

Case Reports

Henoch - Schönlein Purpura, a Rare Disease in Pregnancy

TABASSUM PARVEEN¹, FIROZA BEGUM²

Abstract

Introduction: Henoch-Schönlein purpura is an IgA-mediated small vessel vasculitis involving mainly skin, gastrointestinal system and kidneys. It is predominantly a disease of young children. Most common symptoms are rash (95-100%), sometimes necrotizing involving specially the legs, subcutaneous oedema (20-50%), abdominal pain and vomiting (85%), bloody stool and joint pain (60-80%) involving mainly the knees and ankles. Diagnosis is clinical and not based on laboratory evaluation. Its occurrence during pregnancy is exceptional.

Materials & Methods: On literature search, till date only 17-18 cases of Henoch-Schönlein purpura in pregnancy were found.

Case: Here we present a case report on this rare disease in pregnancy. A 39 year old lady, para 1+ 8 abortions, diagnosed as Henoch-Schönlein purpura at 24 weeks of gestation, treated with steroids and was cured almost completely. She was readmitted at 37 weeks of gestation with few purpuric rashes. Elective caesarean section was planned and a healthy male baby was delivered. But sub-total hysterectomy was needed for intractable bleeding from placental bed. Her post operative period was uneventful.

Conclusion: Very few information we got on literature search on Henoch-Schönlein purpura in pregnancy. Corticosteroids and plasmapheresis have been practiced as treatment during pregnancy. If kidneys are unaffected, obstetrical prognosis is good.

Key Words- Henoch-Schönlein Purpura, Pregnancy

Introduction:

Henoch-Schönlein purpura (HSP) is a small-vessel vasculitis characterized by purpura, arthritis, abdominal pain, and haematuria¹. In 1837, Johan Schonlein, a German pediatrician, described the association of non-thrombocytopenic purpura and joint pain, which he called purpura rheumatica. Later, his student, Edurad Henoch, noted the gastrointestinal and renal involvement in this disease².

The aetiology of HSP is unknown. About 50% patients have preceding upper respiratory tract infections. Multiple infectious agents as well as drugs, foods, insect bites may trigger HSP³. ASO titre is raised in 20-50% cases. It is predominantly a disease of young children; seventy-five percent (75%) are of age group 2-11 years. However in some series as many as 27% were adults. Male to female ratio is 2:1².

Purpura, arthritis and abdominal pain are known as classic triad of HSP⁴. Renal involvement occurs in

40-50% cases and is usually manifested by micro or macro-haematuria with or without proteinuria. Nephrotic syndrome or rapidly progressive glomerulonephritis may develop⁵. The disease tends to last about 4 weeks and then resolves spontaneously⁶.

Because it is uncommon in adults, there is little information on the effects of HSP on pregnancy in the literature. Only 17-18 cases of HSP in pregnancy till date were found on search. Symptomatology of HSP during pregnancy has no specific characteristics. Diagnosis may be difficult when renal, articular or gastrointestinal involvement precede the cutaneous manifestations since many aspects of HSP may masquerade as pre-eclampsia or eclampsia, especially hypertension, nephrotic syndrome, abdominal pain, headache and convulsions⁷. This is a case report documenting the clinical findings and outcome of HSP in pregnancy, a rare disease in pregnancy.

-
1. Assistant Professor- Feto-maternal medicine wing –Department of Obsterics & Gynaecology Bangabandhu Sheikh Mujib Medical university
 2. Professor- Feto-maternal medicine wing –Department of Obsterics & Gynaecology Bangabandhu Sheikh Mujib Medical university

Case Report

A 39 years old lady, para- 1+8 (abortion), hailing from Dhaka, housewife of a middle socio-economic class got herself admitted in Bangabandhu Sheikh Mujib Medical University (BSMMU) on 16th May, 2009 at 37 weeks of pregnancy with diagnosed case of Henoch-Schonlein purpura. Her pregnancy was uneventful up to 24 weeks of gestation. Then she developed fever, sore throat and arthritis. After 1 week she developed epistaxis, blurring of vision followed by complete loss of vision. At the same time she had extensive urticaria, with intense itching distributed bilaterally at the extensor surface of lower extremities, more pronounced below the knee joints. These transformed into typical purpuric rashes within few hours. She needed admission in hospital on 15th February, 2009 and diagnosed as a case of 'Henoch-Schönlein Purpura'. Reports of complete blood count, renal function test were within normal range. Tests to exclude autoimmune disorders like Anti-phospholipid antibody, Anti-Ds DNA antibody for SLE, cANCA, pANCA, Anti-nuclear antibody were negative. Prothrombin time (PT) of control was 11.8 sec and that of patient was 14 seconds, INR- 1.18, APTT – control-28 sec and patient-30 sec. She was diagnosed as a case of HSP and treated with Tab Prednisolon(5mg)- 2 tablet six(6) hourly with gradual tapering dose for two months. She was also given Intra-vitreous Inj. Lucentis (Intra-vitreal triamcinolone acetonide - Ranibizumab), a recombinant humanized monoclonal antibody, monthly for 3 months. Her eyesight gradually returned and other symptoms also subsided but few purpuric rashes and swelling of knee joints persisted which was noted to increase on walking.

She gave history of low back ache and polyarthritis with occasional haemarthrosis for last 15 years which aggravated by trauma. She took advices for several times from orthopedic specialist and was treated accordingly.

She was married for 16 years; age of her only child was 10 years. She had 8 abortions which occurred mostly in the 1st trimester and needed DE&C several times for incomplete abortion. Cause of these repeated abortions could not be identified by doing relevant investigations. Her only living child was the 6th conceptus who was delivered by emergency LSCS at 34 weeks of gestation for preterm labour with less foetal movement. She is non-diabetic, normotensive, non-smoker.

On examination she was obese, weight 82 kg, height 5'2". She was mildly anaemic, nonicteric, non-oedematous and normotensive. Thyroid gland was not enlarged and cardio-respiratory system was normal. Few purpuric rashes of sizes 5-7 mm were noted on extensors surfaces of the lower legs below the knee joints. These were pale-red in colour, did not disappear on pressure. Both the knee joints were swollen and tender.

On per-abdominal examination symphysis-fundal height was found 36cm, liquor volume was adequate. There was a single foetus in longitudinal lie and cephalic presentation.

Elective caesarean section was planned on 18th May, 2009. A healthy male baby of 3.2 kg was taken out having good Apgar score. While delivering placenta, it was noted to be tightly adherent with uterus (placenta accreta). After separation of the placenta, extensive bleeding was noted from placental bed and all the vessels were found to be enormously dilated. All steps were taken to control bleeding but failed and sub-total hysterectomy was done to save the life of the mother. She needed four units of blood transfusions. Post operative period was uneventful. Gradually her condition improved and she was discharged on 10th post-operative day.



Fig.-1: Numerous Purpuric lesions on leg.

Discussion

HSP is an IgA-mediated small vessel vasculitis which commonly involves skin, gastrointestinal system and kidneys. Pathophysiology is small vessel vasculitis characterised by Immunoglobulin A (IgA), C3 and immune complex deposition in arterioles, capillaries and venules⁴.

Various types of infectious agents can provoke HSP³. As we have seen in this case there was

history of fever, sore throat and arthritis for about a week before developing typical features of purpura.

Most common symptoms are rash (95-100%), sometimes necrotizing involving specially the legs, subcutaneous oedema (20-50%), abdominal pain and vomiting (85%), bloody stool, joint pain (60-80%) specially involving the knees and ankles. This patient presented with typical purpuric rashes and arthritis. As neurologic manifestations of the disease focal deficits like aphasia, chorea and cortical blindness are seen occasionally. Shrestha and colleagues found frequent presentations of non-classical features while evaluating 37 adult patients with HSP e.g. epistaxis in 30% and features of eye involvement in 8% cases⁸. Ryder and colleagues reported cortical blindness in a case report⁹. This patient also developed complete loss of vision and was treated with intra-vitrous angiogenic agent and her eye-sight returned to normal. Epistaxis was another presenting feature of this patient.

Diagnosis is clinical and not based on laboratory evaluation. Some tests help in excluding other diagnoses. CBC and differential count usually remain within reference range. However, WBC count and erythrocyte sedimentation rate (ESR) may be found to be elevated. Eosinophilia is sometimes present. Platelet count is usually in the reference range. Normal platelet count rules out idiopathic thrombocytopenic purpura (ITP). A normal platelet count and normal coagulation studies i.e. Prothrombin time (PT), Activated partial thromboplastin time (APTT), Fibrin degradation products (FDP) rule out thrombotic thrombocytopenic purpura (TTP). Renal function including urine analysis shows Haematuria and/or Proteinuria in 10-20% cases. BUN and creatinine levels may be elevated if there is renal involvement. C-reactive protein is usually raised. Circulating IgA level is also raised. Normal lipase level excludes pancreatitis. Immunofluorescence studies characteristically reveal deposits of IgA, IgG, C3, properdin and fibrinogen but not C4 and C1g in the glomeruli. Similar deposits are also seen in dermal capillaries in skin biopsy. In this case, investigations were performed to exclude renal involvement and to exclude other related disorders like SLE, idiopathic and thrombotic thrombocytopenic purpura and others. Almost all investigations were within normal range.

Regarding treatment very limited data are available. Fortunately most patients recover quickly in several

weeks without treatment. Non-steroidal anti-inflammatory drugs (NSAIDs) may help to reduce joint pain and do not seem to worsen purpura. Clinicians often use corticosteroids. Prednisolone 1mg/kg/day for 2 weeks and tapered over 2 weeks has been shown to improve gastro-intestinal and joint symptoms¹⁰. Treatment regimens include IV or oral steroids with or without any of the following: azathioprine, cyclophosphamide, cyclosporine, dipyridamole, plasmapheresis, high dose IV immunoglobulin G, danazole or fish oil³. Treatment with steroids may help to suppress external manifestations but neither steroid nor immunosuppressants have been clearly shown to improve renal outcome. This patient was treated mainly with oral steroids.

Regarding prognosis, HSP is generally a benign disease with an excellent prognosis. More than 80% cases, disease lasts for few weeks. In 10-20% cases there is recurrence and fewer than 5% patients develop chronic HSP¹¹.

Occurrence of HSP in pregnancy is exceptional. In a review in 2003, Kalamantis and colleagues¹² found that only 16 cases of HSP during pregnancy in the literature worldwide. They presented a case report of HSP in a 32-year-old woman as a seventeenth (17th) case.

The effect of pregnancy on the course of preexisting HSP remains to be evaluated. Analysis of four cases reported in a literature showed: aggravation of the disease in two patients^{13,14}, diminution of proteinuria in one patient¹⁵ and disparities of symptoms in one patient¹⁶. The only obstetrical complication of HSP related in the literature was pregnancy-induced hypertension and its possible consequences (preeclampsia, eclampsia)¹⁴.

Cummins DL and colleagues¹⁷ reviewed and evaluated all cases of immunofluorescence-proven HSP of pregnant women, diagnosed as having new-onset HSP, in the Department of Dermatology at the Johns Hopkins Hospital between 1990 and 2002 and three (3) cases were found. Two patients developed new-onset HSP, one at 16 weeks gestation and one at 22 weeks, while the third developed a recurrence of HSP at 12 weeks gestation after 19 years of remission. Authors concluded that pregnancy may be a trigger for onset or recurrence for HSP in susceptible individuals.

This patient presented at 24 weeks of gestation with acute features of HSP. However she had polyarthritis

with occasional haemarthrosis for last 15 years but no definitive diagnosis was made. So we are not sure whether she had pre-existing disease and pregnancy aggravated the condition .

Monica and colleagues¹⁸ presented two cases of HSP with pregnancy. First one was a 35-year-old woman with 27 weeks of pregnancy developed palpable purpura with necrotizing cutaneous lesions on the lower limbs, epigastric pain and arthralgias. Histological examination of a skin biopsy showed leukocytoclastic vasculitis with intravascular fibrin thrombi. The direct immunofluorescence analysis evidenced vascular deposits of IgA and C3 in the upper and mid-dermis. There was no evidence of renal involvement or placental dysfunction. The patient was treated with low-dose oral corticosteroids and a healthy infant was delivered by cesarean section. Examination of the placenta disclosed no signs of vasculitis or infarction. They reported another case of 33 years pregnant woman with HSP, who also had good fetomaternal outcome.

Robert Feldman et al¹⁹ reported a 35-year-old primiparous woman at 31 week of gestation, admitted with 4 weeks history of purpura on the legs, epigastric pain and arthralgias, mainly in the knees. Biopsy of a skin lesion showed leukocytoclastic vasculitis with intravascular fibrin thrombi. The direct immunofluorescence analysis revealed vascular deposits of IgA and C3 in the upper and mid-dermis. She was treated with low-dose oral corticosteroids in a tapering dose . The lesions rapidly resolved. Subsequent antepartum course was unremarkable. At 38 weeks of gestation, a healthy male baby weighing 4100 g was delivered by cesarean section. Histologic examination of the placenta showed no vasculitis or infarction. Direct immunofluorescence did not show evidence of immunoreactants.

Available studies showed that most of the cases of HSP in pregnancy were treated with low-dose oral corticosteroids. Mode of delivery was mostly lower segment caesarean section (LSCS) which matched in our case also. There is probably no risk of IgA vasculitis in the fetus as these immunoglobulins cannot cross the normal placenta. There was no evidence of vasculitis of placental vessels. Examination of the placenta of two reported cases showed no vasculitis or infarction. Fetomaternal outcome was excellent. ^{12,17,18,19}

In this patient, there was history of recurrent pregnancy loss (RPL). Available studies did not show any relation of HSP with RPL. While doing LSCS, we noted abnormal and excessive bleeding from the sinuses. Placenta looked normal except slightly more adherence with the uterine wall and needed manual separation. Subtotal hysterectomy was done to control the haemorrhage. However available literatures, found on search, did not show any such incidence and placenta did not show any dysfunction or features of vasculitis on histopathological examination. However there is definitely further scope of research about the disease in pregnancy, its role on recurrent pregnancy loss and placental changes as well.

Conclusion

Occurrence of HSP during pregnancy is exceptional. Very few information we got on literature search on HSP in pregnancy, most were case-reports. Little experience exists concerning treatment of HSP during pregnancy, although corticosteroids and plasmapheresis have been used. Placenta was not found to be affected with features of vasculitis . If kidneys are spared, obstetrical prognosis is good in this rare disease.

References:

1. Jennette CJ, Milling DM, Falk RJ: Vasculitides affecting the skin. *Arch dermatol* 1994, 130:899-906.
2. Schonlein-Henoch purpura Who named it? Henoch-Schönlein purpura – Wikipedia, the free encyclopedia [last modified on 27 June 2009]; 1-6. Available from http://en.wikipedia.org/wiki/Henoch-Sch%C3%B6nlein_purpura
3. Rai A, Nast C, Adler S. Henoch-Schönlein purpura nephritis. *J. Am. Soc. Nephrol* 1999 Dec;10(12):2637-44.
4. Kraft DM, Mckee D, Scott C : Henoch-Schönlein purpura-a review. *Am Fam Physician* 1998;58(2):405-8,411.
5. Saulsbury FT. Henoch-Schönlein purpura in children. Report of 100 patients and review of the literature. *Medicine (Baltimore)* 1999; 78(6):395-409.
6. Saulsbury FT. Henoch-Schönlein purpura. *Curr Opin Rheumatol* 2001; 13(1): 35-40.
7. Koizumi M, Hagino D, Fukuyama C, Abe K, Inoue K, Arai Y, Takechi K. Schonlein-Henoch

- purpura during pregnancy: case report and review of the literature. *J Obstet Gynaecol Res* 2004 Feb; 30(1):37-41.
8. S. Shrestha, N. Sumingan, J. Tan, H. Althous, L. McWilliam¹ and F. Ballardie : Henoch Schönlein purpura with nephritis in adults: adverse prognostic indicators in a UK population. *QJM* 2006; 99(4): 253-265.
 9. Ryder Hg, Marcus O: Henoch- Schonlein purpura : a case report. *S Afr Med J* 1976 Nov 20; 50(50): 2005-6.
 10. Weiss Pf, Feinstein JA, Iuan x, Burnham JM, Feudtner C. Effects of corticosteroids on Henoch-Schönlein purpura: a systematic review. *Pediatrics* 2007 ;120(5):1079-87.
 11. Blanco R, Martinez-Taboada VM, Rodriguez-Valverde V, Gracia-Fuentes M, Gonzalez-Gay MA. Henoch-Schönlein purpura in adulthood and childhood : two different expressions of the same syndrome. *Arthritis Rheum* 1997; 40(5): 859-64.
 12. Kalmantis K, Daskalakis G, Iavazzo C, Vranos A, Mesogitis S, Antsaklis A: Henoch-Schonlein purpura in pregnancy. *J Obstet Gynaecol* 2008 May; 28(4):403-5.
 13. Plauche WC: Henoch-Schönlein (anaphylactoid) nephropathy in pregnancy. *Obstet Gynecol* 1980; 56:515-517.
 14. Joseph G, Holtman JS, Kosfeld RE, Blodgett WA, Liu YK: Pregnancy in Henoch-Schönlein purpura. *Am J Obstet Gynecol* 1987;157:911-912.
 15. Ray M, Posen GA: Henoch-Schönlein purpura in pregnancy. *Can Med Assoc J* 1985; 132:1385-1386.
 16. Merrill J, Lahita RG: Henoch-Schönlein purpura remitting in pregnancy and during sex steroid therapy. *Br J Rheumatol* 1994; 33:586-588.
 17. Cummins DL, Mimouni D, Rencic A, Kouba DJ, Nousari Ch : Henoch-Schönlein purpura in pregnancy. *Br J Dermatol* 2003 Dec; 149(6):1282-5.
 18. Monica Ray and Gerald A Posen : Henoch-Schonlein purpura in pregnancy. *Can Med Asso J* 1985 June 15; 132(12):1385-1386).
 19. Robert Feldmann , Walter Rieger, Paul G Sator, Friedrich Gschnait, Friedrich Breier : Schönlein-Henoch purpura during pregnancy with successful outcome for mother and newborn. *BMC Dermatology* 2002; 2:1doi:10.1186/1471-5945-2-1.