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## Management of complicated Neurofibromatosis – Our experience

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**N**eurofibromatosis is an Autosomal dominant inherited disorder, with the 6 classical criteria for its diagnosis. Many of these patients presents with varied complications. Neurofibromatosis has been a well-known genetic disorder from 18th century. There has been an extensive work on Neurofibromatosis which leads to multiple criteria for its classification. Neurofibromatosis is associated with multiple tumours ranging from neurofibromas, spinal tumours, schwannomas, gliomas, meningiomas, ependymomas, intraventricular SOL, malignant peripheral nerve sheath tumours etc. Neurosurgeons are quite often challenged to face the following complications - multiple intracranial SOL's, multiple spinal tumours, multiple meningiomas, recurrent spinal tumours, recurrent schwannomas etc.

Neurofibromatosis are also known for their early malignant degeneration to malignant peripheral nerve sheath tumours. Neurofibromatosis type 1 (NF1) is a dominantly inherited tumour predisposition syndrome that targets the central and peripheral nervous system. It is caused by mutations of the NF1 gene which serves as a negative regulator of the cellular Ras/MAPK (mitogen-activated protein kinases) signaling pathway. Owing to the complexity in some parts of its clinical

diagnoses, due to the disease incomplete penetrance and varied penetration, there is a need for better understanding of its molecular relationships. A genetic characterization of this disorder will be helpful in the clinical setting.

At our Institute (Bangur Institute of Neurosciences, Kolkata), we have done a prospective study of all the cases presenting with Neurofibromatosis. In this paper, we are focusing on the rare challenges we faced in managing complications of Neuro-fibromatosis.

### Speaker Biography

Shahid Iftexhar Sadique has completed his Neurosurgery (MCh) at 32 years from IPGME&R (Institute of Post Graduate Medical Education and Research), WBUHS (West Bengal University of Health Sciences), Kolkata, INDIA. He has been giving his unrelentous service to the people of West Bengal for the past 19 years. He has operated many complex Spine, Neurovascular and Neuro endoscopic procedures successfully. He has a number of national and international publications with his field of interest in Neuro Oncology. Presently he is holding a position of Associate Professor of Neurosurgery, at the premier tertiary care Institute, Bangur Institute of Neurosciences IPGME&R, Kolkata, India.

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