Transient internuclear ophthalmoparesis associated with mutism following midline cerebellar tumour surgery in an 8-year boy: A unique case

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Abstract: Mutism and neurobehaviour symptoms are well known features, which may occur following surgical excision of mass lesion of various histopathologies in the posterior cranial fossa, during the postoperative period. Mutism may be rarely associated with ataxia of eyelid closure and paresis of external ocular muscles. However, internuclear ophthalmoparesis is not reported in association with mutism following posterior cranial fossa surgery. We report an 8-year–boy, who developed internuclear ophthalmoparesis following suboccipital craniectomy for decompression of vermian medulloblastoma. The clinical features, aetiopathogenesis, management of transient internuclear ophthalmoparesis associated with mutism and pertinent literature is reviewed in short.

Key words: Posterior fossa, internuclear ophthalmoparesis, medulloblastoma surgery, mutism

Introduction

Internuclear ophthalmoparesis (INO) is one of the most localizing brainstem syndromes, which is caused by a lesion involving medial longitudinal fascicules in dorsomedial brain stem tegmentum of either the pons or the midbrain [1,2]. INO is an oculomotor syndrome characterized by ocular dysconjugancy during horizontal saccades, with slowing of adducting eye movements, with or without ocular limitation, and abduction nystagmus in the other abducting eye. Internuclear ophthalmoparesis is rare occurrence following intracranial surgery. We report an 8-year–old–boy developed INO, in the immediate postoperative period following decompression of vermian medulloblastoma. The development of mutism and associated neuropsychiatric symptoms subsequently many reports have emphasized the importance of impaired eye openings, pseudo
bulbar symptomatology. However, INO was not highlighted in any case report, except as associated signs in association of cerebellar mutism. However, no emphasis was put regarding the onset, course, and neuroimaging in a series of 12 cases reported by Pollack et al [7].

Case report

Patient presented with a six week history of progressive headache and ataxia. One week prior to admission ataxia markedly worsened, with head tilt and diplopia, on admission, he was conscious, irritable, and stable aerodynamically. Fundi examination revealed bilateral papilloedema. Left sixth cranial paresis was present. He power was normal in all limbs. He had marked ataxia, making walking difficult. He had bilateral cerebellar signs. Neck rigidity was present. Routine haematological, biochemical and coagulation profiles were normal. Computerized tomography of brain revealed hypodense mass lesion involving vermis; it was 4 cmX4.5 cm in diameter, causing obstructive hydrocephalus. (Figures 1-3) On contrast administration showed uniform enhancement. Radiologically a diagnosis of vermian, mass with obstructive hydrocephalus was made.

He underwent CSF diversion procedure of ventriculoperitoneal shunt. Two days following ventriculoperitoneal shunt surgery, he underwent elective suboccipital craniectomy in the sitting position. Tumour was exposed and resected via an inferior transventricular approach. The tumour was completely excised using with help of internal decompression of CUSA and haemostasis was achieved with bipolar cattery. The floor of fourth ventricle was not invaded or infiltrated by tumor. CSF was freely flowing through the aqueduct. The content of posterior fossa was lax and pulsating. A histopathological examination confirmed the diagnosis of medulloblastoma.
After reversal of anaesthesia, he was awake with intact neurologically, except left sided sixth nerve paresis and cerebellar ataxia. He was well for forty eight hours, and then developed mutism. He was lying curled up in the bed with eyes closed. He was not obeying commands. He refused oral intake of solid and liquid fluid. (Figure 4) NC CT scan head was done to evaluate for the deterioration in the neurological status revealed oedema around the resected cavity, however no haematoma was encountered in the tumour bed. (Figure 5) Magnetic resonance imaging could not be done in view of economic constrains. After 5 days, he showed progressive improvement in the neurological status and started accepting oral feed; personality carnages also improved. He becomes more cooperative. Ocular muscle examination showed features consistent with the diagnosis of internuclear ophathalmoplegia. His speech also improved gradually. However feature of
ophthalmoparesis did show improvement up to six weeks after surgery, but resolved completely, when seen at 8 weeks of follow up after surgery. The cranial CT scan with contrast-enhanced scan had revealed residual lesion, he received craniospinal irradiation.

Discussion

Common features of cerebellar mutism syndrome include a decreased speech output or mutism, transient irritability or apathy. Although other features transient eye closure, difficulty with feeding, associated with incontinence of bladder and bowel is reported. Siffert et al analyzed 16 cases of cerebellar mutism syndrome and defined more comprehensive neurological dysfunction in the cerebellar mutism syndrome [9]. A severe global cerebellar dysfunction, a communication impairment affecting receptive and expressive languages, severe inattention, apathy, hypokinesis, mood liability, irritability, apathy, hemiparesis, or motor apraxia, transient difficulty with eye opening and lack of control of bowel and balder.

The causes and anatomical basis of cerebellar mutism syndrome in children with posterior fossa surgery is still controversial. Cerebellum has been implicated for cerebellar mutism syndrome [4]. The mutism occur exclusively with midline cerebellar mass lesion, being resected via inferior vermal incision was suggested as bilateral injury to inferior vermal region is crucial for development [9]. Although Dailey et al. had suggested the splitting of the inferior vermis during a tumour resection, but less than 25% of children undergoing similar procedure develops signs of cerebellar mutism syndrome. However several points indicate that anatomical location may not exactly in the inferior vermis, but localized to the adjacent structures. However, many cases of Pollack et al where tumour was located in the superior vermis and inferior vermis was not violated also developed and this anatomical center may not be vermis but instead more laterally located [7].

The onset and recovery is usually delayed hours to days following surgical procedure. Indicating it is caused by intraoperative injury or infarction [7]. Pollack et al also observed postsurgical oedema has been implicated but clinical course often outlast the resolution of oedema. Other mechanism as alteration in neurotransmitter levels and synaptic or transynaptic degeneration of connecting structure may be responsible, which might correlate well with time lag between the time of actual surgical injury until the onset of symptoms. The recovery mechanisms also remain still unclear. After a several week of plateau, children usually notice a fast recovery of speech. However cerebellar ataxia may persist in the majority of cases, so some aspect of clinical recovery may not depend on cerebellar injury [4].

These may reflect the sequel of injury to the afferent or efferent pathway to dentate nuclei, which are involved in the initiation of violation movement [4, 7]. Bilateral impairment of paramedian cerebellar dentate nuclei, its efferent or afferent connection or a combination of factors may be involved [7]. The delayed onset of speech impairment probably related interval of time required for
Oedema resulted due to surgical manipulation in the surgical field has reached the structure [7]. This symptom complex may be associated with a varied duration of speech impairment and spectrum of neurological and neurobehavioral abnormality can vary. In the simple form of spectrum, only impairment of inco-ordinating the complex bilateral integrated movement necessary for speech production. In relatively more severe cases may have additional impairment of inco-ordination of oropharyngeal muscles movement involved in chewing or swallowing movement. However in more severe cases, may have broad spectrum of volitional movement impairment including eye opening and voiding. This more global impairment has also been substantiated on neuropsychiatry evaluation. Accordingly, focal bilateral lesion to dentate nucleus and its connection to while more complex involves paravermian region or its connection.

During recovery, which may occur days to weeks following onset, global akinesia, and urinary retention recovery is followed by eating and finally speech improvement may occur with a dysarthria or bizarre vocal quality and finally regaining of almost normal speech. Wisoff et al described psuedobulbar syndrome after posterior fossa surgery in children, often associated with oedema of bilateral cerebellar peduncle and brainstem, may reflect dysfunction of brainstem [10]. However, unusual personality changes as also observed in mutism syndrome are well described and manifestation of primary brainstem lesion glioma of brainstem and vascular insufficiency from proximal basilar artery ischemia [6, 1]. These psuedobulbar manifestation may also be caused by bacterial meningitis. However, in our case also the INO also improved after four months. INO syndrome is caused by damage to these structures including medial longitudinal fasciculus, paramedian pontine reticular formation, or the ipsilateral sixth nerve nucleus. Commonest causes of INO is multiple sclerosis and infarction of brain stem, however, lesser commoner are tumors of the brain stem and cerebellum and extremely rarely, can be detected in the postoperative period following of the posterior fossa tumors removal [2].

Pollack et al. reported a case, had vermian lesion with metastasis in both hemisphere with fourth ventricle invasion had medulloblastoma developed, mutism 72 hours after surgery, was lying curled in the bed and poor oral intake, improvement started on 18th day and almost complete recovered by 4 weeks. However, no detail of onset or progress or remission of INO was provided. He also developed INO, left arm ataxia after 18 days of surgery and these improved over four weeks, however ataxia persisted [7].

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