

Terson Syndrome - A Rare Presentation of Acute Lymphoblastic Leukaemia in Children

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Abstract: Acute lymphoblastic leukaemia arising from lymphoid precursor cells of the bone marrow, the lymphoreticular system, and the soft tissue can present with medullary and extramedullary involvement. Extramedullary involvement can affect any organ. Presentation with loss of vision secondary to bilateral vitreous haemorrhage along with intracerebral haemorrhage known as Terson syndrome is rare. There are case reports of such presentations in adults but, only a few in children. Here we are presenting a case of a 3-year-old male child who had bilateral visual loss secondary to vitreous haemorrhage along with intracerebral bleed, who was later diagnosed to have ALL and we want to give a brief overview of the literature about this unusual entity. Rapid investigation and timely initiation of treatment are needed to salvage the eye and the vision. **Conclusion:** Every child with acute lymphoblastic leukaemia with involvement of the brain should have at least one ophthalmologic examination.

Keywords: acute lymphoblastic leukaemia, extramedullary involvement, vitreous haemorrhage, intracerebral bleed, Terson syndrome

1. Introduction

1.1 General background

Leukaemias are the most common malignant neoplasms in childhood, accounting for approximately 31% of all malignancies that occur in children younger than 15 yr. Among them, acute lymphoblastic leukaemia (ALL) accounts for approximately 77% of cases. Peak incidence is at 2-3 yr of age and occurs more in boys than in girls at all ages. The initial presentation of ALL usually is nonspecific and relatively brief. Anorexia, fatigue, malaise, irritability, and intermittent low-grade fever, bone or joint pain, particularly in the lower extremities, may be present. Organ infiltration can cause lymphadenopathy, hepatosplenomegaly, testicular enlargement, or central nervous system (CNS) involvement (cranial neuropathies, headache, seizures). Children with leukemic involvement of the CNS may present with papilloedema, retinal haemorrhages, and cranial nerve palsies. Bilateral vitreous haemorrhage³.

ICH is the second most common cause of mortality in ALL after infection⁴.

1.2 Specific background

Terson syndrome is defined as an intraocular haemorrhage associated with intracranial bleeding. It was first described as an intraretinal haemorrhage following subarachnoid haemorrhage by the French Ophthalmologist Albert Terson, but it was found later that it could occur in any case of intracranial bleed or elevated intracranial pressure and includes an accumulation of subretinal, retinal, preretinal (subhyaloidal), or vitreal blood. It is usually bilateral, and in most cases recovers spontaneously within a few months. Vitrectomy may be necessary in the most severe cases.¹

The pathological mechanism responsible for this intravitreal bleed remains unclear. It is thought that raised intracranial pressure forces blood into the subarachnoid space, along the optic nerve sheath and into the preretinal space. The sudden rise in intracranial pressure may lead to a decrease in venous return to the cavernous sinus or obstruct

the retinochoroidal anastomoses and central retinal vein, resulting in venous stasis and haemorrhage.²

Vitreous haemorrhage occurring in children with acute lymphoblastic leukaemia though rare is an important entity that can't be foreseen. There are only a few cases of ALL presenting with vitreous bleed reported in the literature to date. Along with chemotherapy for ALL, pars plana vitrectomy may save the vision in these children before amblyopia sets in⁶.

2. Case Report

We are presenting a case of a 3-year-old male child born to a non-consanguineous couple, third in birth order, who was admitted with complaints of fever, cough, cold and breathing difficulty for one week. The child was quite well 3 months prior to the onset of this illness, he first started to have frequent falls and was not able to follow objects. There was history of pain in both lower limbs because of which the child was unable to walk 20 days back, for which he was taken to local quacks and native casts were applied. Slowly the child lost interest in playing and would prefer to sit in one place rather than play with his siblings.

3. Examination Findings

On admission, the child was pale, febrile, and was having severe respiratory distress with hepatosplenomegaly and bilateral pedal oedema. His weight was 8 kg which was less than the 3rd centile and his height was 79 cm which was at the 3rd centile. He was not able to follow objects and was able to perceive light only. There were intercostal and subcostal retractions along with hepatosplenomegaly and a few palpable cervical lymph nodes.

Laboratory evaluation

Laboratory evaluation of the child revealed anaemia with Hb of 4 gm/dl. along with thrombocytopenia and leucocytosis. His iron studies were low and even he had low serum folate and B12 levels. In view of severe anaemia and impending CCF, the child was given packed red blood cells and

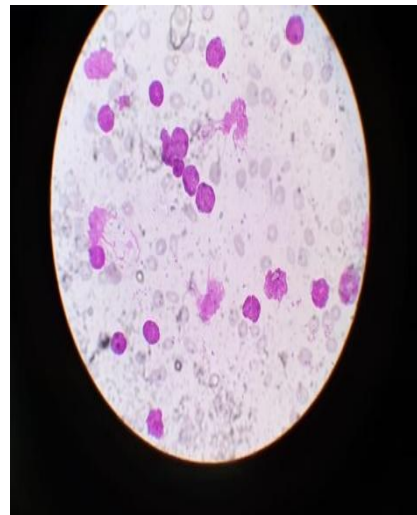
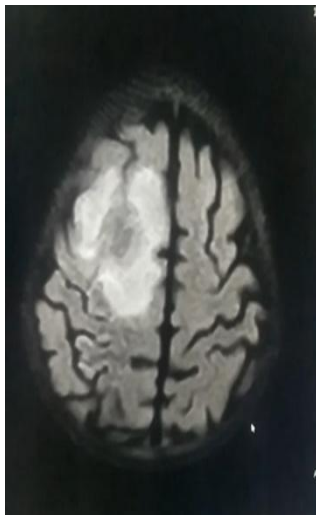
platelets transfusion. At this point in time, the peripheral smear was normal.

Course in the hospital and further follow up

The child was treated for nutritional anaemia and failure to thrive. His ophthalmological examination showed vitreous haemorrhage in both eyes which were suspected to be nutritional in origin so nutritional support was given and the child was discharged. After 20 days the child was readmitted with severe distress, puffiness of the face along with hepatosplenomegaly, and engorged neck veins. The child was anaemic, there was generalised lymphadenopathy. Hemogram showed Hb of 4 gm/dl, platelet counts of 6000, and TLC was 42000. peripheral smear examination showed numerous lymphoblasts. Chest x-ray showed mediastinal widening. There was no improvement in his visual acuity.

He was not able to follow the light. Peripheral smear examination showed numerous lymphoblasts, suggestive of acute lymphoblastic leukaemia. His MRI brain showed intracerebral bleed along with some intracerebral masses. We want to do contrast MRI brain and flow cytometric analysis and bone marrow studies but the due onset of 2nd wave of COVID 19, our hospital was converted to covid care centre after which the parents were denied further stay in the hospital and were discharged against medical advice and were lost to follow up.

Clinical image of the child on the first admission we regret not covering the eyes, its only to show that the child was not able to follow objects (these pictures are being displayed only after taking consent from the parent.)



4. Discussion

Leukaemias being the most common childhood malignancies, we should be familiar with the clinical features which are usually nonspecific and we should have a high index of suspicion in children presenting with fever and lymphocytosis and cytopenias in other cell lines, along with hepatosplenomegaly. Peripheral smear examination is a simple test to look for blast cells. The index child on the first admission was found to have anaemia, thrombocytopenia along with leucocytosis which was wrongly interpreted to be due to infection and the limp the child was complaining

about might be due to bone pains. His peripheral smear examination done during the first admission was after receiving packed cells and also platelet transfusion. So we might have missed the diagnosis on the first admission. When he was readmitted for the second time he had full-blown features of ALL with hepatosplenomegaly with lymphadenopathy after peripheral smear examination we came to the conclusion of ALL in this child with bilateral vitreous haemorrhage and intracranial bleed as the first presentation which is termed as Tersons syndrome. We regret that we don't have the images of vitreous haemorrhage as that was covid pandemic time.

Haematological disorders such as leukaemia may be complicated by haemorrhage. Ocular tissues can also be involved, causing bleed in the anterior chamber known as hyphaema, vitreous haemorrhage, or retinal haemorrhage. Management of Terson syndrome depends on the position of the intraocular haemorrhage. For a vitreous or subhyaloid haemorrhage, treatment could be conservative, we can have a periodic observation for spontaneous resorption if that is not compromising the vision or more aggressive that is pars plana vitrectomy if there is visual compromise. There are reports showing spontaneous improvement in adults within a few months after the acute event, but there are no such reports in children as this Terson syndrome by itself is rare. Therefore some authors prefer to delay vitrectomy and wait for spontaneous resolution and is considered only in severe or bilateral cases that do not show visual improvement.⁵

5. Conclusion

All being the most common malignancy of childhood, all the children admitted with anaemia, thrombocytopenia with leucocytosis should have a peripheral smear examination and a bone marrow evaluation to diagnose them at the earliest. As this case taught us every child diagnosed with childhood ALL should have at least one ophthalmological evaluation as Terson syndrome is one of the sight-threatening complications of ALL though it is a rare presentation of leukaemia. At the same time, it's a treatable cause of blindness in children with ALL.

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