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Case Series

Otorhinolaryngological manifestations of haematological disorders: a case series of five patients

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ABSTRACT

Haematological disorders frequently present with orofacial and ear, nose, and throat (ENT) manifestations, which may serve as the first clinical clue to an underlying condition. Recognizing these signs is essential for timely diagnosis and management. This paper presents a case series of five paediatric patients (aged 4-17 years) who presented with head and neck manifestations of previously undiagnosed or known haematological diseases, including postoperative bleeding, post-traumatic bleeding and spontaneous haemorrhage. Cases included mild haemophilia A diagnosed after adenotonsillectomy (case 1), post traumatic Christmas disease (factor IX deficiency) with extensive neck haematoma (case 2), known haemophilia A with refractory epistaxis requiring embolization (case 3), von Willebrand factor deficiency presenting as spontaneous bilateral epistaxis (case 4), and severe haemophilia A diagnosed after tongue laceration repair (case 5). All cases were successfully managed with factor replacement, fresh frozen plasma, desmopressin, or recombinant von Willebrand factor. Persistent, recurrent or unusual ENT symptoms- particularly when accompanied by unexplained bleeding- should prompt haematological evaluation. A multidisciplinary approach is critical for accurate diagnosis and improved outcomes in this patient population.

Keywords: Haematological diseases, Epistaxis, Coagulopathy, Postoperative bleeding, Multidisciplinary approach

INTRODUCTION

A broad spectrum of haematological conditions seen in internal medicine present with manifestations in the oral cavity and facial area. Recognizing these orofacial signs is particularly important because they may serve as the initial clinical clue that points to an underlying haematological disorder. Disorders affecting the erythroid, lymphoid and megakaryocytic lineages in the bone marrow, along with immune system deficiencies, coagulation abnormalities and human immunodeficiency virus infection, have been extensively documented to involve the orofacial region.[1] Some children with inherited bleeding disorders may not show symptoms until later in childhood, when they are exposed to situations that test their blood's ability to clot, such as surgery, dental procedures, menstruation. Without these challenges, the bleeding disorder might remain undiagnosed. In children with bleeding disorders, bleeding that occurs at a young age can be particularly significant, potentially indicating a more severe underlying condition.[2] Significant bleeding in a patient with no known congenital bleeding disorder can stem from various factors. They may include the use or overdose of anticoagulant medications, acquired vascular or platelet disorders (such as autoimmune thrombocytopenia), or an acquired deficiency in clotting factors. Prompt differential diagnosis and targeted treatment are frequently essential. Acquired deficiencies in clotting factors often result from inhibitory autoantibodies that impair the function or accelerate the clearance of proteins involved in different stages of the coagulation cascade. These antibodies most commonly target factor VIII, leading to acquired haemophilia A. Such cases may arise in individuals with other autoimmune conditions, those with malignancies, or during pregnancy.

The clinical presentation can vary widely, ranging from mild or no bleeding to severe, life-threatening haemorrhages. Inhibitors against clotting factors-excluding those targeting von Willebrand factor or factor XIII - typically result in prolonged screening coagulation tests, such as the activated partial thromboplastin time (aPTT) and/or prothrombin time (PT).[3]

CASE SERIES

We present five patients showing haematological manifestation in head and neck region presented postoperatively, two post traumatic and two unprovoked.

Case 1

A 7 years old girl presented to our department with profuse nasal and mouth bleed. She underwent adenotonsillectomy in a private setup and presented to us on post-op day 5. She was immediately started on haemocoagulase infusion and anterior nasal packing was tried but the haemorrhage could not be controlled. Complete hemogram, blood grouping, coagulation profile, bleeding and clotting time was done. Her haemoglobin was low (8 gm/dl), activated partial thromboplastin time (APTT) raised (51.2) and platelet count was normal. Haematology team was consulted and as per their advice, 4 units fresh frozen plasma (FFP) and 1 unit blood was transfused and injection vitamin K was given. Blood was sent for factor VIII and IX and meanwhile, she was given posterior nasal packing and bleeding was controlled. Her factor VIII level was found to be low (24). She was diagnosed to have mild Haemophilia A and got 1 dose of recombinant factor VIII injection on post op day 2 and another later after 15 days. Posterior nasal pack was removed after 72 hrs and she was discharged after 2 weeks. She was followed up after 10 days of discharge and a diagnostic nasal endoscopy was done which was normal. Her later post op visits were uneventful.

Case 2

An 8-year-old boy presented in the casualty department with swelling in the neck extending cranially from submandibular region anteriorly and occipital line posteriorly and caudally extending up to the first intercostal space and upper third of sternum anteriorly to suprascapular region posteriorly with associated ecchymosis. On oral cavity examination, there was bluish discoloration over bilateral anterior pillars. The patient also presented with hoarseness. On detailed history from the patient's father, he gave history of blunt trauma by a bamboo. The child had no previous history of any bleeding manifestations or did he have and family history. He was not on any previous medications. The patient was kept nil per oral and injectable antibiotics and steroid was started and routine blood investigations were sent. The patient had low haemoglobin (8 g/dl), raised total count (13400); raised APTT (61.6), while platelet count, APTT and international normalized ratio (INR) were normal. The child was started on injection vitamin K and sent for contrast-enhanced computed tomography (CECT) neck and blood was sent for factor VIII and IX analysis. CECT neck revealed post traumatic soft tissue oedema and haematoma in the neck up to superior mediastinum at the aortic arch level displacing and compressing the great vessels of neck, thyroid gland and larynx towards right side. Factor IX level was found to be reduced (6.69%) with normal factor VIII level. Paediatric haematology consultation was taken. The patient was diagnosed with Christmas disease (factor IX deficiency) and he was transfused with one unit of PRBC, 8 units of FFP and 3 doses of recombinant factor IX. The child clinically improved and APTT value came down to 30.1. The patient was kept under observation for another 3 days and then discharged. The follow-up of the patient was uneventful and coagulation profile was within normal limit.

Case 3

A 6-year-old child presented to the casualty with continuous and profuse left nostril bleed following alleged history of fall in school. On taking proper history, it was revealed that he was a known case of Haemophilia A and was on factor VIII supplementation. He was on injection Emicizumab for 2 years every 2 weekly. He was started on haemostatic agents and routine blood investigations were sent. As bleeding did not stop, anterior nasal packing with foley's catheter and merocele was inserted in bilateral nostril. The bleeding then stopped. Blood test revealed low haemoglobin, normal platelet count and PT and INR, while the APTT level was raised. Bleeding time (BT) and clotting time (CT) were within normal limit. The child was given recombinant factor VIII and after 3 days, pack removal was done and endoscopic nasal examination performed under general anaesthesia. On nasal endoscopy, extensive maceration of nasal mucosa involving anterior third of right nasal cavity and anterior two thirds of left nasal cavity was found along with bleeding from medial to left middle turbinate. Haemostasis was achieved with FloSeal and Wellnase nasal pack. On post op day 6, embolization of left sphenopalatine artery was done by interventional radiology with blood transfusion prior to the procedure. Following embolization, he continued to have minimal oozing, for which factor VIII was given that evening and on next morning. He was discharged on stable condition on post op day 11 in stable condition. The follow-up of the patient was uneventful.

Case 4

A 17-year-old male presented to ENT outpatient department (OPD) with spontaneous, profuse, non-traumatic bleeding from bilateral nostril on and off for 10 days. Suspecting for angiofibroma, the patient was admitted and further workup done. Hemogram showed low haemoglobin, APTT and INR was normal. PT was raised. Medicine consultation was taken and on advice von Willebrand factor was done which was found to be deficient. Haematology department started him on

desmopressin and recombinant von Willebrand factor (vWF). The patient was discharged and followed up. The follow-up period was uneventful.

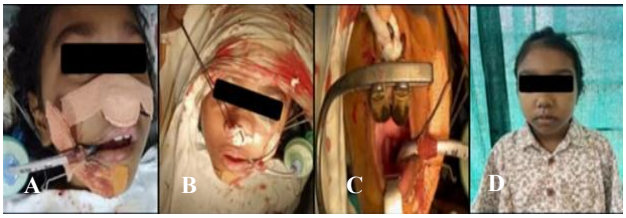

Figure 1 (A-D): Intraoperative pictures where posterior nasal packing was given.

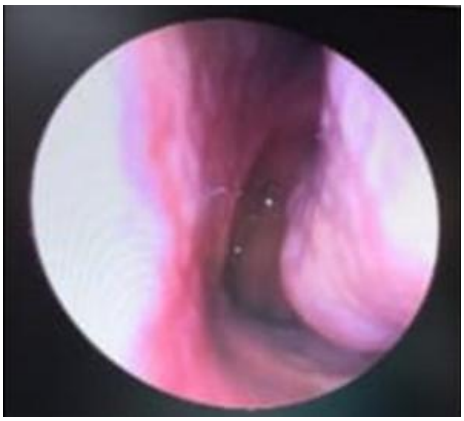

Figure 2: Diagnostic nasal endoscopy done 10 days after discharge.

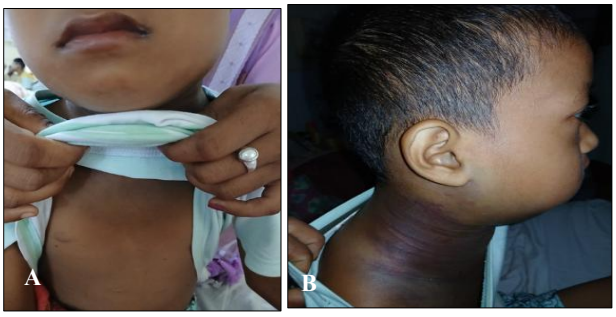

Figure 3 (A and B): 4-year-old boy.

Case 5

A 4-year-old boy, came to emergency room (ER) with bleeding oral cavity for 2 days. His parents gave history of fall 2 weeks back where there were tongue laceration and wound were repaired in a private hospital. Hemocoagulase infusion was started and hemogram was sent. Secondary wound repair was done under general anaesthesia. His Hb was 6 and APTT was 79.1. medicine consultation was taken and 1-unit FFP and 135 ml PRBC was transfused according to body weight. As advised by haematology, factor VIII, IX, XIwas sent and factor VIII came out to be (<0.7) and patient was diagnosed to be a severe case of haemophilia A. Hb and APTT was repeated and it came out to be 11 and 73.8 respectively. The child was on recombinant factor VIII injection and he improved clinically by the end of 2 weeks. The patient was followed up regularly and no complications was found.

Figure 4: Foley's catheter given as posterior nasal packing in emergency room.

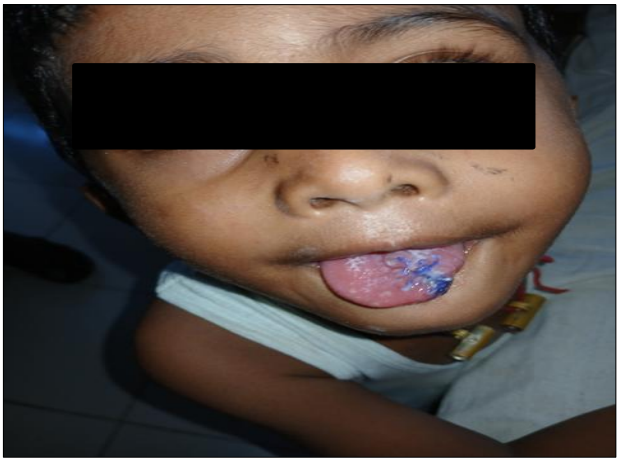

Figure 5: Postoperative visual after repair of tongue laceration and receiving 1 dose of recombinant factor VIII.

DISCUSSION

The five cases presented in this series underscore the critical role of otorhinolaryngologists in recognizing the diverse presentations of underlying haematological disorders. As highlighted in the introduction, otorhinolaryngological manifestations may serve as the initial clinical clue to an undiagnosed bleeding diathesis,

or they may present as refractory complications in known haematological conditions. Our case series demonstrates a spectrum of presentations ranging from post-surgical and post-traumatic haemorrhage to spontaneous, life-threatening epistaxis, requiring varying degrees of medical, surgical and interventional management.

Unmasking inherited bleeding disorders: the role of trauma and surgery

Cases 1, 2 and 5 illustrate a classic paradigm in paediatric haematology: previously asymptomatic children presenting with disproportionate bleeding following routine surgical or traumatic challenges.

Case 1 (mild haemophilia A post adenotonsillectomy) and case 5 (severe haemophilia A post tongue laceration repair) highlight the necessity of maintaining a high index of suspicion for coagulopathies in children presenting with prolonged postoperative bleeding. In case 5, the severity of the presentation was marked by a haemoglobin drop to 6 g/dl and a prolonged aPTT of 79.1. The diagnosis of severe haemophilia A was confirmed by a factor VIII level of <0.7%. The child required aggressive initial resuscitation with 1 unit of FFP and 135 ml of PRBC before transitioning to recombinant factor VIII therapy, which led to clinical improvement over two weeks.

Case 2 is particularly instructive, the 8-year-old boy developed an extensive neck hematoma extending to the superior mediastinum following blunt trauma. Laboratory findings of a prolonged apTT (61.6) with normal PT/INR and platelets, coupled with a factor IX level of 6.69%, confirmed Christmas disease (factor IX deficiency). This aligns with the literature stating that without surgical or traumatic challenges, mild to moderate inherited bleeding disorders may remain undiagnosed until later in childhood.[1]

Epistaxis as a sentinel sign: diagnostic and management challenges

Refractory or spontaneous epistaxis can be a prominent manifestation of both inherited and acquired coagulation disorders.

Case 4 emphasizes the importance of considering coagulopathies in the differential diagnosis of spontaneous, profuse epistaxis in adolescents. Initially suspected of having a juvenile nasopharyngeal angiofibroma, further workup revealed a prolonged PT and deficient von Willebrand, underscoring the utility of standardized bleeding questionnaires.[2]

Case 3 demonstrates the extreme management complexities in a known haemophilia A patient on Emicizumab who presented with refractory unilateral epistaxis following a fall. Despite factor VIII supplementation and initial nasal packing, bleeding persisted. Nasal endoscopy revealed extensive maceration of the nasal mucosa involving the anterior two-thirds of the nasal cavity. Haemostasis was initially attempted with FloSeal and a Wellnase nasal pack, but due to persistent bleeding, the patient required embolization of the left sphenopalatine artery on post-operative day 6. Even after embolization, minimal oozing necessitated further Factor VIII administration, with the patient finally discharged on post-operative day 11. This case highlights that breakthrough bleeding can occur despite modern prophylactic regimens, necessitating a multidisciplinary approach involving haematology, otolaryngology and interventional radiology. This is consistent with Abdel Wahab et al, who noted that recurrent epistaxis in children warrants a thorough investigation for underlying coagulopathy.[4]

Diagnostic evaluation and laboratory correlates

The diagnostic journey across our cases underscores the importance of a stepwise laboratory evaluation. Prolonged aPTT was the hallmark in cases 2, 3, and 5, pointing towards intrinsic pathway deficiencies (factors VIII and IX). Conversely, case 4 presented with prolonged PT and normal aPTT which prompted a specific evaluation for von Willebrand disease. Prompt haematological consultation and targeted factor assays (factor VIII, IX and von Willebrand factor) were pivotal in confirming the diagnosis. This reinforces the importance of evaluating persistence or unusual ENT symptoms accompanied by unexplained bleeding, as these may indicate more severe underlying conditions.[5]

Management strategies and the multidisciplinary approach

Management across the five cases required a combination of supportive care, targeted factor replacement and surgical intervention. Treatment modalities included FFP, PRBC transfusions for severe anaemia (case 5), desmopressin and recombinant vWF in case 4 and recombinant factors VIII and IX (cases 2, 3, and 5).

In case 3, medical therapy failed to control the haemorrhage, necessitating advanced interventional radiological embolization. The successful outcomes-discharge on POD 11 for case 3 and clinical improvement by week 2 for case 5 - demonstrate that a collaborative, multidisciplinary approach involving paediatricians, haematologists, ENT surgeons and interventional radiologists is paramount. Early recognition by the otorhinolaryngologist prevents delayed diagnosis and allows for timely, targeted therapy, significantly reducing morbidity and mortality in this complex patient population.

CONCLUSION

The ENT manifestations of haematological diseases are diverse and clinically significant. A collaborative, multidisciplinary approach and a heightened clinical

awareness are paramount for ensuring timely diagnosis, effective management and improved outcomes for this complex patient population.

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