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Idiopathic Intracranial Hypertension in Sickle Cell Disease: A Paediatric Case Report

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ABSTRACT

In this article, we present the case of a 12-year-old girl with sickle cell disease (SCD), who presented with the severe headache. She had bilateral 6th cranial nerve palsy and papilloedema. The common sickle cell-related vascular causes of headache were ruled out by neuro-imaging. She then had a lumbar puncture and was diagnosed with idiopathic intracranial hypertension (IIH). This case demonstrates that IIH can affect younger children with SCD and should form a part of differential diagnosis when investigating causes of headache in SCD.

Keywords: Sickle Cell Disease; Idiopathic Intracranial Hypertension; Headache in Children

1. Introduction

The common causes of headache in healthy children are tension headaches or migraine [1]. Children with sickle cell disease (SCD) frequently complain of headaches. In one study, the overall prevalence of frequent headache was 32.4% in subjects with SCD which were similar to control subjects. However, younger children with SCD were reported to have headaches more frequently than controls [2].

The cause of headache in SCD is multi-factorial just as in general population but SCD-specific factors such as bone infarction, severe anaemia, or frequent opioid medication use may contribute. However, the headache could be a manifestation of serious cerebro-vascular disease [2].

2. Case Report

We report a child with SCD-SC type who presented with headache and was diagnosed with idiopathic intracranial hypertension (IIH).

A 12-year-old girl with SCD-SC presented to the emergency department with eight days history of headache, vomiting, dizziness and diplopia. She was previously well. She never had a sickle cell crisis and routine trans-cranial Doppler study done on an annual basis had been normal. Her medications were folic acid and prophylactic penicillin.

Examination revealed no signs of meningism or encephalitis. Her weight was 72.7 Kgs and height 173.4 cms with a body mass index of 24.4 (96th centile for age). She had bilateral sixth cranial nerve palsy and papillaedema.

She underwent extensive radiological evaluation including MRI, MRV and MRA for possible SCD-related cerebrovascular disease. All the tests were normal. A bilateral enlarged blind spot was noted on visual field mapping.

She underwent a lumbar puncture. The cerebrospinal fluid (CSF) opening pressure was elevated at 60 cms H_2O. The CSF studies were normal. She was diagnosed with idiopathic intracranial hypertension (IIH) and commenced on oral acetazolamide therapy.

A month later, her headache and eye signs had resolved, but she still had persistent enlarged blind spot and papilloedema. She underwent second lumbar puncture and the opening CSF pressure was 33 cms H_2O. The acetazolamide therapy was continued and after a further month, her visual field normalised and papilloedema resolved. After three months of therapy, acetazolamide was weaned over a further six weeks and stopped with no recurrence of headache or eye signs.

3. Discussion

IIH is classified as a headache syndrome characterised by

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1) raised CSF pressure; 2) normal composition of CSF; 3) normal neurological examination (with the exception of papilloedema and sixth cranial nerve palsy); and 4) normal consciousness [Dandy criteria] [3].

The modified Dandy criteria include an additional criterion of normal CT/MRI brain without evidence of thrombosis [4].

The 90th percentile of opening CSF pressure for age group 10 - 14 years is 28 cms of H_2O [5]. Our patient's opening CSF pressure was significantly elevated at 60 cms H_2O.

There are very few reported cases of IIH in a patient with SCD in medical literature. One case of recurrent IIH in a woman with SCD during pregnancy was reported in 1985 [6]. However, there are only two reports of IIH in SCD in childhood involving five children in total [7,8]. None of these five children had any evidence of cerebrovascular disease. Three of them were 15 - 16 years old with high body mass index. The other two cases were under 10 years of age and had normal body mass index. All reported cases responded well to lumbar puncture and acetazolamide which is the standard initial management of IIH [9].

Our patient was a 12-year-old with high body mass index who responded well to the standard management too.

IIH has been reported in several other types of anaemia but our case is amongst the only few reported in children with SCD.

4. Conclusions

When investigating cases of SCD which present with headache, IIH should be part of the differential diagnosis, particularly in teenagers with high body mass index. However, IIH can affect much younger children with normal body mass index, too.

IIH can be caused by an alteration of one or more of the four determinants of cerebrospinal fluid (CSF) pressure: 1) intrasagittal sinus pressure; 2) resistance of arachnoid villi to the egress of CSF; 3) rate of production of CSF, and 4) compliance of the CSF space [10].

The effect of sickle cell disease on one or more of the above remains to be determined.

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