Endometrial ablation for rare case of Glanzman's Thromboasthenia

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Glanzmans Thromboasthenia is a rare congenital bleeding disorder characterized by deficiency of platelet membrane glycoprotein IIb, IIIa complex associated with primary qualitative platelet deficiency. Apart from other hemorrhagic manifestations, menorrhagia since menarche is a therapeutic challenge to manage in these cases. A rare case with intractable menorrhagia since menarche which was refractory to conservative medical treatment with hormones and ovarian irradiation was treated with endometrial ablation. For more than one year she is asymptomatic with minimal spotting. In recent time endometrial ablation seems to be the answer for such bleeding diathesis.

Case Report

Miss G a 30 year old single female, product of nonconsanguinous marriage was referred for endometrial laser ablation as she had heavy bleeding per vaginum during periods since menarche.

She was a known case of Glanzman's thromboasthenia since childhood as she used to bleed profusely from gums & injury sites and had received multiple blood transfusions. Her menses became nightmare for her and her family when she attained menarche at 12 years.

She continued to bleed for 1-2 months at the gap of one to two months till the age of 15 when a Gynaecologist prescribed her Norethisterone for 21 days every month which she took for 9 months. But she started having very foul smelling bleeding, as clots were accumulated for which she underwent D & C twice.

Her haemoglobin used to fall every cycle by 3 gm% to 6gm%. On an average every cycle 14-18 units of blood and platelet extracts were given in addition to injections of Estrogens or Progesterone to control her bleeding.

She was advised by a haematooncologist for hysterectomy for which she was deemed unfit. So her ovaries were irradiated for 10 sittings which relieved her for 1 year. In 1984 she required another sitting of ovarian irradiation for 10 sittings.

Although she was told that this could keep her amenorrhoic for 10 years she started bleeding profusely with clots only 9 months later.

This time she resorted to homeopathic treatment which relieved her partially for 5 years from 1993 to 1996. She again started bleeding profusely needing hospitalization twice and received 50 units of blood and platelet extracts. Meanwhile she developed hypertension for which she was investigated and no cause was found, thus it was ascribed to chronic hormone ingestion. She was started on Atenolol 25 mg once a day. She was referred for endometrial ablation for intractable bleeding and as a high risk case but because her haemoglobin was low she was kept on Danazol 200 mg for six weeks, to keep her amenorrhoeic and for building up of her haemoglobin.

She was readmitted in February 1997 for endometrial ablation in consultation with a haematologist, after proper counselling that this will relieve her problems to 70-90% only and that she may not be completely amenorrhoeic and second procedure may be required.

She was posted for endometrial ablation with due risk. On investigation her haemoglobin was found to be 11.3 gm% and rest of the investigations like CBC, Platelet count (1.65 lac), Renal Function test, liver function test and fresh coagulation profile were within normal limits. Inspite of having received more than 200 units of blood and platelet extract, she was found to be negative for Hbs Ag, HIV I &II, HCV and VDRL.

On 26th February 1997 she was taken to operation theatre after infusing her 300ml of platelet extract over half an hour before surgery. Adequate blood platelet extracts were kept ready. General anaesthesia was induced with, Pentothal and maintained on isoflurane and spontaneous breathing with mask. She was not intubated because of fear of injury to larynx & bleeding.

At the time of introduction of Sims speculum there was

a little abrasion of fourchette which started oozing and responded to pressure. Examination under anaesthesia revealed antiverted normal size uterus with no palpable pathology in adnexa.

Cervix was dilated to no 8.5 Hegars dilator and 26 French Resectoscope was introduced and a polyp measuring 5 x 4 x 3 mm was removed and was sent for histopathological examination.

The endometrium was ablated with electrocautery upto internal os using cylinder at 80-100 watts. Patient withstood surgery well and intraoperative blood loss was less than 10 ml out of which 6-7 ml was from bleeding from abrasion in the fourchette. This also shows her severe bleeding tendency despite a planned atraumatic procedure. The whole procedure took 26 minutes and no blood transfusion or platelet extracts was required. Post operatively she had little bleeding from vagina which subsided spontaneously. On the 3rd post operative day her haemoglobin was 9.7 gm%. Danazol 200mg once a day was prescribed for one month to improve the results. She was discharged on post operative day 4. Histopathology of the endometrial polyp showed hyperplasia with no evidence of malignancy or atypia. Since more than one year after the endometrial ablation, she has not needed any blood transfusion or platelet concentrates till today. She is amenorrhoic with insignificant intermittent spotting. Further follow up will be required to see whether endometrial ablation is the answer for such intractable menorrhagia in bleeding disorders. As such primary coagulation disorder is found in almost 20% of severe adoloscent menorrhagias.

It's interesting to know that her sister also has Glanzman's Thromboasthenia. She is 24 years of age with one normal delivery needing twenty bottles of blood transfusion to control bleeding after delivery. She also had significant menorrhagia and endometrial ablation was done for her on similar guideline and workup. Now again since more than 4 months she also is relieved of menorrhagia so far. Thus endometrial ablation could be an efficient treatment for excessive menstral bleeding due to coagulation disorders.

Lung Cancer in Follow up Patient of Carcinoma of Uterine Cervix.

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The tendency for multiple primary neoplasms (MPN) to develop in the same or contiguous epithelial surface or in bilaterally paired organs has been thoroughly documented for many years. There is no convincing evidence of predisposition for MPN to develop in unrelated tissues or organs. Metachronous occurrence of MPN in unrelated tissues is a curiosity as it can be important for two reasons. Firstly, it provides opportunity to the clinicians to study the common etiological agents and it alerts them to detect the associated cancer. We present an interesting case of a 62 year old woman who had carcinoma of uterine cervix and lung.

Case Report

A 51 year old lady was diagnosed to be a case of carcinoma of the uterine cervix, stage IIB in the year 1987 at our department. Histopathologically she had a large cell nonkeratinising cell carcinoma. Subsequently she was managed by radiation therapy with a curative intent. A total dose of 50 Gy was administered to the whole pelvis with midline shielding by a tele Cobalt unit. Following the external beam radiation, the patient was evaluated for intracavitary irradiation. A dose of 30 Gy was administered to the point A by remote afterloading equipment. She had an excellent regression of the cervical as well as parametrial disease following radiation therapy. The patient was on regular follow up at the combined gynaecology cancer clinic at our center since then. Eleven years later (1988) she presented with the chief complaints of pain over right chest, productive cough and hemoptysis. X-ray of chest showed a large homogeneous mass over the upper and middle lobe of the right side lung. Bronchoscopy biopsy revealed a poorly differentiated carcinoma. CT scan of the abdomen and pelvis showed no disease. Taking the clinical and histopathological features into consideration a second primary of the lung was diagnosed. In view of the large volume disease the patient is being treated with external beam radiation therapy with a palliative intent.