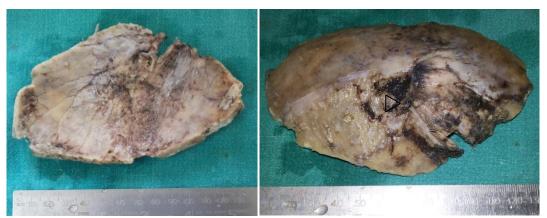


Fig-3: TEMPORAL BONE - The portion of temporal bone above the tumor was removed. The tumor was noted infiltrating through the bone, from the inner surface (*arrow*); into the attached portion of temporalis muscle on the outer surface (*arrow-head*).

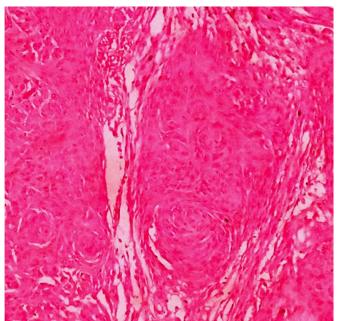


Fig-4: Tumor composed of neoplastic syncytial cell nests with focal whorling pattern consistent with meningiothelial meningioma (H & E, 40x).

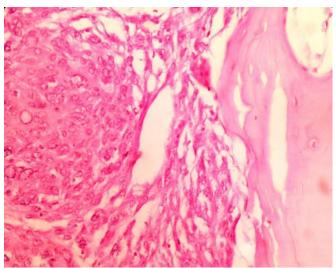


Fig-5: Tumor nests infiltrating into the temporal bone and occupying the marrow spaces noted (H & E, 40x).

DISCUSSION

Meningiomas accounts for the most number of non-glial intracranial neoplasms. Even though meningiomas are slow growing lesions, they are capable of wide spread destruction and extra cranial extension[10, 11]. Approximately 6 to 17% of meningiomas are extra cranial in location[12]. Of these the predominant forms are extra cranial extension of primary intracranial meningiomas (secondary extra cranial meningiomas)[13, 14]. Various mechanisms are postulated for this extension of intracranial meningiomas. Anatomically the preferred pathways are: supraorbital fissure (extension into the orbit), cribriform plate (extension into the nasal cavities and nasopharvnx), floor of middle cranial fossa (extension into paranasal sinuses and pterygoid region), through the suture line of the skull and trans-diploic extension[15]. In the above discussed case report, the meningioma was not in close proximity to any suture line or other natural defects. Besides there was evidence of trans-diploic extension in the form of tumor deposits noted within the marrow spaces of the parietal bone. Most secondary extra cranial meningiomas tend to recapitulate the histologic appearance of their primary intracranial form. The most common histologic types noted are syncytial and transitional type, with syncytial pattern being seen in the present case. The predominant form noted in paranasal sinuses are meningothelial type [10, 11, 16]. When a secondary extra cranial extension arises in an intracranial meningioma, the intervening bone usually shows an osteolytic change [13, 14]. Uncommonly, meningiomas with extra cranial extension may induce an osteoblastic reaction in the intervening bone [17]. However, in the present case the intervening bone showed neither osteolysis nor osteoblastic change.

Clinically most intracranial meningiomas are silent, predominantly presenting with insidious symptoms[18]. If symptomatic, the most common symptom is new-onset, slowly evolving headache due to raised intracranial pressure. The headache may or may not be associated with other symptoms such as confusion, paresis or seizures[18]. Physical examination most commonly reveals various grades of paresis or neural deficits[18]. In this case, the clinical presentation in the patient was due to the left hemisphere sub acute infarct. The meningioma, though visible grossly due to an extra cranial component, was asymptomatic clinically.

Though plain X-ray can be highly informative in meningiomas, the preferred brain imaging is MR imaging or CT scan. Of the three imaging modalities, MR imaging is most informative in differentiating glial lesions from meningioma. CT scan, however, best reveals the bone remodeling seen in meningiomas. Recently, algorithms for effective identification and characterization of meningiomas in MR imaging is being used routinely. MR spectroscopy and PET scan is

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being used increasingly for patients unable to undergo surgery and inoperable cases, respectively[18].

Complete excision is best treatment advised, wherever possible. It has been associated with significantly better long term outcomes when compared to subtotal excision. Even in extra cranial meningiomas, complete surgical extirpation is usually curative. Similar to intracranial meningiomas, chances of recurrence is influenced by the histologic type of the tumor, failure of early diagnosis, inadequate resection of involved bony structures and meningioma en-plaque tumor extension[19]. To aid in effective prediction of recurrence and management of patients Simpson et al have advised a grading system based on degree of surgical excision[20].

CONCLUSION

Intracranial meningiomas, though commonly associated with reactive changes in the overlying bone, are uncommonly associated with bone infiltration and extra cranial extension. Secondary bone changes are often present in such cases of contiguous extension. In this case, however, the interspersed temporal bone showed no reactive change. Identification of extra cranial meningiomas warrants careful clinical and radiologic evaluation of patient to rule out intracranial meningiomas.

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