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Article

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Rare case of multiple meningiomas in non-neurofibromatosis patient at unusual locations

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Abstract: Multiple meningioma is a condition in which more than one intracranial lesion is seen in different location and these lesions may occur with or without signs of neurofibromatosis. Incidence of multiple meningioma range from 1 to 10% in different series. We report a case of multiple meningioma in a 33 years old female who had 14 intracranial lesions both supratentorially and infratentorially, and underwent surgery for large right lateral intraventricular meningioma. She had two meningiomas located in posterior fossa associated with supratentorial meningioma, which has been rarely reported.

Key words: meningioma, Non-Neurofibromatosis, supratentorially

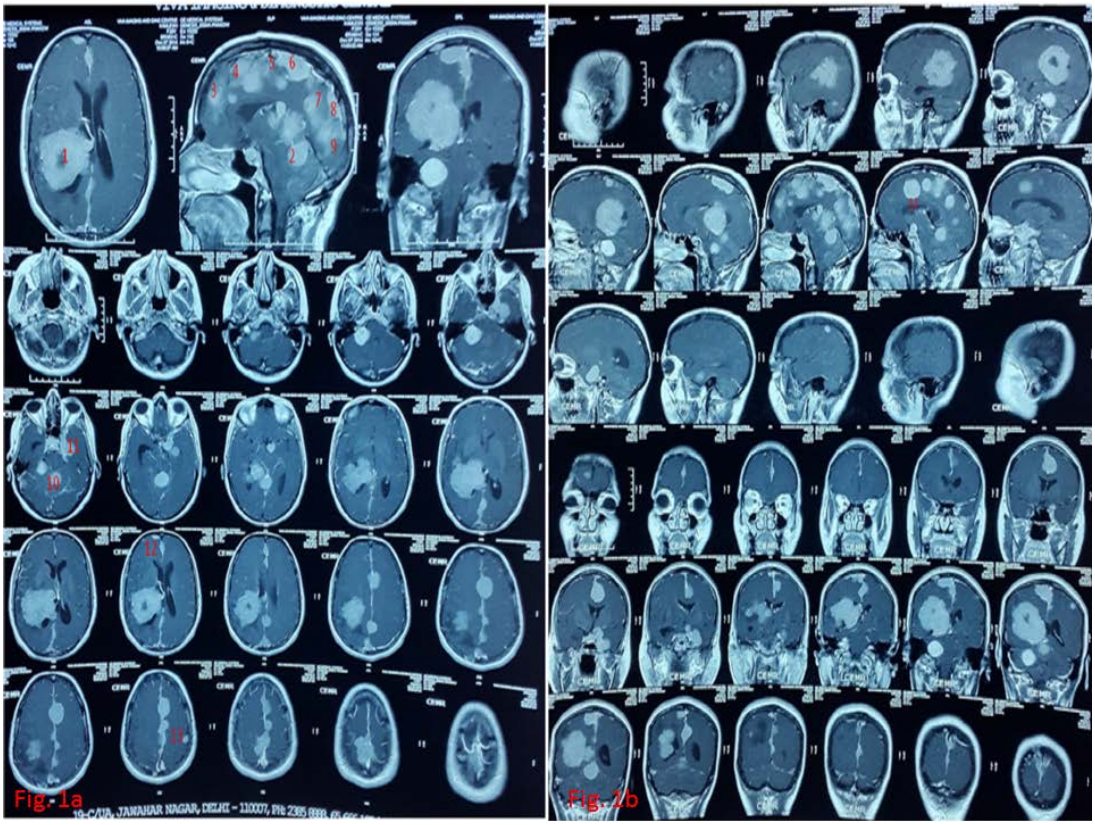
Introduction

The incidence of multiple meningioma is 1 to 2% of all meningioma cases in Cushing and Eisenhardt series.(1) Inspite of multiple intracranial lesions prognosis remains almost same as solitary lesion and in one-third of cases there is occurence of different grades of malignancy simultaneously. The purpose of this case report presentation is to elicit the varied manifestation of multiple meningioma along with its management.

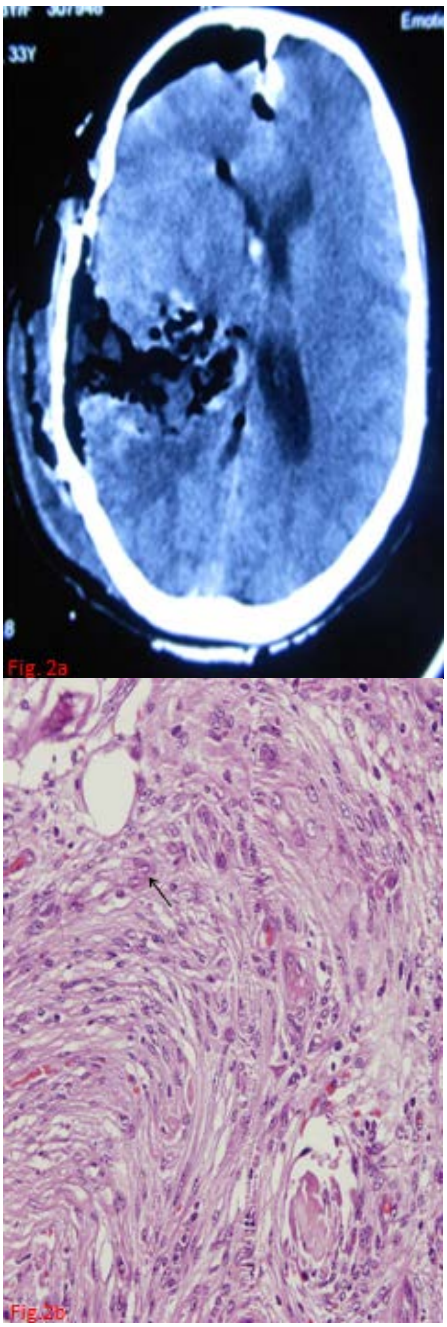
Case report

A 33 years old female patient presented with to us with worsening headache and vomiting of 7 days duration, gradually progressive hearing loss in both ears for past 8 years and difficulty in balance and gait for past 2 years. On examination, patient had features suggestive of raised intracranial pressure with papilledema on bilateral fundus examination. Patient had no signs of neurofibromatosis. She had bilateral sensory-neural hearing loss and ataxia. Contrast enhanced CT scan and MRI

revealed fourteen contrast enhancing intracranial lesions supratentorially and infratentorially, largest located in Right lateral ventricle (Figures 1a & 1b). The Screening of Spine was negative for any tumor. Chromosome analysis revealed normal female karyotype and no numerical or structural anomalies detected at 450-550 banding resolution. Patient underwent right temporo-parietal craniotomy and excision of right lateral ventricle tumor (Simpson grade II) (Figure 2a). Histological examination was suggestive of Grade I transitional meningioma (Figure 2b). Post-operative period was uneventful and patient was followed up for a period of 1 year, however patient refused further treatment and interventions.



Figures 1a and 1b - MRI Scan showing location of meningiomas at various location with the largest being Right Intraventricular in location (1) and two infratentorial lesions (2 & 10)



Figures 2a and 2b - Post Op CT demonstrating excision of Rt Intraventricular Tumor and histopathology showing features suggestive of transitional meningioma (WHO grade 1)

Discussion

Meningiomas arise from arachnoid cap cells and account for 13-19% of all intracranial neoplasms. (2,3) Although most of the meningiomas are benign, 6% are atypical and 1-2% are frankly malignant.(2)Most meningiomas occur sporadically and are of unknown etiology. Genes associated with meningiomas are NF2 on chromosome 22 and DAC-1 on chromosome 18. There is higher frequency of meningiomas in women and progesterone receptors found in 80% of meningiomas. Multiple meningiomas are defined as atleast two spatially separated meningiomas occurring at same time, or more than two meningiomas arising sequentially from 2 clearly distinct regions. (4)

The term 'Multiple meningiomas' was first described by Cushing and Eisenhardt to denote the occurence of multiple tumors in absence of neurofibromatosis or acoustic neuromas. (1)

Pathogenesis of multiple meningiomas has two distinct hypothesis. The first hypothesis suggest tumor arise independently in different location and the second hypothesis suggest that a single transforming event occurs and the original clone of cells spread throughout the meninges in formation of multiple clonally related tumors. (2, 3)

In Locatelli et al series of 10 cases, a total of 227 intracranial meningiomas were reported from 1977 to 1984 and all 10 patients were female.(5) Galabert-Gonzdez et al(6) reported 13 cases of multiple mengiomas between 1983 and 2003 and none of the patient had manifestations of neurofibromatosis.

After the advert of CT, Domericulli et al (2) detected multiple meningioma in 4-5 % of

cases. The most common location of all was supratentorial convexity and parasagittal falx. Multiple meningiomas occurring in posterior fossa are very rare. (7) In our case review we had two tumors located in posterior fossa one at CP angle and the other one in the central tentorium. About 80-90% of multiple meningiomas are benign and are WHO grade I. (8) Tomita et al, described fibrous and anaplastic type of histology in multiple meningiomas. (9) However, in one-third of cases, simultaneous occurence of different grades of malignancy in the lesion is observed.

Surgery is the treatment of choice for multiple meningiomas and prognosis does not differ from solitary benign meningiomas. Surgical decision is based on the following characteristics. Symptomatic meningioma, asymptomatic meningioma greater than 3 cm in size and surgically accessible and symptomatic expanding tumor. (10, 11) Our patient had one tumor which was removed surgically, whereas the other lesion was left to be followed up at frequent interval. The one in CP angle is planned subsequently in second admission.

Conclusion

Multiple meningiomas are rare and are seen more commonly in females. This condition is more commonly associated with neurofibromatosis. Psammomatous, fibroblastic, menigothelial and transitional types are the most common histological subtypes. Surgery is the treatment of choice for symptomatic lesions. Prognosis of multiple meningioma is good and may be same as for solitary meningiomas.

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