International Journal of Pediatric Otorhinolaryngology 76 (2012) 767–771

Contents lists available at SciVerse ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Review article

Predictors of bleeding disorders in children with epistaxis: Value of preoperative tests and clinical screening☆

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ARTICLE INFO

Article history:
Received 8 November 2011
Received in revised form 7 February 2012
Accepted 8 February 2012
Available online 27 March 2012

Keywords:
Epistaxis
Bleeding disorders
Von Willebrand disease

ABSTRACT

Objective: To identify prevalence of previously undiagnosed bleeding disorders in children with severe epistaxis who failed medical therapy requiring intraoperative nasal cautery.
Study design: Retrospective chart review 10/15/2006–12/31/2010.
Setting: Single provider outpatient otolaryngology clinic.
Subjects and methods: Inclusion criteria: children (<19 years) with epistaxis referred to otolaryngology, no known bleeding disorder, failed medical therapy and received surgical nasal cautery. Data collected: duration/severity of epistaxis, bleeding history, family history of bleeding. A screening CBC, PT and PTT were performed on all patients.
Results: Of 248 subjects referred for epistaxis, 47(19%) met inclusion criteria (mean age 9.2 ± 0.5 years; 61.7% male). 31.9% (15/47) had abnormal coagulation studies but on repeat testing only 2 patients had persistent coagulation abnormalities. 15 patients were referred to hematology, 5 were diagnosed with a bleeding disorder (3 – type 1 von Willebrand's disease, 1 – platelet aggregation disorder, 1 – mild factor VII deficiency). Out of the entire cohort 10.6% (5/47) had a bleeding diathesis. Clinical predictive factors for having a bleeding diathesis were explored and included previous emergency room visits for epistaxis ($p = 0.04$). A trend was found in those presenting with epistaxis at a younger age ($p = 0.07$).
Conclusion: Children with recurrent epistaxis despite medical therapy are at higher risk of having a bleeding disorder. In this highly selected group of patients 10.6% (5/47) were found to have a bleeding disorder. Screening coagulation studies (PT, PTT) only revealed 20% (1/5) of patients with a bleeding disorder. Only a subsequent comprehensive hematology evaluation revealed the diagnosis in the majority of patients.

Contents

1. Introduction

Epistaxis is a frequent occurrence in childhood affecting up to 60% of pediatric patients [1] The etiology of epistaxis can be diverse including trauma (digital manipulation is common in pediatrics), medications, allergic rhinitis, dehumidification, septal perforation, neoplasm, hereditary hemorrhagic telangiectasia (HHT), or an acquired or congenital bleeding disorder [2,3]. Otolaryngologists

☆ Paper presented as a poster at AAO-HNS Annual Meeting, San Francisco CA, September 2011.

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doi:10.1016/j.ijporl.2012.02.033

are frequently asked to evaluate pediatric patients for epistaxis and the majority of cases are mild, due to local events and respond to basic medical therapy. The smaller subset of pediatric patients who continue to have epistaxis despite appropriate medical therapy may be at increased risk for having a bleeding disorder.

Mild bleeding disorders in children frequently coincide with a primary complaint of epistaxis. Unlike adults, children with mild bleeding disorders are less likely to present with other bleeding symptoms because of a lack of significant hemostatic challenges such as prior surgery, dental procedures or the onset of menses. Otolaryngologists caring for patients with epistaxis are in a unique position to assist in the diagnosis of an underlying bleeding disorder. Knowledge of whether a child has a coagulopathy prior to surgery can limit complications during cauterization and reduce the likelihood of excessive bleeding when faced with other future surgeries, childbirth, or trauma. Currently, few papers describe the optimal means of evaluating children preoperatively who have persistent epistaxis requiring surgical cauterization. (3–6) The objective of this retrospective study is to determine (1) the value of preoperative blood screening with prothrombin time (PT) and activated partial thromboplastic time (PTT) testing and (2) the prevalence of undiagnosed bleeding disorders in children with epistaxis who fail medical therapy and require surgical cauterization.

2. Methods

This study was approved by the Institutional Review Board at The Children's Hospital of Philadelphia.

Potential cases were identified by querying billing records for a single otolaryngology provider for the International Classification of Diseases (ICD-9-CM) epistaxis code (784.7) from 10/15/2006 to 12/31/2010. Patients were included in the study if they were between the ages of 0 and 18 years, had recurrent epistaxis, failed epistaxis medical treatment with emollient therapy and had subsequent intraoperative cauterization of the septum. Patients were excluded if they had been successfully treated medically or had a known bleeding disorder at the time of initial evaluation or a structural source of bleeding including tumors, HHT or septal perforations. Per the provider's practice, each patient had a preoperative complete blood count (CBC), prothrombin time (PT), and activated partial thromboplastin time (PTT). If these screening tests were abnormal or if there were additional concerns, patients were referred to hematology for further evaluation. The electronic medical record was reviewed to collect subject's demographic information, epistaxis severity, other personal history of bleeding, family history of bleeding, and preoperative coagulation testing. All hematologic and coagulation testing were reviewed by a single pediatric hematologist.

Data analysis was performed using STATA 11.0 (StataCorp LP, College Station, TX, USA). Categorical variables were summarized using frequencies and percentages. For continuous variables the data was summarized as mean $\pm$ one standard deviation. Continuous data were analyzed using the student's *t*-test and categorical data were analyzed using Fisher's exact test. A p-value $\leq .05$ was considered significant.

3. Results

From 10/15/2006 to 12/21/2010, 248 subjects were evaluated by a single physician because they had recurrent epistaxis. Of those excluded from the study, 191 were successfully treated with daily topical emollient therapy that sometimes also contained antibiotics. Eight patients were excluded from the study who did need intraoperative intervention because they had a pre-existing known bleeding disorder including those with an acquired coagulopathy (3 resulting from chemotherapy) or a congenital bleeding disorder (3 type 3 von Willebrand (VWD), 1 idiopathic thrombocytopenia (ITP), and 1 platelet aggregation disorder). Two patients were excluded because they had a structural abnormality requiring repair (septal perforation), and no patients were found to have tumors such as a juvenile angiofibroma (JNA). A total of 47 (19%) subjects met the study inclusion criteria and required surgical intervention (electrocautery of the septal vessels). The mean age was 9.2 ± 0.5 years with a male predominance of 61.7% (29/47). Forty-three percent (20/47) of subjects reported an epistaxis duration of 5 min or longer with bleeding from both nares in 63.8% (30/47). Frequent epistaxis (>25 epistaxis episodes/year) was found in 76.6% (36/47) of subjects. Only two patients had a prior history of iron deficiency anemia that was attributed to recurrent epistaxis. 19.1% (9/47) reported prior emergency room visits for epistaxis. 6.4% (3/47) required emergent nasal packing and 17% (8/47) had prior nasal cautery. Only one subject reported an additional history of bleeding. No patients reported a family history of a specific bleeding disorder but 21.2% (10/47) reported a nonspecific family history of excessive bleeding.

All patients who were offered surgery subsequently underwent cauterization. A screening CBC, PT and aPTT were available for all subjects. Anemia was found in 8.5% (4/47) of patients with a hemoglobin range of 8.1–11.1 g/dL. No subjects had thrombocytopenia. With initial coagulation screening 31.9% (15/47) of patients had either an abnormal PT, aPTT or both. Fig. 1 is a flow diagram for coagulation testing results. Of the 15 subjects with abnormal coagulation studies, 12 patients were referred to hematology. Of the three patients not referred to hematology one had repeat coagulation studies that normalized and two patients with a minimally prolonged PT (<0.2 s prolonged) proceeded to surgery without repeat coagulation testing or a hematology consultation. Three additional patients with normal coagulation studies were referred to hematology secondary to additional concerns of anemia or no family history secondary to adoption (Refer to Fig. 1). Five of the 15 patients (33%) referred to hematology were subsequently diagnosed with a coagulopathy. The most common diagnosis was type 1 VWD in three children, platelet aggregation disorder in one and factor VII deficiency in one subject. One patient had an abnormal test result that subsequently normalized, thought to be related to the presence of a temporary inhibitor (discussed below).

Exploratory analysis was completed to determine if there were any potential clinical predictors for identifying patients who have a coagulopathy. These factors included additional personal history of bleeding, family history of a specific bleeding disorder, family history of bleeding, anemia from epistaxis, history of emergent nasal interventions, history of ER visits for epistaxis, prior nasal cautery, patient sex, age, duration of epistaxis, location of epistaxis, and frequency of epistaxis. Statistical analysis showed a significant correlation between a prior history of ER visits and the presence of a diagnosable coagulopathy (p value = 0.04). Children diagnosed at a younger age showed a trend suggesting they were at higher risk of having an underlying coagulopathy (p value = 0.07). Refer to Table 1.

4. Discussion

In our single institution study of pediatric patients with recurrent epistaxis despite medical therapy 10.6% (5/47) were diagnosed with an underlying bleeding diathesis. While this is a high proportion, it is likely an underestimate because only 31.9% (15/47) of the subjects had a complete hemostatic evaluation. Accomplishing hemostasis at the site of vascular injury is a complex process including vessel constriction, platelet adhesion at the site of injury facilitated by binding to von Willebrand factor