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Obstructive Hydrocephalus with Impaired Ventricular Compliance Resulting In Visual Deterioration In An Adult With Vestibular Schwannoma

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Abstract: *Obstructive hydrocephalus is often associated with ventriculomegaly. Intraventricular pressure is often transmitted to the visual pathway resulting in impaired vision. However, in rare circumstances of impaired ventricular compliance, intraventricular pressure rises without concomitant ventricular dilatation. We present the case of a 38 year-old lady with a right sided vestibular schwannoma (Hannover grade 4b) and features of raised intracranial pressure (progressive blindness from papilloedema) with normal sized ventricles (Evans ratio 0.28). Her unusual presentation of elevated intraventricular pressure and normal sized ventricles resulted in a long referral chain as to the cause of her worsening vision and a delay in her care. She had an external ventricular drainage with marked opening pressure and subsequently sub-occipital craniectomy and tumour resection with satisfactory outcome. Her vision improved postoperatively. There is a need for high index of suspicion in patients with obstructive tumour and visual impairment with normal sized ventricles.*

Key Word: *hydrocephalus, ventricular non-compliance, vestibular schwannoma.*

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I. Introduction

Obstructive hydrocephalus is a well known complication associated with vestibular schwannoma and has been reported in about 3.7-42% of patients.[1,2,3,4] It results from the mass effect of the tumour obstructing the cerebrospinal fluid (CSF) outlets of the fourth ventricle. In some other cases, it results from malabsorption of CSF at the arachnoid granulations due to the high protein concentration secreted by the tumour or meningeal adhesion following minor haemorrhages from the mass lesion.[1,2,5] Generally, there is associated ventricular dilation as the fluid accumulates leading to raised intracranial pressure which manifests clinically as recurrent headaches, early morning vomiting and visual impairment.[6] In rare cases, Intraventricular pressure rises without concomitant ventricular dilation due to possibly impaired ventricular wall compliance for some unexplained reasons.

We present a case of a young lady with obstructive hydrocephalus and blindness following a large vestibular schwanomma without concomitant ventricular dilation despite CSF being under high pressure on ventricular cannulation.

II. Case report

A 38 year old female with five year history of progressive painless hearing loss and one year history of recurrent pancranial headache, initially low grade but lately distressing, worse early hours of the morning and relieved transiently by vomiting or analgesics.

She subsequently developed progressive painless visual loss of about six months, characterized by blurring of vision but no peripheral visual loss or diplopia. There was no associated endocrinopathy. She visited several optometrists and ophthalmologist who recommended change in reading glasses on several occasions without significant improvement. A diagnosis of glaucoma was made at some point in her care and treatment instituted without any significant benefit. Her vision continuously worsened despite all intervention until she needed aid to move about. She had CT scan and MRI done which showed a posterior fossa tumour necessitating referral to the neurosurgeon. Other symptoms she noted within this period were gait abnormality with occasional falls to the right, right handed tremors and subtle left sided hemibody weakness.

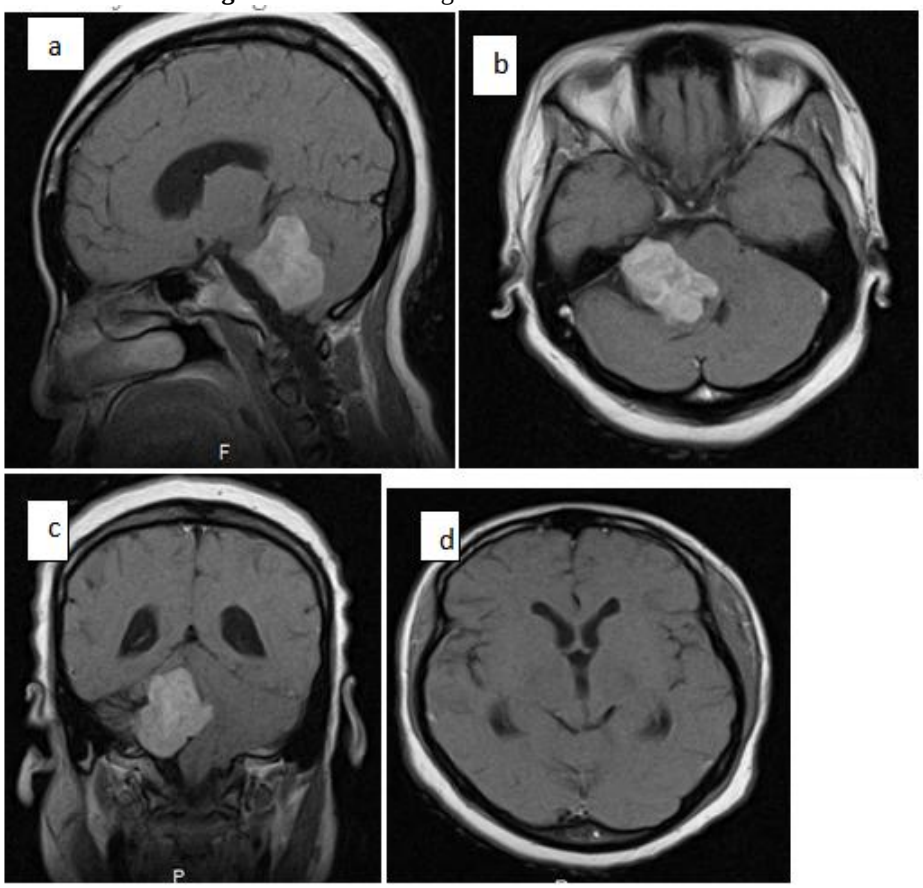
Examination showed a young lady with depressed mood, requiring an assistant to guide her while walking and a subtle ataxic gait. Speech was good and memory was succinct for both short and long term

events. She had NPL on the right and light perception on the left. Fundoscopy showed marked papilloedema bilaterally (Frisen grade IV). She also had sensorineural hearing loss on the right ear but the left ear was normal. There was mild right sided facioparesis (House and Brackmann grade II). She had positive right sided cerebellar signs and subtle left sided hemibody weakness. Deep tendon reflexes were exaggerated in the lower extremities with a positive babinski on the left, sensation was preserved in all modalities.

MRI showed an oblong shaped mass in the right CP angle measuring 41 by 45 by 33mm, isointense on T1 and heterogeneously hyperintense on T2 and FLAIR. There was associated brainstem compression with effacement of the 4th ventricle (Hannover grade T4b). The lateral and third ventricles were not enlarged (Evans ratio 0.28). The mass enhanced heterogeneously with contrast.

She had emergency external ventricular drainage which revealed a marked opening pressure. CSF chemistry and culture were normal except for elevated protein (60mg/dl). She later had right lateral sub-occpital craniotomy and gross tumour excision via a retrosigmoid approach. She had postoperative pseudo-meningocele which was managed conservatively with pressure bandaging. Her vision gradually improved to counting fingers at one meter and she can ambulate unaided. Also, she had significant resolution of the cerebellar symptoms and hemibody weakness and is gradually being reintegrated into the society.

Figure1. MRI showing vestibular schwannoma



Contrast MRI (a) Sagittal (b) axial (c) coronal (d) axial views showing a right CP angle tumour (Hannover gradeT4b) causing marked ponto-cerebellar compression with effacement of the fourth ventricle. Note the normal sized ventricles.

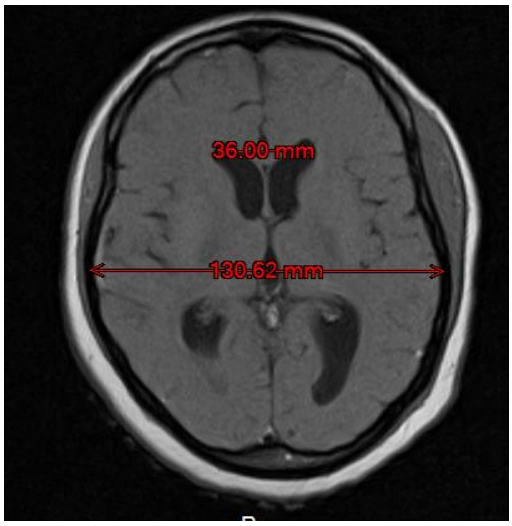


Figure 2. Evans ratio estimation.
Frontal Horn (FH) ratio / Biparietal Diameter (BPD) 36/130.6 Evans ratio of 0.28

III. Discussion

Ventricular compliance refers to pressure volume relationship where rise in transmural pressure results in increase in volume of the ventricle**.**[7] Impaired ventricular compliance results when elevated ventricular pressure is not associated with a concomitant increase ventricular size. Our patient had markedly elevated intraventricular pressure and papilloedema due to obstructive effect of the tumour without concomitant associated dilation of the ventricles. Her Evans ratio was 0.28 which is within the normal allowable range for standard lateral ventricles. This occurrence is rare as few cases have been reported in literature and none to the best of our knowledge in this part of the world.[8,9]

Grainger and Dias[8] suggested that high protein secreted by VS may impair reabsorption of CSF, albeit intermittently causing a communicating type of hydrocephalus. However, our patient, though had elevated protein did not have features of hydrocephalus. It is possible that the underlying mechanism for non-compliance of the ventricles may be related to excess protein and other factors elaborated by VS which initiates a low grade inflammation in the ventricles, meninges and surrounding brain parenchyma.

Furthermore, it is believed that the ependyma acts like an elastic membrane lining the ventricles and possibly due to low grade inflammation of the ependymal cells and associated periventricular gliosis results in less tendency to distend in response to raised intraventricular pressure[10,11] These changes result in loss of elasticity of the ependymal wall and failure of ventricular compliance. However, some other studies have demonstrated alteration of the viscoelastic properties of the brain parenchyma as the aetiological factor in impaired ventricular compliance as seen in certain pathologies and inflammatory conditions.[12]

Her unusual presentation caused a diagnostic dilemma as the initial managing physician attributed her visual loss to glaucoma rather than papilloedema from raised intracranial pressure. This caused a delay in her care as all efforts to revert her deteriorating visual symptoms using medical management for glaucoma was unsuccessful. This calls for a high index of suspicion and low threshold for intervention in managing patients with obstructive intracranial lesion, visual deterioration and normal sized ventricle. The reported rarity of VS in the sub-region[13] underscores the need for increased vigilance as these tumours could masquerade as in the index case

She had emergency external ventricular drainage to relieve raised ICP and subsequently a definitive surgery to resect the tumour. This is an expedient method of managing obstructing VS,[1,14] as it relives the pressure and allows time to adequately plan the definitive procedure. Furthermore, in the long run, it prevents insertion of a permanent hardware as the natural CSF pathways opens up after tumour excision as she had since been stable with improving vision and non dependent on shunts.

IV. Conclusion

Our report demonstrated visual deterioration following papilloedema in a patient with elevated intraventricular pressure and normal sized ventricle. This impaired compliance may be related to excess protein elaborated by the tumour which initiates a low grade ependymal inflammation and periventricular gliosis. There is a need for a high index of suspicion of raised intraventicular pressure as a cause of papilloedema in brain tumours with potential high CSF protein.

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