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Adenotonsillectomy In A Child With Factor VII Deficiency

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I. Introduction:

Tonsillectomies have been performed from the ancient times for tonsillar hypertrophy. They've been done time and again with minimal complications and very few contra-indications. One of these relative contra-indications would be coagulopathies- given the tendency for haemorrhage. Factor VII deficiency is one among those coagulopathies - a rare autosomal recessive factor haemorrhagic deficiency. Here we present a six year old child with factor 7 deficiency on whom we done an adenotonsillectomy.

II. Case History:

A 6-year-old male child was brought to the ENT outpatient department with complaints of recurrent upper respiratory infections since childhood (approximately ten episodes a year.) Child also had complaints of mouth breathing. On examination; grade three tonsillar hypertrophy was noted and an X-RAY Nasopharynx was suggestive of adenoid hypertrophy. An adenotonsillectomy was planned for the patient.

Routine investigations were carried out for surgery, the prothrombin level was identified to be high (18.7 s) with INR normal (1.78). Paediatric opinion was obtained and on evaluation of clotting factors, the child had low factor 7 levels. On probing of family history, there was no history of bleeds and it was a non- consanguineous marriage. Surgery was cancelled due to expected complications and family's wishes

A couple months later child was again brought to OPD, this time with obstructive tonsillitis (grade four tonsillar hypertrophy) and dysphagia. On discussion with paediatric haematologist and paediatric oncologist, child was taken up for adenotonsillectomy with necessary precautions.

Before surgery, two units of fresh frozen plasma was given, each unit was transfused over 2 hours. One more unit of fresh frozen plasma was given intra-operatively and post-operatively over 4 hours.

We used coblation method to remove both tonsils and blind curettage for adenoidectomy. The whole procedure took less than thirty minutes and bleeding was watched for meticulously during the entire process.

Post -operatively, patient was monitored for two days. There were no episodes of bleeding either via the oral cavity or nasal cavity. Tonsillar fossa and posterior pharyngeal wall were normal.

Child was discharged on post-operative day 2. Parents were counselled about the complications and danger signs. On review child was vitally stable with a healthy fossa and no complaints of bleeding. Parents were further counselled about the disease and the need for genetic testing.

III. Discussion:

Patients who have episodes of tonsillitis and known coagulopathies themselves or have family members with such a history represent a very significant problem and one that should be approached with great caution. Although such patients may undergo tonsillectomy uneventfully, there is a possibility of significant and prolonged bleeding at any point from the time of the procedure through healing of the fossae some weeks later.

Factor VII deficiency is the most common among rare autosomal recessive bleeding disorders, with an incidence estimated at 1 in 500,000. This is one of the vitamin K dependent proteins produced by the liver.

When vessel injury occurs, nonvascular cells (fibroblasts, vascular and smooth muscle cells) with tissue factor on their surface are exposed to blood. Tissue Factor will bind to FVII and FV11a (in the presence of calcium) forming the FVIIa/TF COMPLEX. Once formed, this initiates the extrinsic and intrinsic pathways of blood coagulation eventually leading to formation of fibrin clot and haemostasis.

Bleeding symptoms can range from easy bruisability to life threatening haemorrhage. Clinical features include bleeding from skin or mucous membranes, such as epistaxis, gingival mucosa, or menorrhagia. Bleeding into the joints (hemarthrosis), soft tissue bleeding and bleeding into stomach, intestines and urogenital tract can also occur, resulting in blood in urine and stools.

Diagnosis involves PT levels. Factor VII is the only plasma coagulation factor deficiency in which the PT alone is prolonged (APTT would remain normal). INR values are also elevated.

Management includes recombinant factor VIIa (optimal replacement therapy), prothrombin complex concentrates (which contain factors 2,7,9,10) and plasma derived factor VII concentrates.

Because FVII has a short half-life (3–4 h), repeated transfusion may be necessary. In our case, we transfused the child before, during and after the surgery hence ensuring adequate maintenance of clotting factors.

IV. Conclusion:

Factor VII deficiency is a rare congenital disorder which presents with isolated elevated PT levels. With adequate care and precautions one can go ahead with performing a tonsillectomy on patients with tonsillitis.