Fig-IV- Excised cyst with a tooth

Fig.V- Residual defect after excision of cyst

Fig.VI- Post-op picture after 2nd operation

In children, best results are obtained by marsupialization as enucleation may lead to damage to involved tooth. In adults the tooth is unlikely to erupt therefore enucleation together with removal of associated tooth is preferred⁽¹⁾. In the present case the cyst was very large and caused expansion of surrounding bones therefore enucleation was done in first stage and reconstruction in the second stage

CONCLUSION

It can be concluded that a simple dentigerous cyst can acquire a huge size and cause distortion of the face, so that it requires major reconstructive surgery for restoration of a normal face.

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Address for correspondence:

Dr. Chandra Shekhar
Associate Professor and Head of the Dept.,
Dept. of ENT,
Patna Medical College, Patna.

Clinical Report

DISSEMINATED INTRAVASCULAR COAGULATION SYNDROME - ENT SURGEON'S PERSPECTIVE

A.Ravikumar,* Vivek Sasindran,** K.Senthil,*** John Samuel,**** Thirumaran ****

Abstract: ENT surgeon is seldom confronted with a systemic disease severe enough to progress to Disseminated Intravascular Coagulation syndrome (DIC), where he has to undertake a definitive surgical procedure for malignancy or perform an emergency tracheostomy. This calls for a judicious calculation of the pros and cons with an element of risk that needs to be accepted. We report two such patients who have been treated in such a scenario where the outcome varied depending on the severity and time of onset of the underlying coagulation disorder.

Keywords: Bleeding Disorders, Disseminated Intravascular Coagulation, Hepatitis B infection

Key for Abbreviations:

DIC- Disseminated Intravascular Coagulation
INR- International Normalized Ratio
FFP- Fresh Frozen Plasma
PT- Prothrombin Time
PTT- Partial Thromboplastin Time
APTT- Activated Partial Thromboplastin Time
USG- Ultrasonography
CT- Computed Tomography
TFA- Tissue Factor Activity

INTRODUCTION

Disseminated intravascular coagulation is a condition characterized by widespread activation of coagulation process, which results in intravascular formation of thrombin and ultimately thrombotic occlusion of small and midsize vessels^[1,2].

CASE 1:

38 yr old male patient presented with complaints of difficulty in breathing for 4 months. He was asymptomatic till 6 months ago when he developed Hepatic Encephalopathy following Hepatitis

*Professor & Head- Dept of ENT, Head & Neck Surgery, **Post Graduate, *** Associate Professor, **** Assistant Professor- Dept of ENT, Head & Neck Surgery, Sri Ramachandra Medical College & Research Institute (DU), Porur, Chennai

B infection and was intubated and ventilated for 2 weeks duration. The breathing difficulty developed 2 months later and progressively worsened. Examination revealed a dyspnoeic patient in stridor but comfortable at rest. Indirect laryngoscopy showed a normal endolarynx confirmed by videolaryngoscopy. However the subglottis and the trachea were narrowed. CT scan of the neck showed a stenotic segment extending from just below level of the vocal cords to the second tracheal ring. However oxygen saturation was maintained on room air. Investigations showed low platelet count (40,000) with markedly raised INR (4.06). Liver enzymes and fibrin degradation products were elevated (> 4000). Hematologist opined that patient had Disseminated Intravascular Coagulation syndrome. Before performing any surgical procedure to relieve his airway obstruction, the deranged clotting profile was corrected by infusing FFP and platelet concentrate (3 units each) twice daily for 3 days. The platelet count rose to 1,00,000 and the INR lowered to 1.6 and could not be improved any further. After this he was taken up for elective tracheostomy under local anaesthesia after obtaining high risk consent. Trachea was opened between the second and the third tracheal rings and size 8 Portex cuffed tracheostomy tube used. Very minimal bleeding was encountered during the procedure, which was uneventful. Intraoperatively 2 units of FFP and 1 unit of platelet were transfused. Post operatively 2 more units of FFP were transfused daily for 7 days, till the coagulation parameters were stabilized. Definitive treatment of tracheal stenosis resection anastomosis was considered highly risky and haematologist opined against such a major procedure. Weighing the options, by informed decision making, the patient and his family chose to maintain a tracheostomy as opposed to the morbidity and possible mortality associated with resection anastomosis. At the time of discharge Portex tube was changed over to Shiley's non cuffed tube with speaking valve since the patient chose to remain on tracheostomy tube indefinitely. A total of 64 units of FFP and platelet concentrates were transfused during his 20 day stay in the hospital. Patient was relieved of stridor and could speak at the time of discharge; however his coagulation parameters were still deranged inspite of aggressive management, due to underlying liver disease. He is still being followed up regularly for last 18 months and has no respiratory or speech difficulty.

CASE: 2

56 yr old female presented with hoarseness of voice for 3 years. She had no other associated complaints like difficulty in breathing, blood stained sputum or choking over food. Clinical examination revealed exophytic growth occupying the region of the right vocal cord in the mid third. There was no cervical lymphadenopathy. Videolaryngoscopy confirmed the findings. CT scan of the larynx showed supraglottic extension to aryepiglottic fold with no involvement of the pre-epiglottic space. Two biopsies of the growth taken earlier by microlaryngoscopy were suggestive of malignancy. She also had a short neck and hypognathia which caused difficulty in visualizing the larynx adequately. Clinically given the malignant nature of the growth it was decided to perform a vertical partial laryngectomy by laryngofissure approach. Elective Tracheostomy under local

anaesthesia followed by laryngofissure under general anaesthesia was done. Tissue sent for frozen section biopsy was reported as squamous cell carcinoma. Using a KTP 532 laser (0.2 mm fibre with 4-6 watt power) a right frontolateral partial laryngectomy was performed. Right vocal cord with anterior commissure and part of the thyroid cartilage were excised and submitted for HPE. The total blood loss during the surgery was 120-130 ml. Patient was stable for 36 hrs post op. Drain was 10-15 ml. About 36 hrs post op patient developed breathlessness and bradycardia, was resuscitated, shifted to the ICU and placed on ventilatory support. 3 hrs later patient developed bleeding from the nose, oral cavity and tracheostomy site. Hemoglobin, total counts and platelets counts were 6 gm%, 22,000 and 66,000 cells/cmm respectively. PT, PTT and INR were raised. FDP was high (> 2000). USG abdomen revealed perinephric collection and right sided pleural effusion. The patient was diagnosed to have Disseminated Intravascular Coagulation syndrome. In spite of transfusing several units of FFP and packed cells as per the hematologists advice, urine output was only 40 ml on the third post op day. She was diagnosed to have metabolic acidosis with acute renal failure. Patient had a cardiac arrest for the second time and was resuscitated. On the fourth post operative day PT, PTT and INR were high but bleeding had stopped from all the sites. FFP, packed cells and whole blood transfusions were being continued. Patient had anuria and heparin free hemodialysis was done. On the successive post operative days transfusion of blood products were being continued and hemodialysis was also done. On the fifth post operative day drain was removed. Patient developed abdominal distension and intraperitoneal bleed was suspected on ultrasound examination. CT scan of the brain revealed multiple infarcts with intracranial hemorrhage. Sutures in the neck were removed on the sixth post operative day. On the seventh post operative day patient had bradycardia and was resuscitated. For the next three days patient had persistent anuria and high fibrin degraded products. Hemodialysis was done along with transfusion of blood products. On the fourteenth post operative day, patient had cardiac arrest and could not be resuscitated. Retrospectively it was evident that it was the surgical trauma and also the underlying latent deranged coagulation mechanism that predisposed to the onset of DIC. This could not be detected preoperatively in the routine haematological investigations that were performed.

DISCUSSION

DIC usually results from entrance into or generation within the blood of material with tissue factor activity (TFA), which initiates coagulation. The endothelium may be disrupted so that tissue factor is released from tissue damaged by trauma, ischemia, excessive metabolic stress, heat, chemicals, infectious organisms, tumors, or activation of the complement attack sequence^[3].

MECHANISM OF DIC ^[4]

Infection is the commonest cause of DIC. About 10-20% of patients with gram negative bacteraemia have evidence of DIC ^[3]. Liver Diseases may also cause chronic DIC but the mechanism is still not clear. Malignancies also cause DIC by invasion of tissues and release of tissue factor, activation of leucocytes and secretion of tissue factor. Direct activation of the prothrombinase complex by mucin or a specific cancer procoagulant is also described ^[1,5]. In our study, case no.1 had DIC due to Hepatitis B and in case no.2 DIC is supposed to have been triggered due to release of tissue factor following manipulation of the malignant growth and surgical trauma as described above.

Clinically, a pathological degree of bleeding in a sick patient should alert doctors to the possibility of DIC especially in the background of severe sepsis or malignancy. Depending on the relative rates of formation and breakdown of fibrin, the syndrome may be asymptomatic or cause severe bleeding or thrombosis ^[3]. Haemorrhage is the commonest presentation and is caused by generation of free plasmin and depletion of coagulation factors and platelets ^[6]. Our first patient with chronic DIC did not have overt clinical manifestations whereas the second patient presented as acute DIC. In acute DIC the hemorrhagic features are also associated with hypovolaemia, hypotension and shock. Acute renal failure, generalized cortical and brain dysfunction are due to microvascular thrombosis in the kidney and brain respectively. ^[3]. Thrombosis and hemorrhage in the lungs cause hypoxia and progressive respiratory failure. Micro vascular thrombosis is the primary mechanism in most cases of DIC and end organ failure is a major cause of death. The second case reported here had all the above complications and succumbed eventually.

There is no single laboratory test that can establish or rule out the diagnosis of DIC ^[4]. Thrombocytopenia due to thrombin generation is an almost universal finding in acute DIC. It is sufficient to measure platelet count, PT, PTT, APTT and fibrinogen concentration ^[3]. High plasma concentrations of fibrinogen degradation products are not specific for DIC. D-dimer latex agglutination test is used to measure fibrin degradation products. Thromboelastography (TEG) measures the viscoelastic properties of blood as it is induced to clot under a low shear environment resembling sluggish venous flow.

Treatment of the underlying cause is essential for treating DIC. The condition will not resolve until the trigger mechanism is removed. Patients may be treated with blood components to replace depleted coagulation factors, platelets, and natural inhibitors of thrombin and plasmin to reduce bleeding while the underlying problem is corrected. Administration of vitamin K to

all patients with suspected DIC will replenish depleted stores. Parenteral antibiotics should be administered without delay if bacterial infection is suspected. Measuring the concentration of platelets, fibrinogen and assessing the PT and APTT are essential for guiding management ^[3]. However the decision to start replacing blood components is determined by whether the patient is bleeding and whether an invasive procedure is required. Low dose of heparin, thrombin inhibitor, plasmin inhibitors such as tranexamic acid or aprotinin are generally considered to be contraindicated because of the risk of increased end organ damage from microvascular thrombosis ^[5]. However they are judiciously used when patients continue to bleed despite treatment with blood components as was necessary in our second patient.

CONCLUSION

In the practice of modern medicine it is obvious that Bleeding time and Clotting time estimation are inadequate guides to evaluate bleeding disorders. It's mandatory to perform platelet count, PT, PTT, APTT and INR in all major surgical cases. These results will be able to point towards latent blood disorders that need to be addressed adequately before embarking on any major surgical procedure. It will also protect the surgeon against unnecessary litigation for a complication not directly due to surgical procedure.

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Address for correspondence:

Prof. A. Ravi Kumar
Professor & Head- Dept of ENT, Head & Neck Surgery,
Sri Ramachandra Medical College & Research Institute (DU),
Porur, Chennai- 600 116.
India. E-mail:- entsrmc@yahoo.co.in