Case Report

*ACQUIRED HEMOPHILIA A* IN DIABETIC WOMAN

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Background

Acquired hemophilia is a rare condition in which autoantibodies, usually IgG class are produced against factor VIII or IX. Age distribution is bimodal. Although hemophilia is a hereditary disease, approximately 20-30% of patients have no family history of clotting disorders.

Case report

A 55-year-old lady with major complaints of gums bleeding, bruises on the both of leg and thighs that are varying colour (multiple spontaneus haematomas. No previous history of bleeding and no history of blood clotting disorders in the family. She has diabetes mellitus for 8 years was treated metformin three times a day, and )controlled level of blood glucose. Laboratory findings showed decreased haemoglobin (5,7 mg/dl) with prolonged aPTT 129 s(28 s control) but still got the normal PPT that is 17.2 s (14.4 s control) and normal INR 1,3, factor VIII activity got decrease with result of 1,5% (control 91,1%) but factor IX activity still normal 118,7% (control 108%). The Factor VIII inhibitor was 19.52 Bethesda Unit. After exclusion of other possible pathological condition and on the basis of lab criteria we diagnosed the case as acquired hemophilia A.

Discussion

The diagnosis of Acqured hemophlia A (AHA) shoud be considered in patient who present with bleeding and prolonged aPPT. The pattern of bleeding in AHA differs from that in congenital hemophilia A. Bleeding tends to occur in soft tissue, muscle, retroperitoneal space, iatrogenic bleeding is also common. The diagnosis is then confirmed by a Bethesda positive assay for F VIII inhibitor titre. Diagnostic test in AHA are clotting factor measurement (isolated low FVIII level) and quantification of the inhibitor titer (presence of inhibitor). Most often the cause is idiopathic in 50% of patient or it can be associated with autoimun disorders, hepatitis and diabetes. Chronic disease or diabetes mellitus makes misrecognition tends to self antigen.1,3

Conclusion

A 55-years-old lady with diabetes got symptoms gum bleeding, multiple spontaneuos hematomes and very low in factor VIII activity and presence of factor VIII inhibitor. It is concluded that this patient is diagnosed of acquired hemophilia A.

Keywords: Acquired Haemophilia A, activated partial thromboplastin time (APTT), Factor VIII.

References

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Laporan Kasus

*ACQUIRED HEMOPHILIA A* PADA WANITA PENDERITA DIABETES

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Latar Belakang

*Acquired* Hemofilia adalah kondisi sangat jarang di mana autoantibodi, IgG diproduksi terhadap faktor VIII atau IX. Distribusi usia adalah bimodal. Meskipun hemofilia adalah penyakit keturunan, sekitar 20-30% pasien tidak memiliki riwayat keluarga gangguan pembekuan.

Laporan kasus

Seorang wanita berusia 55 tahun dengan keluhan utama perdarahan gusi, memar di kedua kaki dan paha yang warnanya bervariasi (*multiple spontaneus hematoma*). Tidak ada riwayat perdarahan sebelumnya dan tidak ada riwayat gangguan pembekuan darah dalam keluarga. Ia menderita diabetes mellitus selama 8 tahun diterapi dengan metformin dan kadar glukosa darah terkontrol. Hasil laboratorium menunjukkan penurunan hemoglobin (5,7 mg / dl) dengan PT 129 (kontrol 28), PPT normal yaitu 17,2 s (kontrol 14,4 s) dan INR normal 1,3, aktivitas faktor VIII mengalami penurunan dengan hasil 1,5% (kontrol 91,1%) tetapi aktivitas faktor IX masih normal 118,7% (kontrol 108%). Inhibitor Faktor VIII adalah 19,52 Unit Bethesda. Berdasarkan kriteria laboratorium, diagnosis kasus ini adalah *acquired hemofilia* A.

Diskusi  
 Diagnosis *Acqured hemophlia* A (AHA) harus dipertimbangkan pada pasien yang datang dengan perdarahan dan pemanjangan PPT. Pola perdarahan pada AHA berbeda dari pada hemofilia kongenital A. Perdarahan cenderung terjadi pada jaringan lunak, otot, ruang retroperitoneal, perdarahan iatrogenik juga sering terjadi. Tes Bethesda positif untuk titer inhibitor F VIII. Tes diagnostik dalam AHA adalah dengan mengukur faktor pembekuan (tingkat FVIII rendah terisolasi) dan kuantifikasi titer inhibitor (inhibitor). Penyebab tersering pasien dikaitkan dengan gangguan autoimun, hepatitis dan diabetes. Penyakit kronis atau diabetes mellitus menimbulkan *misrecognition* terhadap antigen penderita.1,3

Kesimpulan  
 Seorang wanita 55 tahun dengan diabetes mengalami gejala perdarahan gusi, hematom spontan di paha dan perut dengan aktivitas faktor VIII yang sangat rendah dan munculnya faktor VIII inhibitor. Pasien ini didiagnosis menderita *Acquired hemofilia* A.

Kata kunci: *Acquired Haemophilia* A, *activated partial thromboplastin time* (APTT), Factor VIII.

Daftar Pustaka

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