Case Report

A YOUNG FEMALE PATIENT WITH NEUROFIBROMATOSIS TYPE II: RARE CONDITION HAVING SHORT HISTORY WITH HUGE PARASAGITTAL MENINGIOMA

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ABSTRAT

Neurofibromatosis is a rare neoplastic disease with which we come across. In addition to that a very large Parasagittal Meningioma was encountered which has invaded across the anterior & middle superior sagittal sinus. It may present with spinal tumors. The case of Neurofibromatosis type II we present in a young female patient who had a history of loss of vision, head ache, decrease hearing from left side and single episode of fits for the last few months. She was managed initially conservatively then surgical intervention was contemplated in two stages. In first stage the hyperostotic bone flap was removed and placed in abdomen. Then while removing tumor in the next stage the anterior one third of the superior sinus was tied and separated from the falx for attempting the complete removal.

Key words: Neurofibromatosis Type II, Huge Parasagittal Meningioma, Simpson Grade I.

INTRODUCTION

Meningiomas show a rising incidence with age. It is assumed that meningiomas are responsible for about 15% of all male intracranial tumors and 30% of the female ones. As a rule the tumors are reported to be 1.5 to 3 times more frequent in women. Majority of the patients who suffer from Neurofibromatosis type 2 develop meningioma1,2,3. NF II is an inheritable disorder with an autosomal dominant mode of transmission. Incidence of the disease is about 1 in 60,000.4 Inherited Schwannomas, Meningiomas, and Ependymomas") is an inherited disease. The main manifestation of the disease is the development of symmetric, non-malignant brain tumors in the region of the cranial nerve VIII, which is the "auditory-vestibular nerve" that transmits sensory information from the inner ear to the brain. Most people with this condition also experience visual problems. NF II is caused by mutations of the "Merlin" gene,5 of which seems, to influence the form and movement of cells. The principal treatments consist of neurosurgical removal of the tumors. The underlying disorder does not have any therapy due to the cell function caused by the genetic mutation. In the literature review, the largest of these is the 404-gm. meningioma (also reported as 430 gm.) which was removed by Cushing6 at the fourth operation on a patient. Cushing reported a similar meningioma weighing 400 gm. removed at the eighth operation on a patient who had a total of 1,350 gm. of tumor tissue removed in a series of 11 operations over a period of 12 years. This tumor was reported in Cushing's monograph "Intracranial Tumors"'7 as weighing 1,475 gm. Cushing also reported a removal of a 341-gm. glioma' and a 341-gm. Round cell sarcoma with operative deaths. Davidoff 8 reported an 835-gm. meningioma but it was composed largely of hyperostosis of the skull with relatively little intracranial tumor. Later it has been published that the part of tumor was excised weighed 1380gm, and is the second largest and the heaviest meningioma that has been successfully excised.9 The idea of reporting this case is to highlight the significance of invasion and the attachment with superior sagittal sinus is still the most challenging part of the surgical removal. The unusual gaint Meningioma is

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itself the reportable aspect, in addition to this it was associated with Neurofibroma type 2 which is one of the rare diseases.

CASE REPORT

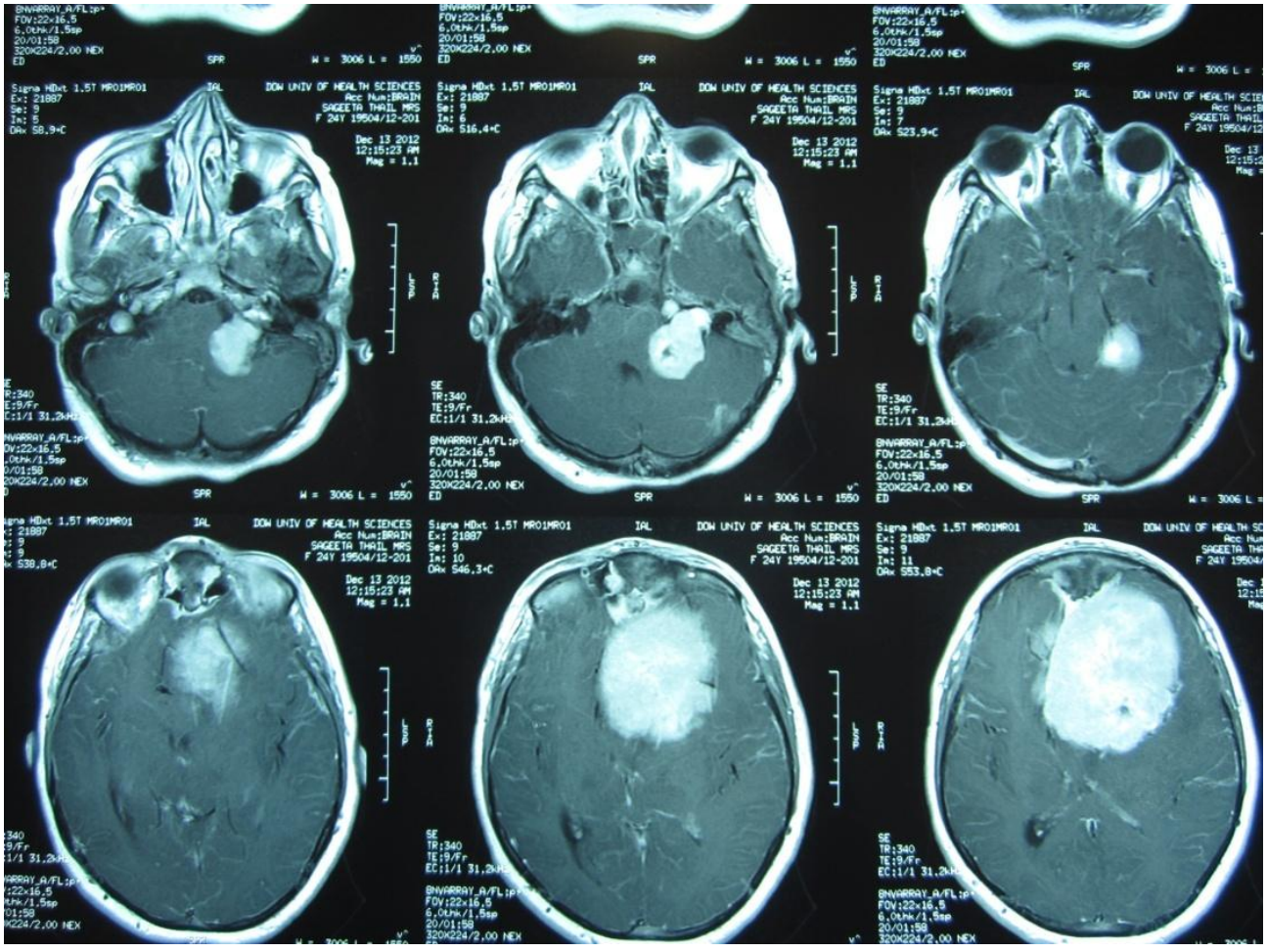
Twenty two years old married female presented with progressive loss of vision for the last one year and headache for four months. According to the patient she was alright one year back then she noticed diminution of vision. It started from the left eye, first in the outer zone then inner zone was also involved. Two months later, she became unable to see from her left eye. Furthermore she started complaining of headache for the last four months which was progressive in nature, increasing in intensity with time, more in frontal region and throbbing in character not associated with particular time partially relieved by analgesics. She also started decreased vision of right eye too. Over a period of six months she lost her vision completely. On enquiry she added that she has developed difficulty in hearing from her left ear for the last twenty days which was not noticed even. There was no significant personal history and neither a positive history for constitutional symptoms.

After consultation with Opthalmologist and Neurophysician, who performed some workup and prescribed some medicines. Finally she was referred to Neurosurgery Department Civil Hospital Karachi Dow University of Health Sciences for further workup and management. There was no history of Diabetes Mellitus, asthma, jaundice and surgery. On examination a young women of average height and built lying comfortably in bed, conscious and oriented. She was found to be stable vitally at the time of admission. There was no other abnormality found on the systemic review and head to toe examination. While on neurological examination her higher mental functions were normal and the speech was also normal. But she could not walk with out support. On cranial nerve examination her sense of smell was intact but visual acuity was consistent with the finding of no light perception. Pupils are dilated bilaterally and not reactive to light. Fundoscopic finding was bilateral optic discs atrophy. Rest of the examination was completely normal including motor and sensory system. There was no positive cerebellar sign and the signs of meningeal irritation were also negative. The routine investigation including blood biochemistry and X-ray revealed no abnormality. While performing the first stage of surgery the modified bicoronal scalp incision was given and bone flap removal was planed in such a manner that approximately more 2cm maximum margins of the tumor are exposed across the midline. During the procedure profuse bleeding from bone while performing craniotomy leads the procedure to convert in two stages. Patient lost the approximate 1500cc blood hence the total removal was planed for the next stage. The surgery was uneventful patient smoothly recovered from the anesthesia and post operatively she was managed in high dependency unit. Couple of day's later next stage of surgery was performed and the tumor was removed en bloc from all sides. And the middle part was detached from the falx after tying the superior sagittal sinus. Moreover most of anterior part of the sinus was already invaded and occluded by lesion. During surgery eight pints of blood were transfused with fresh frozen plasma. Again the patient recovered well and managed in the Intensive care unit for the initial 48 hours and remained better for further one week. During this post operative period she developed cerebrospinal fluid leak on third postoperative day. Pharmacologic agents were used and later on Epidural drain was placed. Next week when the problem did not resolve the subgalial drain was also placed. Unfortunately her clinical condition deteriorated and she could not survive despite taking all measures. The macroscopic pathological finding of the specimen showed size as 13x11x7cm and weighted approx 1500gm. While the histopathology revealed WHO Grade II meningioma labeled as clear cell meningioma comprising of moderately cellular and highly vascular tumor.

DISCUSSION

Meningioma with unusual and large size has been described in literature. Meningiomas are rather frequently encountered in neurosurgical practice. These tumors arise from the arachnoid cells and account for 10-20% of brain tumors seen in a general neurosurgical oncology population and may rarely occur extradurally or even extracranially [10]. It's predominance in females is already documented. Meningioma variably express hormone receptors for progesterone, androgen, estrogen and placental growth

factors [11,12,13] as well as exogenous hormones, [14] and their response to increased serum progesterone levels during the pregnancy may account for the accelerated growth. This explains the sudden presentation as a neurosurgical emergency in some circumstances.[15,16] Similarly there is a possibility of genetic factor relation with Neurofibrmatosis type 2 in large meningiomas progression but to confirm this assumption some larger studies would be required. We thus have to treat such patients early provided the patients could have arrived earlier in our setup. In the literature the FIG massive bleeding from diploic veins while performing craniotomy in large meningioma surgery is well documented. It has also been mentioned that in the earlier period when the resources and expertise were limited these patients with such a large meningiomas or lesions used to die on operating table secondary to excessive bleeding. With advances in terms of preoperative planning and expert team work such events have been reduced but still haemostasis is challenging for such huge lesions among neurosurgeons. Meningioma is usually considered as a slow growing tumor but different meningiomas behave in atypical manner. Therefore they present very late and at times sudden presentations are dealt as neurosurgical emergency. However not all meningiomas are equal and the data from the literature indicates that the clear cell meningioma encountered in our patients is exceedingly rare [17,18,19]. Patient recovered well in both stages of surgery but was unfortunate due to cerebrospinal fluid leakage, meningitis and later on sepsis, all these factors were found to be the reasons of mortality in this patient.



I: PREOPERATIVE MRI SHOWING BOTH LESIONS.

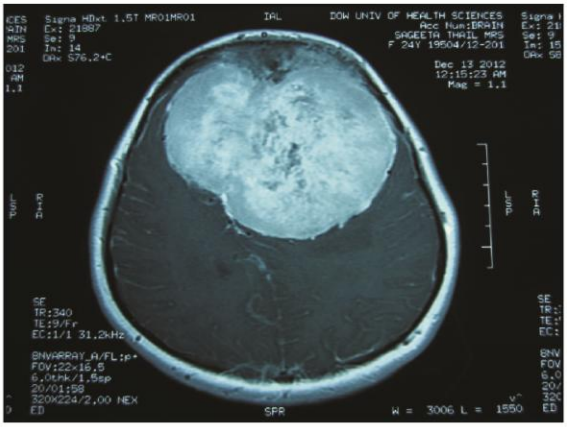
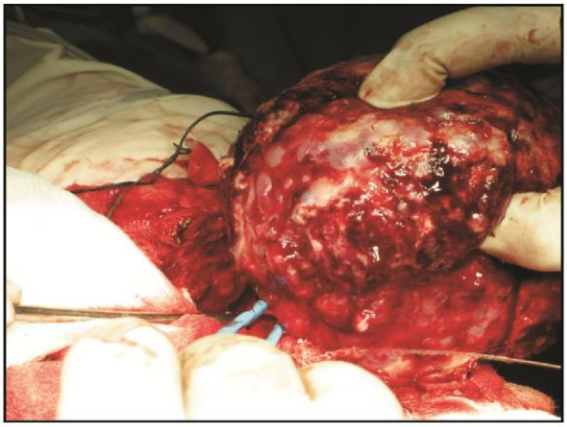
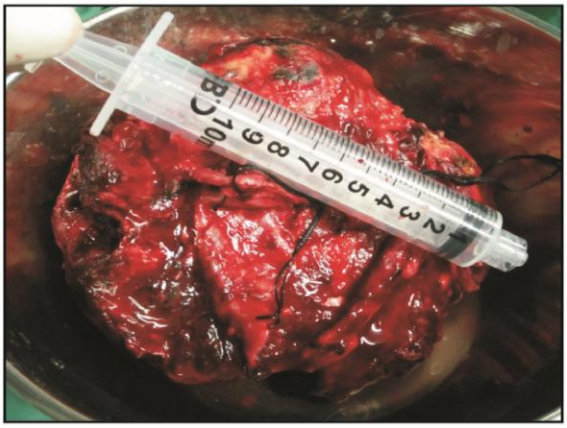


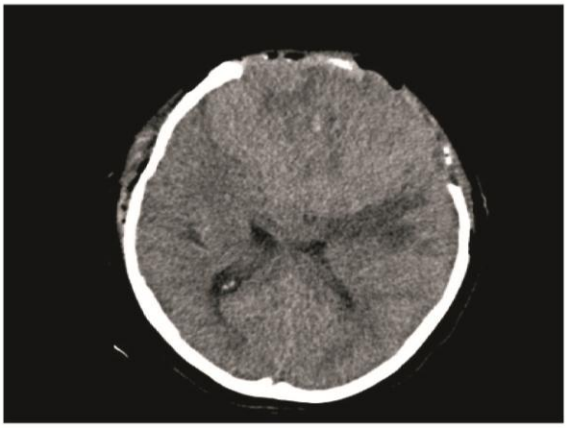
FIG II: PREOPERATIVE MRI SHOWING PARASGITTAL MENINGIOMA



FIGIII: PEROPERATIVE LAST STEP OF EN BLOC REMOVAL



FIGIV: POSTOPERATIVE LSEION SIZE 13 BY 13CM



FIGV: FIRST STAGE POSOPERATIVE CT SCAN

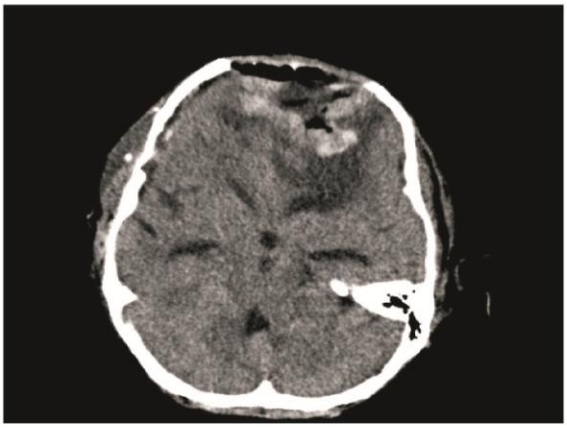


FIG VI: SECOND STAGE POSTOPERATIVE CT SCAN

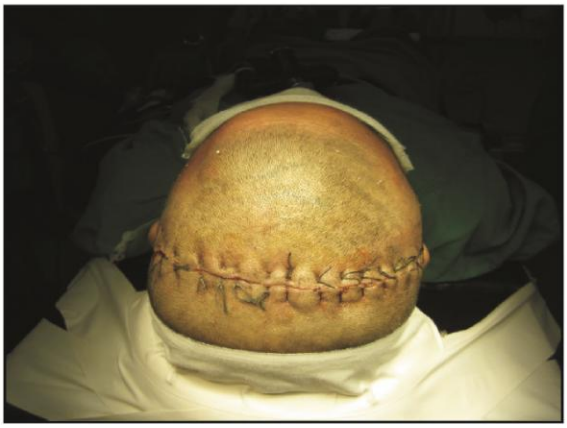


FIG VII: POSTOPERATIVE SURGICAL WOUND

CONCLUSION

Simpson's grade I excision of meningioma nearly give complete cure and large defect ask for timely reconstruction to restore function in turn minimizes the chances of infection. Timely screening of such patients with neurofibromatosis type II is important and may be helpful to avoid getting such a large lesion in late stages.

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