

# Spontaneous Suprachoroidal Hemorrhage in a Thirteen-year Old Child with Thrombocytopenic Purpura

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**Abstract** We report a paediatric idiopathic thrombocytopenia purpura (ITP) patient with primary spontaneous suprachoroidal haemorrhage (SSCH). The patient was diagnosed of ITP based on platelets count and morphology. Eye ultrasound indicated SSCH with total choroidal detachment. After improvement of the patient platelet count using immunoglobulin treatment, two surgical vitrectomy were performed on the patient with 3-weeks interval. After six months-follow up, the patient revealed normal IOP and hand motion vision in the left eye. We recommend that ITP should be considered as one of the risk factors for vision threatening ocular haemorrhages even for paediatric patients.

**Keywords:** *idiopathic thrombocytopenia purpura, surgical drainage, total choroidal detachment*

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## 1. Introduction

Idiopathic thrombocytopenia purpura (ITP) is an autoimmune defect characterized by production of antibodies attacking the platelet surface antigens, usually presenting with subcutaneous or mucosal bleeding [1]. Haemorrhage in ITP is extremely variable with regard to body site and clinical severity [1]. Ophthalmic involvement is very rare [2]. We report the first case of spontaneous suprachoroidal haemorrhage (SSCH) in a patient with ITP. Moreover, this is the youngest reported patient to have SSCH without any systemic or ocular risk factors and without being on any anticoagulation therapy.

There are two types of SCH, surgical and spontaneous. Surgical SCH has been described in literatures very frequently. But the spontaneous SCH (SSCH) is extremely rare condition and only described in case reports or case series. It was reported mostly in old age patients on anti-coagulants, thrombolysis therapy, age related macular degeneration, acute angle closure glaucoma, and decompensated liver disease [3,4]. To our knowledge, it has never been reported to occur in a paediatric case or a patient with thrombocytopenic purpura without any associated ocular pathology or injury.

## 2. Case Report

Thirteen-year old boy came to the ER with history of bruises all over the body, redness in left eye and epistaxis for two weeks. There was no history of any such problem earlier, though, he had fever reaching 39 degree Celsius and upper respiratory tract infection three weeks prior to the presentation date. He did not give any history of lower or upper GI bleeding, change in urine colour or history of trauma. There was no history of eye pain, discharge or eye trauma. The patient is known asthmatic, with one ER visit 6 months prior to this presentation. No previous hospital admissions or surgical operations. The patient has not been on any medications at home. Family history was unremarkable for haematological diseases.

Prothrombin time, partial thromboplastin time and international normalized ratio (INR) were normal. Blood smear showed thrombocytopenia with large platelets and presence of reactive lymphocytes. The complete blood picture showed low platelet count ( $10 \times 10^9/L$ ) in the first day. On the basis of clinical findings, he was diagnosed by a haematologist as idiopathic thrombocytopenia purpura.



**Figure 1.** Ophthalmic ultrasound when the case was first introduced showing 360 degree bullous kissing haemorrhagic choroidal detachment

On the second day of admission, ophthalmology consultation was asked for. On examination, vision was light perception in the left eye. He had left sub-conjunctival haemorrhage, left relative afferent pupillary defect (RAPD), quiet and deep anterior chamber and absent red reflex. IOP was 34. There was no fundus view, and ultrasound showed 360 degree bullous kissing choroidal detachment (Figure 1). The patient was diagnosed as SCH with total choroidal detachment. The case was discussed with the haematology consultant and it was planned to start him on systemic cortisone along with intravenous immunoglobulin (IVIG). Patient was given two doses of IVIG, and CBC in the next day showed some improvement, although still very low ( $49 \times 10^9/L$ ). The patient did not require transfusion of platelets, since his platelet count kept on improving, and reached normal level on the fourth day of admission. Topical anti-glaucoma medications (i.e. dorzolamide/timolol, brimonidine tartrate, and bimatoprost) and systemic acetazolamide was also started. Surgical drainage was planned 4 days from the admission date (i.e. once the platelet count was stabilized).

Surgical drainage of SCH was done on the planned date. The patient was discharged the next day on 60 mg of oral prednisolone, along with moxifloxacin ophthalmic solution, prednisolone 1% ophthalmic solution and omeprazole 20mg capsule orally for weeks. And patient was booked for follow up surgery three weeks later.

On first follow up one week after the surgical drainage, the vision was light perception and IOP was 14 in the left eye.

Three weeks after the discharge patient came for the second planned surgery. The procedure performed

was 23 gauge pars plana vitrectomy, inferior retinectomy, air fluid exchange, endolaser and injection of 1000 centistoke silicone oil in the left eye with encircling scleral buckle. On the first post-operative day, patient had hyphema, IOP was 25, and the patient was put on antiglaucoma medications along with the post-operative steroid and antibiotics.

Two weeks after the second surgery, vision was Hand motion (HM) in left eye, IOP was 15mmHg, and early cataract had formed. Fundus details were not clear. There was organized vitreous haemorrhage and peripheral retina was flat. Patient was continued on tapering systemic and topical steroids with anti-glaucoma medications. Six weeks after the second procedure, on follow-up, vision remained HM, IOP was 12 mmHg. On fundus exam there was sub-retinal fibrosis and atrophic retina.

Same visual acuity and IOP was recorded three months after the surgery. Pupil was dilated with posterior synechia, and fundus examination showed chronic tractional retinal detachment under silicon oil. Systemic prednisolone was tapered and prednisolone eye drops were continued BID. The patient was given follow-up 3 months later (i.e. six months after the surgical procedure).

Six months after the surgery, the vision in the left eye did not change, IOP was 12mmHg, eye exam revealed white and quiet anterior segment, with dilated pupil, and fundus exam showed chronic retinal detachment with silicon oil and fibrosis.

Meanwhile patient continued follow-up with haematology with monthly CBC. His haematological condition remained stable.

### 3. Discussion

Idiopathic thrombocytopenia purpura (ITP) is an autoimmune haematological disease characterized by decreased platelet count with normal bone marrow and absence of other causes of thrombocytopenia. There are circulating antibodies directed against platelets surface antigens causing their destructions and thus decreasing their numbers [1]. It affects both adults and children. Children usually develop this after a viral infection and recover fully without treatment while adults usually develop chronic course. ITP commonly manifest as bleeding from the skin, mucosal membranes, GIT, CNS and vagina [2]. Normal platelet count is 150,000-400,000/ $\mu$ L. The count ordinarily must be reduced below 50,000/ $\mu$ L before untoward bleeding is observed, even that usually does not occur without trauma. Spontaneous bleeding is unlikely unless the platelet count is below 20,000/ $\mu$ L. Despite of having low platelet count (<20,000/ $\mu$ L), the great majority of children with ITP do not have major bleeding at diagnosis [2]. Risk of bleeding increases as the platelet count decreases.

In medical literatures, very little has been written about ocular complications in children or adults with ITP. There are few case reports in this regard [2]. Case reports of ocular haemorrhages (sub conjunctival, retinal and vitreous) as first presentation of ITP have also been reported [3]. Two case reports have described retinal haemorrhage at diagnosis or shortly thereafter in the absence of previous trauma occurring prior to or concurrently with intracranial and vitreous haemorrhage [5]. After extensive review of medical literatures we could not find any report of SSCH in patients with ITP. The only reported SCH case report of in a patient of ITP was introduced by Srinivasan et al. [4] and it was a postoperative expulsive haemorrhage for an old patient [4].

SSCH is extremely rare event, described only in isolated case reports. In patients with this occurrence, systemic or ocular risk factors or a history of systemic anticoagulation has been described [6]. Decompensated liver disease with resultant coagulopathy has also been described as a risk factor for ocular bleed [2]. The ocular risk factors include glaucoma, aphakia, axial myopia, age related macular degeneration and inflammation. Other associations of SSCH are systemic hypertension, increasing age, atherosclerosis and significant vascular diseases. The fragility of choroidal and ciliary vasculature is believed to be increased in presence of these factors, making them susceptible to rupture [7].

Case reports have described spontaneous hyphemas and sub conjunctival, suprachoroidal, vitreous, subretinal and orbital haemorrhage with the use of tissue plasminogen activator, streptokinase, low molecular weight heparin and warfarin, especially in cases with associated ocular and systemic risk factors [8,9].

In almost all the previously reported cases of SSCH, the patients were of old age. They had ocular or systemic risk factors or they were on systemic anticoagulation for some reason. We reviewed 31 reported cases of spontaneous SCH with age ranging from 24 - 92 years. The mean age is 66.13. The youngest reported case of SSCH was for a 24-year old female with cystic fibrosis and diabetes [9].

She was on warfarin for 2 months.

This case is considered unique as it is the first SSCH case reported in paediatric patient, as a SSCH case with no history of any cardiac, vascular or ophthalmic disease, and a patient was not on any anticoagulant therapy. And, to the best of our knowledge, this is the first report of SSCH in an ITP patient.

The presentation of SSCH patients tend to be with eye pain or loss of vision. The presentation of our case was totally different as he presented with sub conjunctival haemorrhage only. There was no complaint of pain though his intraocular pressure was high. Usually patients with SSCH shows shallow anterior chamber (AC), in contrary our patient had a deep AC. Chak and Williamson [10] also presented a case of SSCH with deep AC. Our patient also didn't complain of loss of vision at presentation. While in most of the cases, patients presented with loss of vision associated with or without pain.

The visual prognosis for patients with SSCH is quite predictably frustrating, common for most of patients and depends on the severity of SCH.

### 4. Conclusion

Poor prognosis and frustrating outcome of SSCH makes it important to know the possible risk factors and associations of this condition, so as to prevent further complications. Ocular involvement in ITP is not very common, but this might be due to the fact that there is not much written about it in literature. It should be considered as one of the risk factors for vision threatening ocular haemorrhages. Moreover, unexplained ocular haemorrhages prompt the need to investigate the life threatening systemic haematological conditions.

### Conflict of Interest

All authors declare no conflict of interest to disclose.

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