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Case Report

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A rare case of a diffuse intrinsic pontine glioma presenting as pathological laughter and apparent hyperactivity in a child

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Abstract

Introduction: Diffuse intrinsic pontine gliomas generally present with cranial nerve palsies with cerebellar and pyramidal-tract involvement. The pons has generally not been regarded as a structure that mediates complex affective behaviour. However pathological laughter has been reported with pontine pathology in various forms.

Case Presentation: We present a case of pontine glioma presenting as pathological laughter and apparent hyperactivity in a previously healthy 11-year-old boy.

Discussion: Literature states that pontine gliomas are the subgroup of brainstem gliomas with the worst prognosis. This patient remained with deficits in motor functions and coordination despite intense treatment with radiotherapy.

Keywords: Pontine, glioma, pathological laughter, attention deficit hyperactivity disorder

INTRODUCTION

Diffuse intrinsic pontine gliomas (DIPG) are paediatric high-grade gliomas characterized by infiltrative tumours of the brainstem. These are astrocytomas histologically[1] and account for 10-20% of all pediatric brain tumours[2].

They are amongst the most challenging tumours to treat. Surgery is not an option due to the infiltrative nature of the lesion, and the effects of radiation therapy are temporary, and no chemotherapeutic agent has demonstrated significant efficacy[3].

Children with pontine glioma usually present classically with ataxia, motor deficits and cranial nerve palsies[4]. Pathological laughter and crying is defined as a condition with relatively uncontrollable episodes of laughter, crying or both[5]. The episodes either do not have an apparent triggering stimulus or are triggered by a stimulus that would not have led the subject to laugh or cry before the onset of the condition.

It is an uncommon symptom usually caused by bilateral, diffuse cerebral lesions. The current view is that it results from any lesion in the cortical pathways that inhibit the centre for pathological crying and laughter, located in the brainstem[6]. Midbrain involvement causing pathological laughter is rare[7]. However, there are case reports

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of a pontine abscess and pontine gliomas presenting with pathological laughter and behavioural changes[8,9].

We present the case of a previously healthy 11-year-old boy who presented with pathological laughter and behavioural abnormalities who was subsequently found to have a pontine glioma.

CASE PRESENTATION

An 11-year-old schoolboy was referred to the consultant psychiatrist for the evaluation of hyperactivity disorder, following the routine school medical inspections.

The teachers reported that he was laughing aloud, for no apparent reason on several occasions. Once he started laughing, it appeared that he had difficulty controlling his laughter. At other times, he was reported to be seemingly not interested in participating during teaching sessions and would keep his head on the desk saying he was tired, while at other times seemed distractible and unable to sit still.

This behaviour was out of character to his usual self, which prompted the teachers to inform the matter at the school medical inspection. Furthermore, his parents revealed that there were subtle changes in his gait, with a tendency to fall, which were unfortunately overlooked.

He had also complained of headaches frequently and would neglect to attend to his schoolwork. Before the onset of these symptoms, he had a good academic record at school and had scored 178 out of 200 marks for his grade 5 scholarship exam the previous year.

On examination, he was laughing and giggling aloud, for no obvious reason. It appeared that he was having difficulty controlling his laughter. It was difficult to engage him in the conversation as he was unable to focus on the questions being asked from him.

On examination, he had a broad-based gait with a tendency to veer off to the left side and was unable to perform the tandem gait. He had horizontal nystagmus as well as past pointing and dysdiadochokinesia. HIs motor system was normal except for brisk reflexes. The cranial nerves were normal. Bilateral fundoscopy and sensory system examinations were normal.

Following an assessment by the paediatric neurologist, it was decided that he required immediate radiological imaging. While lesions in this region ideally require an MRI, at the time of presentation, the only available imaging was a CT scan. He subsequently underwent an MRI scan at the National Hospital Colombo. Imaging revealed a pontine glioma measuring 2.5cm by 3cm. He was transferred to the cancer hospital where he underwent a series of radiotherapy treatments. This resulted in the reduction of symptoms, including his pathological laughter, although he was never able to reach his premorbid levels of schooling, and attend to activities of daily living due to the neurological deficits that remained. At the time of writing this article, almost one year after his initial presentation, he was readmitted with recurrence of symptoms, with ongoing radiotherapy.

DISCUSSION

This was a rare presentation of a pontine glioma. The child was referred for an assessment of hyperactivity disorder by the medical officer at the school medical inspection, upon being alerted by the school teachers. However, several features in the history weren't typical of a child with attention deficit hyperactivity disorder. The age of hyperactivity disorder onset is generally before the age of 6 years with a significant impact on schoolwork[10], whereas this child presented at the age of 11 years while performing very well at school until the onset of symptoms. Although he was seemingly clumsy, it was due to the incoordination resulting from the direct effects of the tumour. His pathological laughter was interpreted as him disregarding the rules and regulations of the class, though this was again due to the effects of the tumour.

This case also highlights the importance of a comprehensive physical examination particularly when the symptoms do not fit into a uniform pattern of a particular disorder.

The traditional and currently accepted view is that PLC is due to the damage of pathways that arise in the motor areas of the cerebral cortex and descend to the brainstem to inhibit a putative centre for laughter and crying. In that view, the lesions `disinhibit' or `release' the laughter and crying centre. The neuroanatomical findings in a recently studied patient with PLC, along with new knowledge on the neurobiology of emotion and feeling, allowed us to revisit the traditional view and propose an alternative. Here we suggest that the critical PLC lesions occur in the cerebro-ponto-cerebellar pathways and that, as a consequence, the cerebellar structures that automatically adjust the execution of laughter or crying to the cognitive and situational context of a potential stimulus, operate based on incomplete information about that context, resulting in inadequate and even chaotic behaviour."

Irradiation has been described as an effective element for the treatment of pontine gliomas. Intensive chemotherapy seems to be important in achieving a better outcome survival[11]. Despite undergoing routine radiotherapy, he showed very little improvement in overall symptom improvement.

It is pertinent for clinicians to be mindful of the significant degree of overlap between neurological and psychiatric symptoms which can easily be overlooked when working in a busy and overcrowded outpatient setup.

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Author contributions
Dr A S Rajapakse examined the child and made the initial draft
Dr S R Perera did the complete writeup of the article

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Availability of data and materials
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Ethics approval and consent to participate
Informed verbal consent was obtained from the parents of the patient

Competing interests
No conflicts of interest by either author

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