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# Nasal pseudotumour in a haemophilic: A case report

Sulochana B, Balakrishna, Kailesh Pujary, Annamma Kurien, Anjalin D'Souza, Roseminu Varghese\*

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## Abstract

Haemophilia is an X linked inherited coagulation disorder. A haemophilic pseudotumour is one of the rarer complications of haemophilia, which results from repetitive bleeding. These have become scarcer over the years with better treatment facilities for bleeding disorders like factor replacement (Pakala, Thomas, & Philip, 2012). The present case report details the history and clinical course of a 23-year-old case with recurrent nasal bleed and nasal pseudotumour, which showed a positive outcome after surgical intervention and factor replacement. At 3-month follow-up, the patient was evaluated by endoscopy, CT scan, and there was no evidence of recurrence.

**Key words:** Nursing management, nasal pseudotumour, haemophilia A

## Background

Congenital bleeding disorders are caused by dysfunction or deficiency of coagulation factors. The most common congenital bleeding disorders are caused by the lack of factor VIII in haemophilia A and lack of factor IX in haemophilia B. Spontaneous bleeding into joints and muscles and haemorrhage is a feature of severe haemophilia (Paola Coppola et al., 2010) but does not prevent the progressive joint deterioration leading to crippling hemophilic arthropathy. Therefore, primary prophylaxis, ie, regular infusion of concentrates started

after the first joint bleed and/or before the age of two years, is now recognized as first-line treatment in children with severe hemophilia. Secondary prophylaxis, whenever started, aims to avoid (or delay. Nasal pseudotumour is a rare complication in haemophiliacs with an incidence of 1-2% (Purkait, Mukherjee, Naskar, & Bhadra, 2013). The most common basis for pseudotumour is due to repeated bleeding at the fracture bone or sub periosteal space and bleeding into the soft tissue. The haemophilic pseudotumours are encapsulated with necrotized tissue and blood with clot due to inadequate resorption of extravagated blood from frequent bleeding episodes (Rijal, Neogi, Ansari, Khan, & Yadav, 2010). The current case reported here was showing recurrent nasal bleed and was not responding to conservative management and therefore was posted for surgical removal of the pseudotumour.

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## Case report

### Case presentation

A 23-year-old male, (54 kg weight) diagnosed to have Haemophilia A (F VIII <1%) presented with unprovoked, frequent episodes of nasal bleed more on the right side than on the left for a year. Initially, 10 to 20 drops of bleeding on exertion was noted and later

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progressed to 10 to 20 drops of blood spontaneously. Each episode lasted for one minute. On observation, 2-3 episodes were noted in a week, with the last episode noted two weeks earlier. History of haematemesis and melena were also reported. Patient informed a history of nose block more on the right side than on the left for one year, which was insidious in onset and gradually progressive. Patient also reported history of mucoid and blood tinged discharge. He experienced complete absence of smell perception for eight months, which was insidious. No history of altered smell or taste perception and fever was reported prior to the onset of symptoms. He had a history of prominence of the right eye and blurred vision for six months. There was no history of diplopia, use of spectacles, or disturbance in colour vision. He also reported right Iliopsoas haematoma and haemarthrosis of left knee and elbow for which he was been treated with, on demand factor infusion in the past.

#### Physical examination

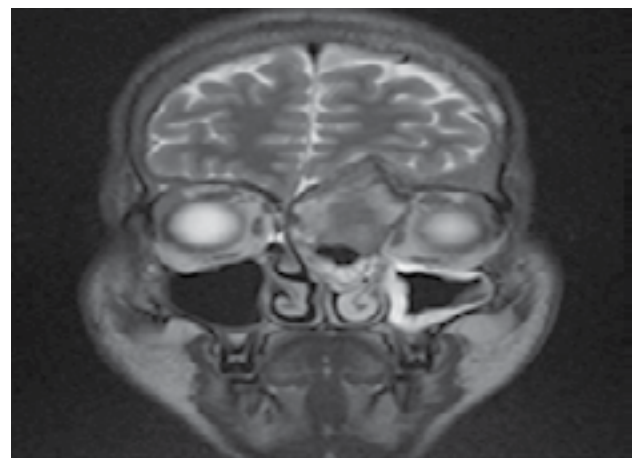
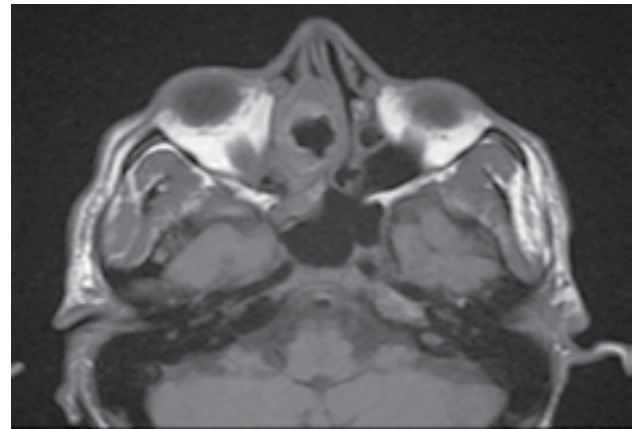
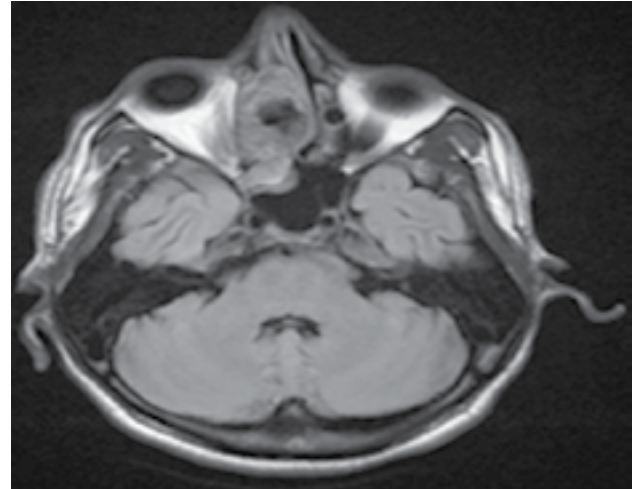
On physical examination, patient's Glasgow Coma Scale (GCS) score was 15, bilateral equally reacting pupils, full extra ocular eye movements and blurring of vision was present on the right eye. Examination of the nose showed normal external nasal framework, vestibule, and columella. Anterior rhinoscopy revealed mild deviation of septum to left. Bilateral middle turbinate was edematous and right polyp was present between the middle turbinate and septum. Posterior rhinoscopy was not performed as the patient was gagging and was not co-operative. Oral cavity and bilateral palatal movements were normal and gag reflex was present. On inspection, right eye was pushed forward and lateral. Visual field, corneal, and conjunctival reflex was normal. Bilateral carotid pulsations were felt and trachea was centrally located. On examination, bilateral external ears were found to be normal and tympanic membrane was intact. Cranial nerves II, III, IV, VI, VII, and IX were also found to be normal.

#### Lab reports

Admission laboratory reports were significant for Hb of 6.0 gm/dl and Hct of 24.4%. Activated partial thromboplastin time (aPTT) was 85.6 seconds (control 27.5s); Factor VIII assay was less than 1% (ref range: 50-150%). Inhibitor test screen was negative.

#### Radiological examination

CT brain was suggestive of intra cranial, frontal space occupied lesion extending in to the nasal cavity and Esthesioneuroblastoma (Figure 1). MRI scan showed a lesion involving right stenoid cavity, infiltrating right ethmoid, and frontal sinuses with possible extra cranial extension in to the right orbit.



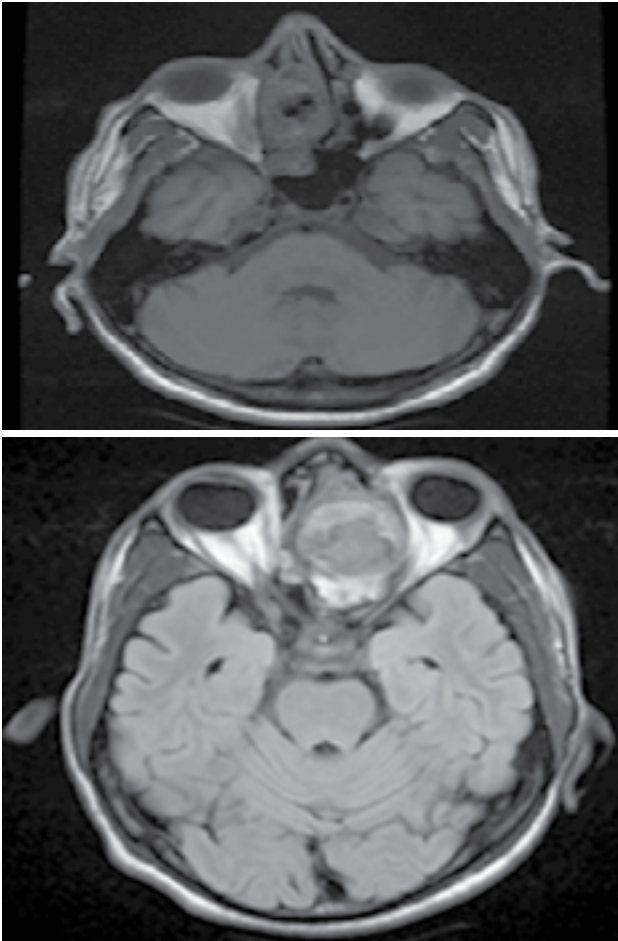


Figure 1: CT of brain before excision of sino nasal mass

### Surgical procedure

Endoscopic excision of sino-nasal mass was performed for two hours and 45 mins under general anesthesia. An encapsulated lesion in the right fronto ethmoidal sinus region extending into the right nasal cavity was found intra-operatively. The presence of polypoidal, mucoid discharge was present in the right maxillary and right frontal sinus. Cystic cavity filled with blood clots were excised piece meal and was sent for frozen section. Frozen section suggested a benign lesion with no evidence of malignancy. Right concha bullosa, tumor adjacent to dura, tissue from roof of ethmoid were sent for histopathological examination. The final histopathology report was suggestive of benign organized haematoma.

### Nursing management

With adequate pre- and post-operative monitoring of the clotting factors, nursing care, and timely replacement therapy, haemophilic patients will have a conservative

prognosis. Better clinical outcomes are achieved, only if factor concentrates are readily available and if the symptoms for bleeding are monitored. The main goal is to prevent bleeding intra-operatively and post-operatively (McDaniel, 2013).

Patient was transfused with blood, fresh frozen plasma (FFP), and factor VIII for management of symptoms on admission as per the lab values and started on Ferrous Sulphate and Multivitamins. Nurses were trained for the proper infusion of the clotting factor and close monitoring of the patient for bleeding symptoms. A care plan was developed to provide individualized care. The patient was started with factor VIII infusion pre-operatively based on the protocol and has been continued for a week i.e. 20 IU/kg bd. Pre-op health education was provided to the patient to ensure co-operation of the patient and its family members. During the surgery, around 200 ml of blood loss was reported. Patient was put on central venous catheter with ultrasound assistance to prevent multiple pricks and potential central venous pressure (CVP) inversion complications and underlying surgery related complications. Patient was observed for cerebrospinal fluid (CSF) leakage during the post-operative phase, because it is one of the complications, as the tumor was extended into extra cranial space. Immediate post-operative period was uneventful. Patient was on antibiotics, proton pump inhibitors, clear nose drops, anticonvulsants, anti-emetics, analgesics, antihistamines, and clotting factor VIII; 2000 IU slow IV over 15 mins every 12th hourly. After which, patient was continued with clotting factor VIII weekly twice 20 IU/kg.

### Follow up

Patient was followed up for three months. On follow-up, patient inhibitor status was tested and were within normal range. There were no bleeding episodes reported. At three months CT scans (Figure 2) were repeated. There was no evidence of recurrence of the pseudotumour and there was no sign of herniation into the cavity.



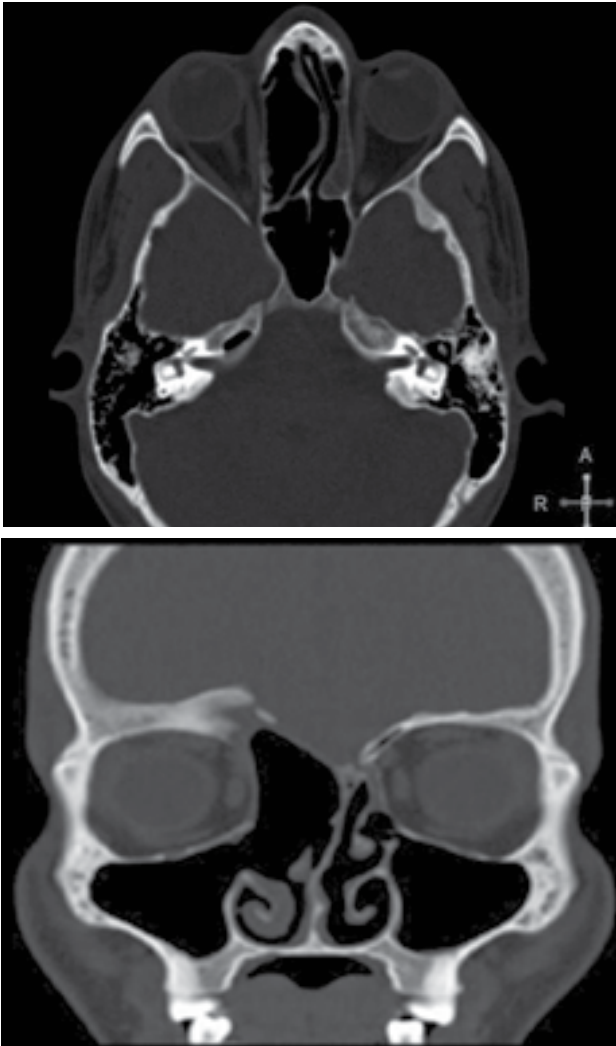


Figure 2: CT of brain after four weeks of surgery

## Discussion

Haemophilic pseudotumour is an uncommon complication of factor VIII and IX deficiencies in the coagulation cascade and occurs in bones and soft tissues. A pseudotumour consists of chronically encapsulated blood collection due to recurrent extra articular haemorrhage. As the swelling progresses, increasing pressure leads to the slow destruction of adjacent structures (Karunanithi, Sethi, Reddy, and Vivekanandam, 2009). The pseudotumours are dangerous and give rise to haemophilia complications occurring 1-2% of persons with haemophilia. They are chronic and slow growing resulting into repetitive bleeding, which is surrounded by thick fibrous capsule, in the centre presence of haematoma (Kapoor, Sastri, Malhotra, Kumar, and Singh, 2006). Specialized

nursing practice in less resource constraint countries is an essential need, when we take care of patients with chronic diseases like haemophilia and other bleeding disorders. As the tumor increases in its size, it will extend to the adjacent structure and causes compression (Stafford, James, Allen, & Dixon, 2003). Needle biopsies are avoided to prevent infection, fistula formation, and haemorrhage. Radiological investigations like CT and MRI are the significant assessment tools pre-operatively to understand the possible involvement to take a decision on surgical approach (Heaton et al., 2000).

As the clotting factor concentration are not easily available in circumstances like surgical interventions, coordinated care is obligated. Nurses play a pivotal role in the management of patients with haemophilia in terms of education, counseling, and coordinating care with health care team members. When taking care of patients with chronic disorders, it is essential to create enhanced patient-provider relationship for effectiveness. Advanced nursing role provides an opportunity to assess patient's needs to improve and explore patient's medical history and perform focused physical examinations. At the completion of assessments, the management goals are identified and plans of care can be established effectively.

## Conclusion

Individualized patient care is important for better patient outcomes. Nurses caring for patients with haemophilia should keep in mind the potential post-operative complications. Specific observations should be made to evaluate any signs and symptoms of bleeding. Factor replacement therapy and post-surgical follow-up will provide better prognosis and collaborative approach will enhance better patient outcome.

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