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Case Report

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A case of juvenile addisonian pernicious anaemia

Abstract

We present a 26year old juvenile pernicious anaemia with polymyositis and peripheral neuropathy. She had a history of undiagnosed cause of anaemia severe enough to require hospitalisation, blood transfusions at the age of 3-years and a 6-year history of lethargy, severe generalised weakness, months of darkened knuckles and palms

Keywords: dermatomyositis, addisonian pernicious anaemia, autoimmune, paraneoplastic

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Case description

A 26-year-old female patient presented with worsening progressive generalised body weakness, cramps and numbness in extremities, darkening of palms (Figure 1) and soles with darkening and scaling of knuckles, difficulty walking due to mild to severe weakness of legs and lower back ache worsened by walking. She had an unclear history of being unwell in childhood and receiving blood transfusions at the age of 3years although no diagnosis had been offered then. Thereafter she was well until 6years ago when she began to complain of constant lethargy to date. Then 3months prior to admission she continued to worsen and developed darkening of the soles, palms and knuckles followed by more progressive weakness a week ago to a stage where she was struggling to do daily tasks for example combing her hair, getting up from a chair, to even walking .She comes from a middle class family and has a balanced diet and frequent meals, no history of any chronic illness or previous surgery and there was also no family member with a similar clinical picture or history. On examination in addition to the above positive findings were premature greying of hair, conjunctival pallor, tachycardia(112), reduced power across all limbs with a power of 3/5 in both lower limbs and 4/5 in upper limbs and tenderness on palpation across large muscle groups.

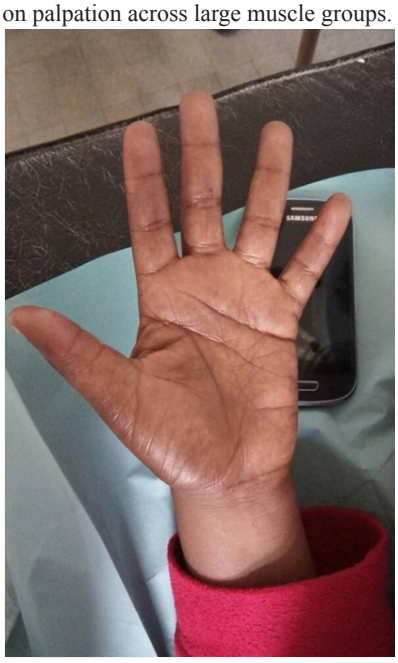

Figure 1 A 26-year-old female patient presented with worsening progressive generalised body weakness, cramps and numbness in extremities, darkening of palms.

Several investigations were done during her 5 day hospitalisation: BMA (hyper cellular bone marrow with mild to moderate megaloblastic changes in all cell lines), SERUM VITAMIN B12 deficient (-98pmol/L)-b12 deficiency as normal range(156-672) ,Intrinsic factor antibodies–positive >1.53AU/Ml ,MUSCLEBIOPSY (no pathology- although only done several days after patient had been on steroids), FBC (HB;10 RBC; 3,41 MCV;89 PLT;127 NEUTRO 37.5% -anisocytosis, normochromic anaemia, combined neutro & thrombocytopenia, SERUM CK; 35iu/L(several days after patient commenced on steroids), CRP;5.0mg/L, RF; NEGETIVE, ESR;26, ENA; NEGETIVE, URIC ACID;0.25mmol/L, TSH:0.80Uiu/Ml, FE;42.1umol/L, CA;2.10 mmol/L, ALB;46g/L, Ba meal: mild gastroesophageal reflux. Electro diagnostic studies: unavailable locally.

We made an impression of juvenile addisonian pernicious anaemia with polymyositis and peripheral neuropathy and commenced her on prednisolone 30mg po od, paracetamol 1g po tds, vitamin B12 injections lifelong, continued on folate 5mg pood from her family Dr, Gradually during the week on treatment the patient improved she regained her strength and was able to get up from a chair, comb her hair, walk a few steps with power going up to 5/5 upper limbs and 4/5 lower limbs and there was remarkable improvement in tenderness across the large muscle groups, back pain ,numbness in her hands and feet and the darkening on her knuckles. She was discharged and reviewed after a month by then she had no complaints at all including the dark palms which have normalised.

Discussion

Pernicious Anaemia is an autoimmune atrophic gastritis that causes a deficiency in Vitamin B12 due to its malabsorption. Vitamin B12 malabsorption is the result of deficiency of intrinsic factor, a protein that promotes its transport to the terminal ileum for absorption. Generally, it is considered as a disease of the elderly as its prevalence is 0.1% in general population and 1.9% in subjects over the age of 60years. Pernicious anaemia in age group below 30 years is less common (<4%).[1] It is thought to be rare in Africa, with the biggest burden of anaemia cases attributed to the neglected tropical diseases (NTDs), Malaria and HIV[2] In our country Zimbabwe however, Vitamin B12 deficiency is the primary cause of megaloblastic anaemia and, contrary to textbook statements, is often due to pernicious anaemia.[3]

We appreciated the importance of clinical findings and history before we received our lab results to help formulate our diagnosis on the patient with a history of unknown anaemia in childhood so severe she needed multiple transfusions and with history of chronic fatigue, dark palms, pallor, premature greying of hair, stocking and

glove neuropathy.[1] It was interesting to note that our patient presented with a condition that is not prevalent in her age group according to epidemiological statistics.[1] The central physiologic principles are that clinically important deficiency is more likely to occur (and progress) when intrinsic factor-driven absorption fails than when diet is poor and that most causes take years to produce clinically obvious deficiency therefore treatment of vitamin B12 deficiency due to malabsorption/PA is parenteral hydroxocobalamin and is life long, with recurrence of symptoms on discontinuation.[4]

It would also be important to highlight that anaemia in childhood so severe to require blood transfusions and to continue into adulthood does need to be followed up and although Pernicious anaemia during childhood and early adulthood is seen in a small subset of individuals hence the diagnosis is often overlooked it still needs to be considered as a differential.

Acknowledgments

None.

Conflict of Interest

The author declares no conflicts of interest.

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