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INTRAVENTRICULAR SCHWANNOMA - A RARE CASE REPORT

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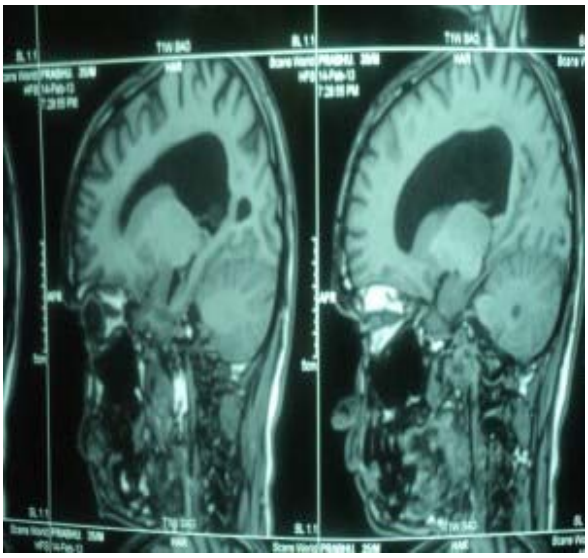
Abstract :
The Intraventricular Schwannoma is a rare benign intracranial tumor. Its exact origin is unknown. We report a case of 33 years old male, presented with focal seizures involving the right Upper Limb with headache and projectile vomiting. He was evaluated and diagnosed to have a Intraventricular tumor involving the Left Lateral ventricle with hydrocephalus. He underwent Right VP Shunt and subsequently, the definitive surgery was done and the lesion was excised completely. Postoperatively, he had improved. Histopathologically, the tumor showed the features of Schwannoma. Since the tumor was excised completely, no further treatment is necessary..

Keyword :
Intraventricular Schwannoma, a rare intraventricular tumor.

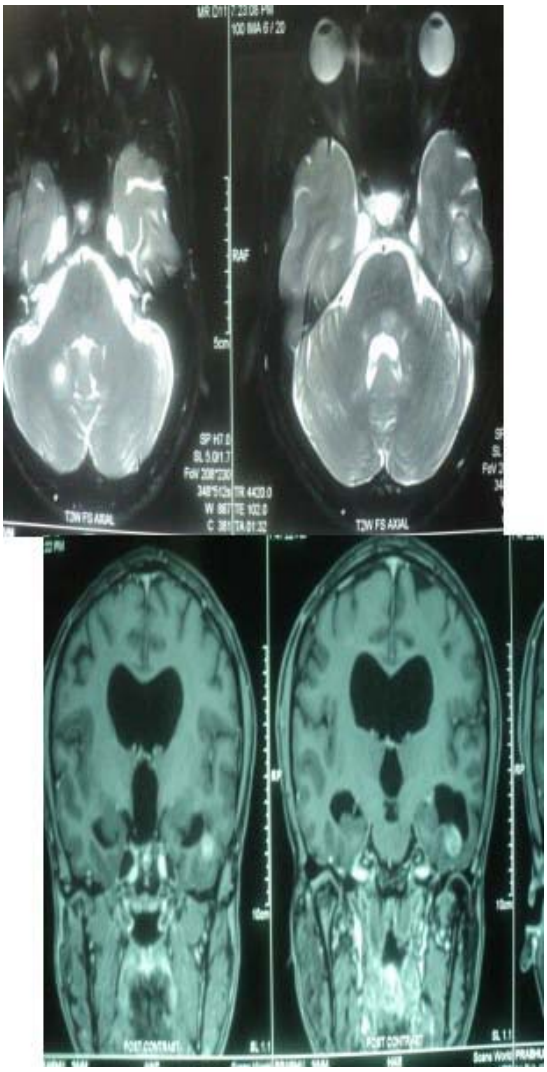
CASE REPORT :
A 33 years old man presented with focal seizures involving right Upper Limb for about one year duration. The seizures were not preceeded by aura, not associated with tongue bite or drooling of saliva. It was not associated with loss of consciousness or automatism, no postictal confusion or weakness. He had 2 to 3 episodes per day and for the past 3 months, he had increased seizure episodes ranging from 5 to 6 episodes per day. Each episode last for about 5 to 10 minutes.He had headache for the past 6 months which was of dullaching, holocranial, intermittent and no diurnal variation. The headache was severe for the past 15 days, got relieved by vomiting. He also gave history of projectile vomiting for the past 5 days.

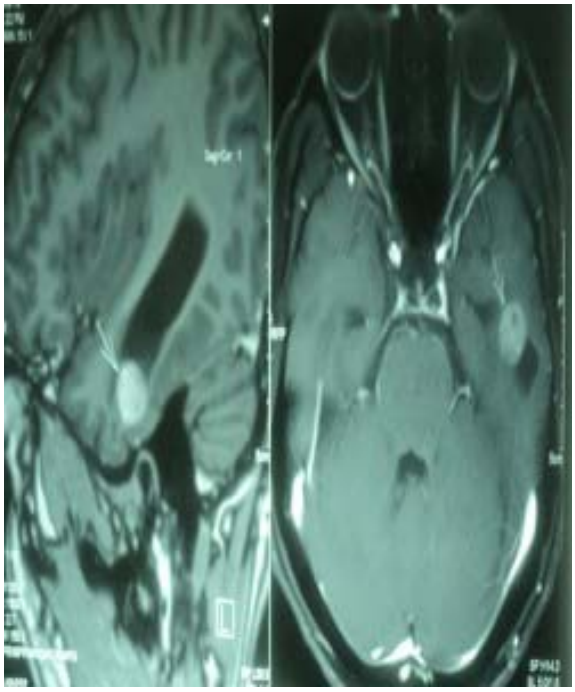
He had no symptoms suggestive of Higher mental function or Cranial nerve disturbances. His Bladder and Bowel functions were normal. He had no constitutional symptoms related to inflammatory pathologies. There was a family history of Neurofibromatosis Type I. His mother was affected with features of NF-1, but remain asymptomatic.

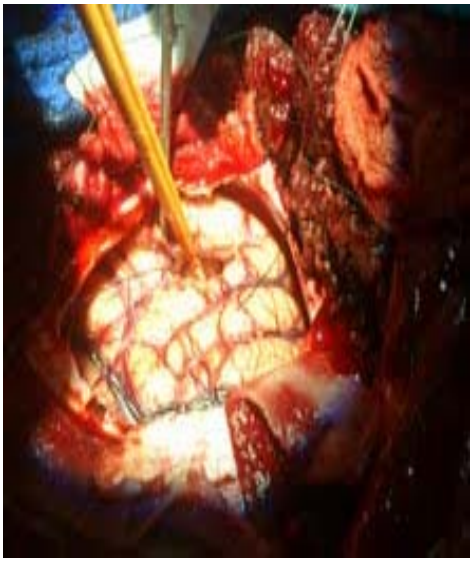
On General examination, he had multiple subcutaneous nodules and café- au-laut spots distributed through out his body. On Neurological examination, he had bilateral papilloedema. His spinomotor system, sensory system, Spine and Cranium were normal. No cerebellar or Extrapyramidal signs were present.

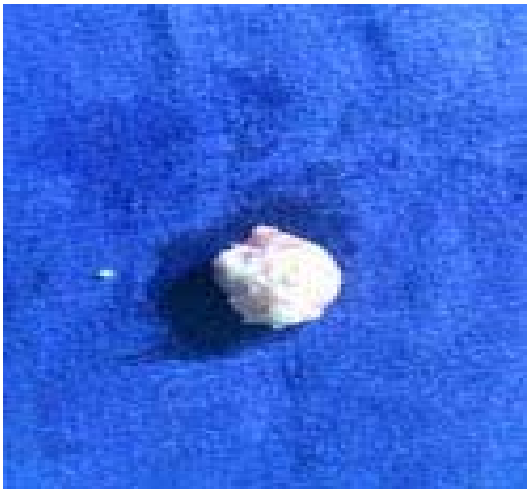


He was evaluated with MRI Brain which showed a well circumscribed T1 hypointense and T2 hyperintense lesion present in the left lateral ventricle with dilated ventricles. On Gadolinum, it showed homogenous enhancement. MRI Spine screening was normal.

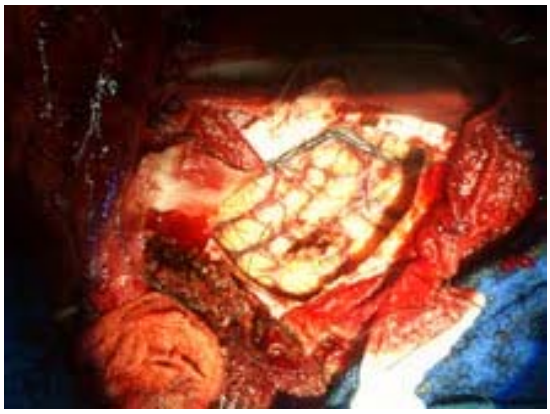




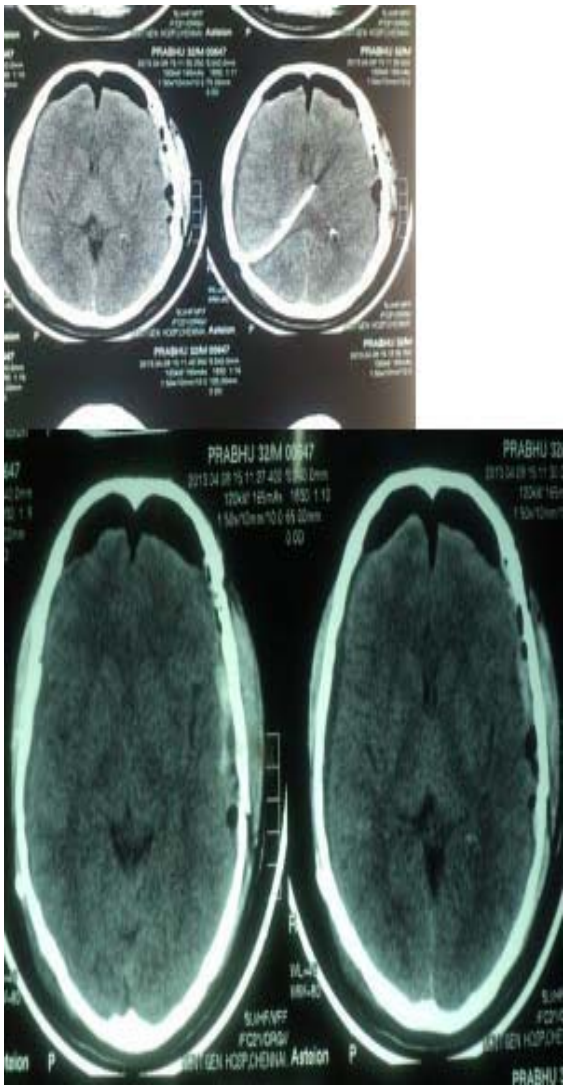


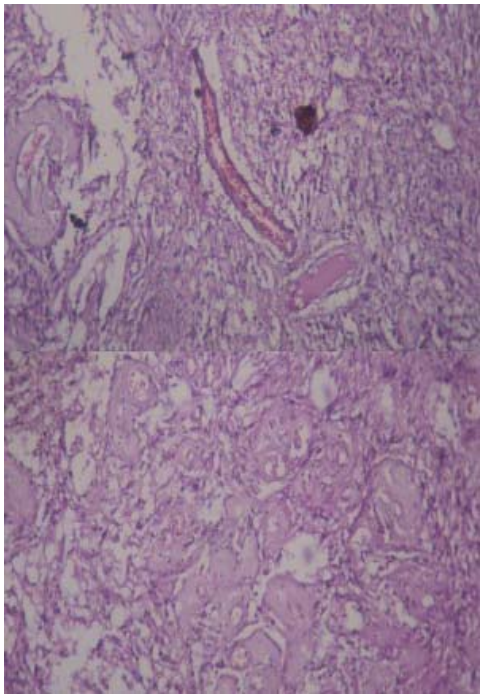


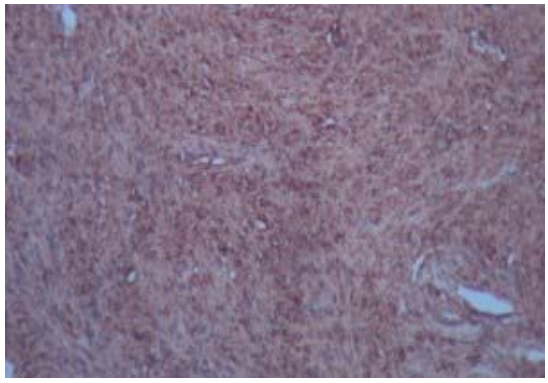
We proceeded with Right Ventriculoperitoneal shunt. Subsequently, we did Right FrontoTemporoParietal craniotomy and through Transcortical incision, the right lateral ventricular wall was opened and the lesion was accessed. The tumor was about 3.5 X 2.5 cm, yellow white in colour, firm in consistency with nodular surface and was excised completely. A part of the tumor was sent for "squash" at our institute, which was reported as "Schwannoma".



The postoperative period was uneventful and he got relieved from his symptoms. The Post operative CT Brain showed the complete excision of the tumor and no residual lesion was found.







Post operatively, the Histopathology showed the neoplasm composed of spindle to oval cells with spindle shaped nuclei. The cells are arranged in fascicles and show nuclear palisading in some foci. There are extensive areas showing Xanthomatous changes, hyalinised blood vessels and cystic degeneration favouring SCHWANNOMA " (WHO Grade I).

He was discharged from the hospital after the suture removal. He is in regular follow up without any complications or recurrences till now.

DISCUSSION :

Schwannomas comprise about 8% of all Intracranial nplasms (Ref :Arquivos de Psiquiatr. Vol. 67 No.4 Sao Paulo Dec. 2009). Majority arise from the Vestibulocochlear nerve , others from the sensory fibers of 5^{th}, 7^{th}, 9^{th}, 11^{th} nerves.

The Schwannomas can present as primary meningeal, Intracerebral and Intraventricular tumors. Majority arise either in the lateral ventricles or in the fourth ventricle (Ref : 1. Arquivos de Psiquiatr. Vol 67 No.4 Sao Paulo Dec. 2009, 2. Barbosa MD, Fernandes R 2001,Cystic Intraventricular Schwannoma – case report, Neurocirugia (Astur) 12:56-60). The first reported Schwannoma is from the Third ventricle.

Since the nerve fibres of Central Nervous System is not invested with Schwann cells, the exact site of origin is unknown. However, similar to the hypothesis, about the histogenesis of Spinal Intramedullary tumors and Intracerebral Schwannomas, which are slightly more common than Intraventricular Schwannomas, different possibility may be considered.

The most popular hypothesis is that it may arise from the sympathetic plexus around blood vessels. These vascular nerves can be found around the medullary vessels, in the choroid plexus and in the meninges.

Alternative hypothesis says its origin from the ectopic neural crest derived cells which has been displaced during embryogenesis. This may point to a link between Intraventricular Schwannomas and the so called Neurocristopathies, which comprise a spectrum of dysgenetic & neoplastic disorders associated with alteration in migration, growth and differentiation of Neural crest tissue during embryogenesis.

The Neurocristopathies comprise the lesions originating from the Neural crest derived cells in abnormal locations, during the process of embryogenesis. The abnormalities in the process of neural crest derived cells migration, growth and differentiation may produce a spectrum of neoplastic and dysgenetic disorders.The prominent examples include, Neurofibromatosis and Hirschprung disease, Waardenburgh syndrome. (Ref : Bolande RP Neurocristopathies : Paediatric Patho Lab Med 17 : 1-25)

The Intraventricular Schwannomas are usually solitary, unless associated with specific genetic syndromes like Neurofibromatosis.

Neurofibromatosis refers to a number of inherited conditions, that are clinically and genetically distinct & carry a high risk of tumor formation in the brain, spinal cord, meninges. It is a autosomal dominant disorder, the severity in the affected individuals can vary and this may be due to the variable expressivity. Approximately, half of the cases are due to de novo mutation & no other affected family members are seen. It affects both males and females equally.

The Neurofibromatosis are as follows : (1) Neurofibromatosis Type I : Autosomal dominant disorder caused by the mutation or deletion of NF-1 (“neurofibromin”) gene, located at chromosome 17 (17q11.2), in which the nerve tissue grows tumors (Neurofibromas) that may be benign & may cause serious damage by compression of nerves and other tissues. (2) Neurofibromatosis Type II : due to mutation of “Merlin” gene located in chromosome 22 (22q11). They have the risk of developing cancers in the Brain & leukemias. They have the increased propensity to develop Bilateral Acoustic in the Schwannomas. (3) Schwannamatosis – they develop painful schwannomas in the cranium, spinal cord and peripheral nerves. Our patient had

clinically demonstrable cutaneous lesions like cutaneous nodules & café-au-lait spots, skin biopsy was done.

Another proposed hypothesis is that of "Schwannosis". It is actually a multipotent cell theory, stating the transformation of multipotent cells into Schwann cells & proliferation of foci of Schwann cells. Schwannosis is otherwise described as the non-neoplastic proliferation of Schwann cells in spaces around the blood vessels of brain & spinal cord. This theory is based upon the ideas that Schwann cell, a specialized mesenchymal element is derived from primitive multipotent cells that can give rise to other types of specialized mesenchymal elements in the appropriate cellular milieu. Based on this theory, there is differentiation of multipotent mesenchymal cells into schwann cells, stem cells & neoplastic transformation from autonomic cells found in the intrinsic arteries & choroid plexus. This is supported by the identification of nerve fibres in choroid plexus of 4 th ventricle by Benedict in 1874 & Stohr in 1922.According to this theory,Schwannomas refer to hamartomatous lesions of schwann cells & reticulin fibres that are formed from the conversion of pial mesodermal cells into schwann cells.

Another proposed theory is the "Cancer Stem cell Theory ". This centers on the idea, that tumors arise from a small subpopulation of stem cells,which have the ability to self-renew, produce daughter tumorigenic cells & can give rise to non tumorigenic cancer cell phenotypes. Recently, stem cells have been detected in Glioblastoma, Medulloblastoma, Ependymoma. Stem cells have the ability to reform parent tumors, when implanted into nude mice & can partially differentiate in vivo. It has been shown that there is upregulation of certain stem cell markers like "Nestin", CD44 in some Schwannomas. A study shows about 92% of human vestibular schwannomas demonstrated an upregulation of CD44 at mRNA level compared to normal Schwann cells.

Recently, another proposed hypothesis is that these Schwannomas arise from aberrant Peripheral nerve fibres due to abnormal proliferation from localized TRAUMATIC INJURY (e.g., focal axonal proliferation in the pons adjacent to the infarcted brain tissue or traumatic Schwannomas associated with spinal cord compression. They may not be true neoplasms, but rather regenerative proliferation of injured peripheral nerves found within central nervous system.

Neuroradiologically, the intraventricular Schwannomas cannot be differentiated with certainty from the other intraventricular tumors like Ependymomas, Choroid plexus tumors. Histologically, the diagnosis is straightforward, with the differential diagnosis of pilocytic astrocytoma and Fibroblastic meningioma. However, the abundance of reticulin fibres and lack of GFAP Immunoreactivity rules out a pilocytic astrocytoma with strong expression of S100 and absence of Immunoreactivity for EMA (Epithelial membrane Antigen) excludes fibroblastic meningioma.

About 55 cases of Intraventricular Schwannomas have been reported, so far, in the literature worldwide, to the best of our knowledge. In India, less than 10 cases have been reported so far. This case is presented for its rarity and its association with Neurofibromatosis Type I.

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