PERIPARTAL CARDIOMYOPATHY

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Peripartal Cardiomyopathy is a disorder of heart muscle which presents clinically with the onset of heart failure in the last trimester of pregnancy or two weeks to five months after delivery.

Some authorities include only the post partal cases so as to exclude those patients with occult heart disease in whom failure was precipitated by stress of the last trimester of pregnancy (Hull and Hidden 1938, Melvin 1947). Post partal non valvular myocardial disease of unknown origin was known to physicians for more than a century (Virchow 1870). But this syndrome is uncommon and has been reported sporadically mainly in the American literature since 1937.

Hull et al and Gouley, McMillan and Bellet - (1937 -38) were the first to suggest its existence as a clinical entity and to stress the possible importance of puerperal factor in its etiology. Most of the reported cases have been amongst the Negro community in the U. S. A. There had also been reports of a few cases from Great Britain, West Indies, Africa, China, Holland, Brazil and India. The preponderance among the Negro community in the United States led some workers to suggest the importance of racial factor in its etiology. But this disease has been described in other races as well, and the increased incidence amongst the Negroes in Southern U.S.A. may be due to their comparatively lower socio-economic condition.

Etiology and Pathology

The exact etiology of this condition is not known, so various theories have been postulated. Suggested factors include:-

- (1) Immune response to foetal tissue.
- (2) Auto-immunity to the muscle product of rapidly involuting uterine muscle fibres.
- (3) Viral infection.
- (4) Nutritional deficiency.
- (5) Endoerinal imbalance.

Histological examination has revealed varying degree of hypertrophy of myocardial fibres with fibrosis, interstitial oedema and occasional focal accumulation of lymphocytes. These changes are almost indistinguishable from those of ordinary congestive cardiomyopathy.

On histochemical examination however the decrease of succinicdehydrogenase is much less marked as compared to alcoholic Cardiomyopathy. The absence of specific pathological changes and similarity in clinical picture to other types of congestive cardiomyopathy has led some workers to doubt the existence of this entity, but we think that it is important to maintain this syndrome as a separate entity for the following reasons.

(1) Peripartum cardiomyopathy has a definite time relationship with pregnancy, 93% of the cases have occurred in the post partal period and not in the period of maximal stress. (Seftel and Susser -1961)

(2) If cardiac failure occurs in successive pregnancies, it once again manifests in the post partal period.

(3) This syndrome has got certain clinical characteristics as for example it is more common in multiparous women and in toxæmia and twins (Meadows 1961). Heart size returns to normal in more than half of their patients of this syndrome (8 out of 14 cases described by Seftel and Susser in 1961). Such recovery is very uncommon with non alcoholic primary congestive cardio myopathy (Goodwin, personal communication).

Clinical features

There has been considerable uniformity in the clinical pattern described by various authors. Demakis et al, 1971 found Cardiomegaly and gallop rhythm in all their cases. Murmurs were inconspicuous Oedema was present in 48% of their cases. Initially ECG was abnormal in all their cases. The main changes being left ventricular hypertrophy with

inversion of T, wave, normal or low voltage QRS complex with inverted T waves or non specific ST-T changes. Embolism has been reported frequently and was the main cause of death in the cases reported by Gouley, McMillan and Bellet. Pulmonary embolism occurs more frequently than systemic, Gilchrist (1963) had described an anginal type as well.

Age and Parity

In Meadow's series the maximum incidence was found in third pregnancy. Most authorities have described the highest incidence amongst the highest age and parity group especially in women with many children born in quick succession. Susser and Stein showed that the age and parity of their patients were significantly greater when compared with those of the local child bearing population. There has however been a case report of paripartal cardiomyopathy in a 14 year old primipara (AHJ 72. 1968). Women with twin pregnancy are said to be more often affected (Hull and Hafkesbring 1937, Woodford 1952, Meadows 1957). Walsh et al 1965 have suggested inadequate nutrition to be an important factor in their series. Others have disputed this point and there was no evidence of malnutrition in any of the three patients described by Brown et al in British Heart Journal 1967.

Prognosis

There has been conflicting reports on the prognosis of this condition. Majority of the patients made final recovery in Melvin's series (1947); whereas in the series of Walsh et al the mortality was as high as 60%.

Case Reports

Case No. I

ND aged 25 years was admitted on 11. 9. 72 with the complaints of Dyspnoea & palpitation for one month & swelling of the legs since 2 weeks. She had also developed thrombophlebitis of left leg for last 10 days. She had normal delivery 2½ months before admission. During pregnancy she had mild dyspnoea and swelling of the legs in the last month which disappeared immediately after delivery. She was a second para and had no disability during or after the first pregnancy six years back. She had no history suggestive of heart disease in the past. She came from a lower middle class family and her diet was deficient in protein. She used to take alcohol occasionally in small amounts.

ON EXMINATION:-

Pulse 120/mt

B. P. 100/80 mm of Hg.

Gross evidence of congestive cardiac failure.

Apex 5th space $\frac{1}{2}$ " outside the mid-clavicular line.

Grade II/VI Pansystolic murmur in mitral area and Protodiastolic gallop.

INVESTIGATIONS:-

Hb 10.2 gm %

ESR 6 mm 1st hr.

Total protein 5.8 gms %

Albumin 3.9 gms

Globulin 1.9 gms:

Paper Electrophoresis different fraction showed normal ratio.

Blood sugar 120 mg %

Serum cholestrol 330 mg %

X-ray Chest showed moderately enlarged heart and left oblique view showed an enlarged left ventricle.

ECG showed low T waves with slight depression of ST Segments. She was put on bed rest and standard treatment of CCF. Heart size rapidly diminished and ECG became normal in about a months time. She has been followed for 5 months but slight enlargement of the heart persists.

Case No. 2

CM. Aged 22 years was admitted to the hospital on 18. 10. 72 with the complaints of breathlessness and swelling all over the body for the last two weeks. She had normal delivery on 31. 8. 72. She was a third para and she had no complaints during this or previous pregnancies. There was no history suggestive of heart disease in the past.

NUTRITIONAL HISTORY:- She came from a lower middle class family and her diet was obviously deffeient in protein. She did not take any alcoholic drinks.

ON EXAMINATION

Pulse 100/mt

BP 105/85 mm of Hg.

Apex beat in left 5th space $\frac{1}{2}$ " out from the mid clavicular line.

Grade II/VI systolic murmur maximum in the pulmonary area.

There was a distinct presystolic gallop. She was in gross CCF with oedema of legs, engorged neck veins, enlarged tender liver and crepitations at the bases of both lungs.

INVESTIGATIONS

Hb 9.5 gms.

ESR 35 mm 1st hr.

Blood sugar 100 mg%

Blood urea 18 mg%

Serum cholesterol 250 mg%

Total protein 5.3 gms%

Albumin 3.1 gms.

Globulin 2.2 gms:

Paper Electrophoresis- different fraction showed normal ratios.

Urine NAD

Chest X-ray showed markedly enlarged heart with enlargement of left ventricle.

ECG showed inverted T waves in lead I, II and V - V₁ - V₆. She was put on absolute bed rest and standard treatment for CCF. She made a remarkable recovery and her heart size became normal on 1.12.72 ECG also showed marked improvement.

Case No. 3

CV, aged 35 years was admitted in Bir Hospital on 23.10.72 with the complaints of breathlessness, palpitation, swelling of the legs and chest pain for the last 10 days. She had her delivery on 28.6.72 which was normal except for retained placenta for which manual removal had been done. She was a 4th para and there was no history of any disability during this or previous pregnancies. There was nothing in the past history to suggest heart disease and family history also was not significant. She came from a low socioeconomic group and her diet was deficient in protein. There was no history of excessive intake of alcohol.

ON EXAMINATION

Pulse 84/mt

BP 110/80 mm of Hg.

Heart was enlarged clinically and there was grade II/VI systolic murmur, maximum in 3rd left space and there was a protodiastolic gallop. She was in obvious CCF.

INVESTIGATIONS

HB 9.2 gms.

Blood count normal.

ESR 22 mm 1st hr.

Blood sugar 118 mg %

Serum cholesterol 260 mg %

Total serum protein 7.8 gms %

Albumin 2.2 gms.

Globulin 5.6 gms:

Electrophoresis showed raised β level

X ray Chest Showd enlarged heart.

ECG revealed Rt. Bundle Branch Block with inverted T waves from V₁ - V₅. She was

kept on absolute bed rest and usual treatment for CCF were administered. She made a remarkable recovery after a month, there was no evidence of failure. Heart size returned to normal, T waves became upright in V₄ & V₅ Bit Rt. Bundle Branch block persisted.

Discussion

All our cases were multiparous. The clinical features were similar to those described by other workers. In one case the symptoms started in the last month of pregnancy and in the two remaining cases started during the post partal period. In the two post partal cases the heart size returned completely to normal but in the first case the heart was a little enlarged even after five months. All these cases came from the low socioeconomic group with deficient protein diet. The serum protein was low in first two cases and in the third case although the serum protein was normal, the albumin was only 2.2 gms. So it seems very likely that protein deficiency may have an important role in the genesis of peripartal cardiomyopathy.

The serum cholesterol was high in first and third case and was in the upper limit of normal in the second case. Serum cholesterol is usually much lower in women of this age and socioeconomic group in Nepal. Hypercholesterolaemia has also been reported by Walsh et al in their series of ten cases. Further studies of detailed lipid and lipoprotein pattern in peripartal cardiomyopathy may provide a clue to the etiology of this condition.

All our cases showed a very remarkable response to therapy and in two cases the heart size became normal rapidly and in the remaining cases also it came down to almost normal

size. In Meadow's series of seventeen cases the heart size came down to normal in only 8 cases these had a very good prognosis and were found to be unlikely to develop peripartal cardiomyopathy in subsequent pregnancies. In the remaining cases where cardiomegaly persisted, the average survival was only 4.7 years and these cases were likely to develop peripartal cardiomyopathy in subsequent pregnancies. We insisted on our patients to have absolute bed rest and this early institution of complete rest is probably an important determinant factor in the favourable response, of our cases. We found all the three cases unduly sensitive to Digitalis, so this drug has to be given cautiously in this condition. Potassium supplement should be given routinely in all cases to prevent digitalis toxicity. One of our cases developed thrombophlebitis. This is significant in view of the comparative rarity of this complication in our country. Pulmonary and systemic embolism has indeed been reported by many workers and has been the cause of death in many cases. So anticoagulant therapy is worth trying. Further pregnancy should be avoided if heart size has not returned to normal. But oral contraceptive should not be given in view of the increased risk of thrombosis.

Summary.

1. 3 cases of Peripartal cardiomyopathy seen in Bir Hospital during the last seven months have been described with review of the literature.
2. Low Protein diet & Hypoproteinaemia is probably an important factor in the genesis of this condition.
3. These cases were found to be unduly sensitive to Digitalis, so this drug should be given cautiously & routine Potassium supplement is advocated.
4. All the cases responded very well & this favourable outcome is most probably due to early institution of absolute bed rest in the hospital.

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