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Acute Idiopathic Thrombocytopaenic Purpura with Cerebellar Haemorrhage: A Case Report

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Summary

Obi GO. Acute Idiopathic Thrombocytopaenic Purpura with Cerebellar Haemorrhage: A Case Report. *Nigerian Journal of Paediatrics*, **1986; 13:87.** The case of a 12-year old school girl who presented with idiopathic thrombocytopaenic purpura, is reported. Therapy with prednisolone was associated with an initial partial response. However, three weeks after discharge from the hospital, she developed intracranial bleeding with typical signs of a cerebellar lesion. She eventually recovered completely from the neurological deficit, which was an unusual development in the natural history of idiopathic thrombocytopaenic purpura.

Introduction

IDIOPATHIC thrombocytopaenic purpura (ITP) represents a spectrum of thrombocytopaenic disorders in which no obvious cause can be detected. Acute and chronic varieties of ITP have been identified, but in recent years, it has become increasingly evident that ITP may be an autoimmune disease. Probably in keeping with the apparent rarity of autoimmune diseases among black people in general,[1 2] ITP is uncommon among patients of negroid stock; some cases have however, been described in Kenya.[3] Furthermore, Wintrobe[4] and Keller[5] have opined that a condition known as *Onyali* which has for some time, been recognised as a distinct clinical entity in East and South Africa,[6] is simply a more severe variant of ITP.

Intracranial bleeding is a rare complication of ITP[5 7-9] and the few reported cases have been mainly cerebral or subarachnoid, and were invariably fatal. We report here, the occurrence of acute ITP complicated by cerebellar bleeding, which was followed by complete neurological recovery, in a 12-year old girl.

Case Report

MO, a 12-year old school girl, presented on October 6, 1983, with bleeding gums, petechiae and bilateral subconjunctival bleeding; there was a history of fever a week earlier. There was no relevant history of drug ingestion. Examination confirmed the bleeding gums, petechiae in the mouth, arms and trunk, but no haemorrhagic bullae. Initial haematological investigations were

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haemoglobin (Hb) 10.2g/dl, packed cell volume (PCV) 0.31; the platelet count was 28 x 10^9/L (28,000/cmm) and leucocyte count 5.9 x 10^9/L (5,900/cmm): the differential count was normal. The lupus erythematosus (LE) test was negative.

The marrow showed an increase in the number of megakaryocytes; a proportion of these were enlarged with well-defined granularity of the cytoplasm but no platelet formation. In others, nuclear bodies were present in the cytoplasm which showed no granularity or platelet formation. A diagnosis of acute idiopathic thrombocytopaenic purpura was made.

Treatment and Course

The patient was commenced on oral prednisolone on October 7, 1983 and transfused with two units of fresh whole blood as facilities for the preparation of platelet concentrates were not available. Some improvement occurred over the following 3 weeks, although slight gum bleeding persisted. However, on October 28, there was a sudden onset of headache, pain in the eyes, vertigo and nausea. Examination at this time showed that the patient was conscious and well orientated, but was ataxic, with a sensation of vertigo. The speech was dysarthric and there was bilateral horizontal nystagmus. By November 18, 1983, the bleeding had stopped, ataxia became more pronounced, and the patient was now confined to bed; the platelet count was 45 x 10^9/L (45,000 cmm). For the next three weeks, there was no improvement in the neurological complications and the patient remained confined to bed because of severe ataxia; both the dysarthria and the nystagmus persisted. As the hospital bills had increased, the patient's relations requested for a discharge, in spite of her condition. She was allowed to go home, on oral prednisolone 30mg daily, but was to report back at the haematology clinic two weeks later. She however, did not attend the clinic until a month later, although she had in the meantime, been taking her medications as directed. She denied attendance at any other hospital or clinic, or the taking of any other medication. Physical examination now revealed complete recovery of the patient with absence of all previous neurological deficits. The Hb was 13.5g/dl, leucocyte count, 6.1 x 10^9/L (6,100/cmm) and platelet count, 98 x 10^9/L (98,000/cmm). The patient has continued to attend the out-patient clinic monthly and has remained quite well.

Discussion

This case satisfied the criteria for the diagnosis of acute ITP, with a history of pyrexia suggesting recent infection a week prior to the onset of bleeding, very low platelet count, typical bone marrow appearance and early remission with steroid therapy[4 7 8]. The exclusion of secondary causes is supported by the absence of splenomegaly, superficial lymph node enlargement and bone tenderness and by the finding of a characteristic bone marrow morphology[9].

What is remarkable about our case is the development of cerebellar signs three weeks after the onset of symptoms and while the patient was still hospitalised. The cerebellar lesion was acute as shown by the classical signs of loss of balance, vertigo, bilateral nystagmus, dysarthria and ataxia[11 12].

Intracranial haemorrhage in ITP although the most serious complication, is comparatively rare [7 8 10 13]. Komrower and Watson [13] reported 4 cases of intracranial haemorrhage in 43 children and in a review of the literature on the disease, they found intracranial bleeding in 19 out of 278 patients. The haemorrhages were mainly subarachnoid and rarely, intracerebral. Choi and McClure[10] observed only 2 cases of cerebral bleeding out of 235 cases of ITP in children. In all the cases described, the occurrence of cerebellar bleeding was most uncommon. Our case must therefore, be a most unusual presentation of ITP, for not only was the haemorrhage cerebellar, it was also followed by complete recovery of the neurological deficit. Another notable feature of our case was the absence of haemorrhagic bullae in the oral mucosa. Haemorrhagic bullae have

been described in East and South African patients in the condition known as *Onyalai*[6]. They appeared to be unique in African patients, but Wintrobe[4] and Koller[5] have observed that haemorrhagic bullae are the result of severe acute thrombocytopaenia rather than a specific feature of any pathogenic form and may in fact, also occur in drug induced thrombocytopaenia.

Immunosuppressive agents have been the main line of treatment of ITP, the principle being the suppression of production of autoantibodies, by the direct killing or inhibition of the proliferation of antigen stimulated lymphocytes[14]. The most often used agents are corticosteroids, purine analogues, vinca alkaloids and the alkylating agent, cyclophosphamide[14 15]. A corticosteroid such as prednisolone, is the drug of choice for initial management because of the rapidity of response; reduction of purpura may even precede a rise in platelet count. Corticosteroids usually control the clinical state until spontaneous cure occurs[14]. Recently, favourable response has been obtained to treatment with vinblastine loaded platelets,[15] plasmapheresis[15] and large doses of intravenous IgG[16 17].

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