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Double jeopardy: the management of ruptured corpus luteum and Factor V deficiency in adolescence: a case report

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Introduction

- Factor V deficiency is a **rare inherited bleeding disorder**.
- Bleeding manifestations may become clinically significant during **menarche and ovulation**.
- Intra-abdominal bleeding can mimic an acute surgical emergency, creating a diagnostic and therapeutic dilemma.

Case History

13 year-old girl, Known case of moderate Factor V deficiency

First presentation

- Menarche 15 days earlier with **severe menorrhagia**
- Severe abdominal pain + vomiting
- Distended abdomen with **guarding and rigidity**
- **Vitals- Normal Blood pressure with Tachycardia in range 110-120/min**
- **Hb: 4.4 g/dL**
- **PT/INR: 42.7 s / 3.7**
- **aPTT: 89 s**
- Imaging: **Significant hemoperitoneum**
- Suspected ruptured hemorrhagic/corpus luteal cyst

Initial Management

- Conservative management as patient was initially stable
- Blood component support
- **Ovulation suppression with OCPs**
- Discharged after stabilization

Recurrent Bleeding episode

1 month later

- Shock → Recurrent hemoperitoneum → Acute renal compromise
- Emergency laparotomy performed
- Severe coagulopathy resulted in difficult surgical hemostasis
- Laparostoma was required
- Histopathology report - necrosis and hemorrhage
- Postoperative course marked by persistent spikes of fever, rapid drop in hemoglobin to 3.1 g/dl on D4 and development of 8.8* 3.5 cm subcutaneous abdominal wall hematoma.
- Regular FFP, PRBC and even recombinant factor 7 was administered.
- Growing hematoma caused compression resulting in obstructive uropathy and Acute kidney injury.
- Due to the refractory compression effects and continuous hematoma expansion, a secondary relaparotomy was performed, and a laparostoma was created to manage the recurrent hemorrhage.
- Her postoperative course was again strictly monitored and she was discharged successfully in stable condition with laparostoma and was given GnRh analogue- injection leuprolide 11.25 mg intramuscular for 3 months and was advised for regular follow up.
- After 1 month she succumbed to sepsis at home.

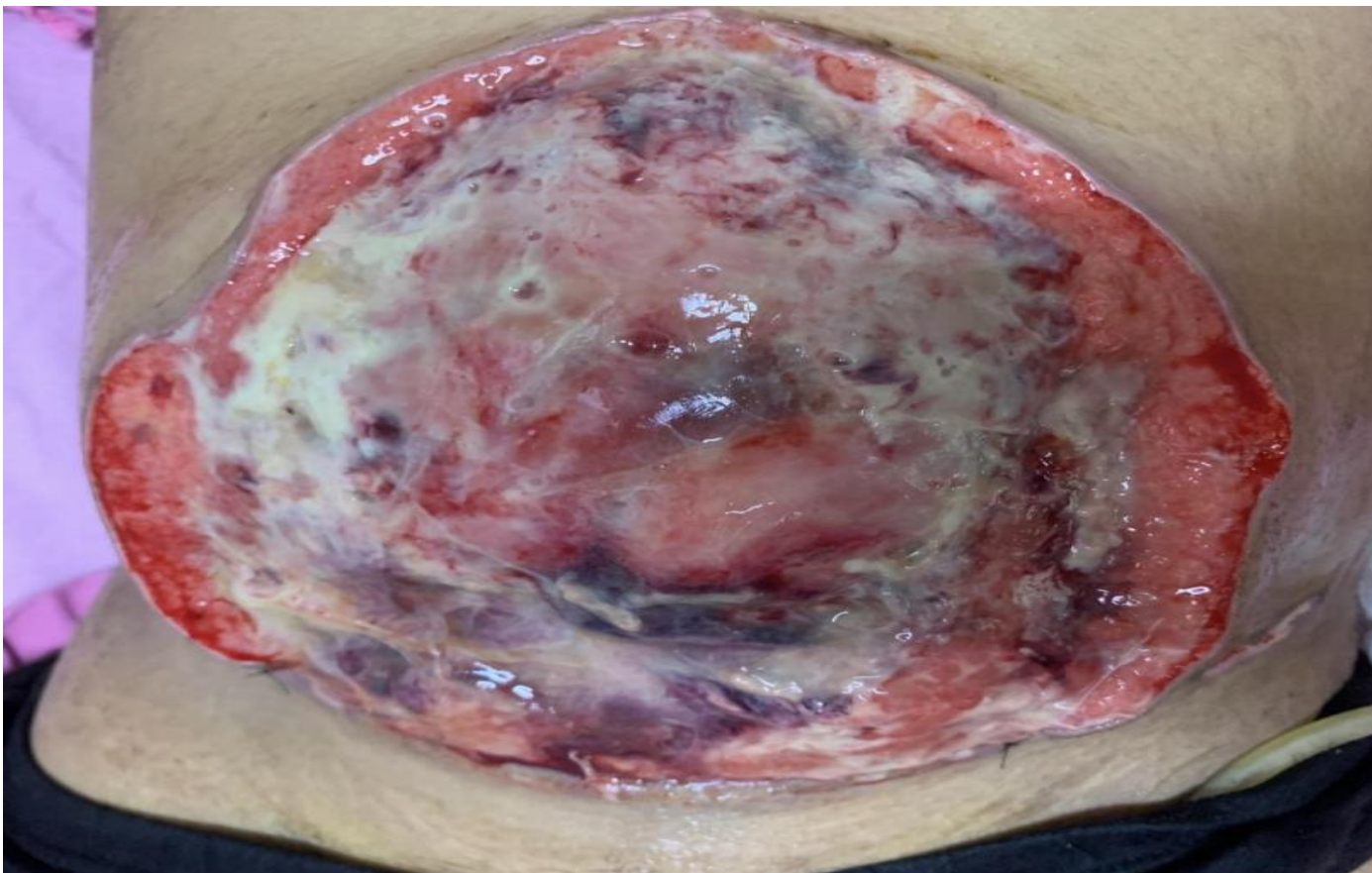


Figure 1: Granulation tissue over the open wound

Figure 2: Laparostoma covered with a uro bag covering

Learning from this case

Think beyond the surgical diagnosis: Hemoperitoneum in a patient with a bleeding disorder may have a coagulopathic component.

Recognize the risk at menarche: Menstrual and ovulatory bleeding may precipitate life-threatening hemorrhage.

Correct the coagulopathy: FFP remains the principal source of factor V replacement.

Early multidisciplinary management is critical: Delayed access to hematological and surgical care can be catastrophic.

Discussion

Factor V deficiency presenting around menarche can result in life-threatening menstrual and intra-abdominal hemorrhage. Early recognition, prompt FFP replacement, hormonal suppression, and coordinated multidisciplinary management are essential to prevent catastrophic outcomes.

Conclusion

- Menarchal females presenting with heavy bleeding or acute abdomen - always suspect coagulopathy disorders.
- Early diagnosis and management can result in better outcomes.
- Always screen the other family members for congenital disorders.