Severe Intrauterine Fetal Growth Restriction Associated with Fetal TAR Syndrome and Primary Hypothyroidism.

Majumdar *Subrata*, Kundu *Sarmila*, Dutta Roy *Chaitali*, Mukherjee *Sabyasachi*
Institution of Post Graduate Medical Education and Research and SSKM Hospital, Calcutta 700020.

Key words : IUGR, Fetal TAR syndrome, primary hypothyroidism

Congenital anomalies of the fetus are an important cause of fetal growth restriction. Thrombocytopenia with absent radii (TAR syndrome) is a rare type of structural abnormality of the fetus. We report a case of TAR syndrome with unilateral absence of radius associated with primary hypothyroidism responsible for intrauterine fetal growth restriction. Mrs A G 27 year old primigravida married for 2 years with LMP 27-10-2000 and EDD 3rd August 2001 first attended our antenatal clinic on 11th November 2000 with amenorrhoea of six weeks. She was advised routine investigations, folic acid supplementation of 5 mg daily and monthly antenatal checkup. First trimester and early 2nd trimester were uneventful. USG (4-3-2001) showed a single live fetus of 18 weeks corresponding to LMP with adequate liquor. Placenta was located on anterior wall of uterus away from the internal os. Subsequent antenatal visits showed -

	Weight (Kgs)	BP (mmHg)	Fundal height (weeks)	Gestational Weeks	Abdominal girth (cms)	Symphysis Fundal height	USG
18th Mar 2001	58	120/80	20	-	-	-	
14th April 2001	60	114/76	24	-	-	-	
26th May 2001	60	120/80	24	28	83	24	Single live fetus of 24 weeks
10th June 2001	60	116/76	24	30	84	25	

As her weight gain and fundal height were stationary, she was admitted on 10th June for observation and advised bed rest, high protein nutritious diet, 100 gms of glucose per day, hematinics and calcium supplementation along with biweekly infusion of amino acid solution and 10% fructodex. USG done on 15th June showed maturity of 28 weeks (33 weeks by LMP). Liquor was diminished. Subsequent examinations showed -

	Wt (kgs)	BP (mm Hg)	Fundal height (weeks)	Gestational Weeks	Abdominal girth (cms)	Symphysis Fundal height	USG
17th June 2001	60	110/70			84	26	Maturity of 28 weeks
30th June 2001	61	116/76	26	35	85	26	Maturity of 30wks (35wks by LMP) with decreased liquor
12th July 2001	61	120/80	28	36+	85	29	

On daily clinical examination, fundal height was static and the amount of liquor was found to diminish. The patient complained of less fetal movements since 10th July 2001. She was given moist O_2 inhalation, rest in left lateral position, and Inj. sodium bicarbonate 40 ml and kept under strict observation.

Emergency cesarean section was done on 12th July 2001 for fetal distress and severe fetal growth restriction. A female baby in breech presentation was delivered in an

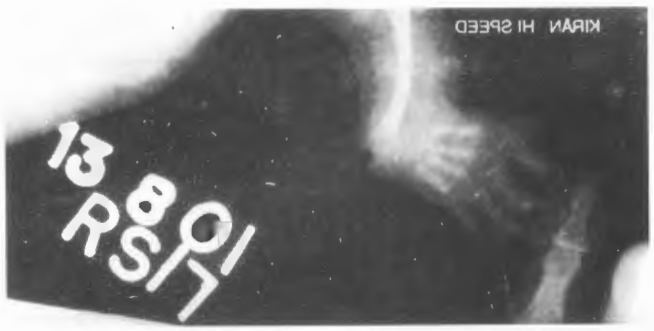
Paper received on 9/1/02 ; accepted on 3/5/03

Correspondence :
Majumdar Subrata
Institution of Post Graduate Medical Education and Research and SSKM Hospital, Calcutta 700 020.

asphyxiated condition with Apgar scores of 4 and 8 at 1 and 5 minutes respectively. Liquor was thickly meconium stained. Placenta showed sings of calcification.

Examination of the baby revealed birth weight 1 kg, length 37 cm, head circumference 30 cm and abdominal circumference 27 cm. Right forearm was shortened and curved. Radius was absent. Right thumb was attached by a fleshy flap. Petechial spots were seen all over the body. CNS examination showed diminished reflexes and activity. Platelet count – 85,000 cmm. TSH 2.9 / mIU / ml (Normal < 25) T4-10.8 ugm / ml (Normal 11-34). Echo cardiography was normal.

The baby was diagnosed as a case of thrombocytopenia with absent radius (unilateral) i.e. TAR syndrome along with primary hypothyroidism. She was given platelet transfusion along with thyroxin and she gradually improved. Reconstructive surgery for the right upper limb was to be considered at a later date.



Photograph - 1 : Xray Right arm showing absence of radius, 1st metacarpel and 1st phalanyx (absent right thumb)

Photograph - 2 : New born showing deformity of the right forearm and hand with absent right thumb.

Discussion

Thrombocytopenia with absent radius (TAR syndrome) is a very rare type of skeletal anomaly associated with congenital thrombocytopenia, the incidence being 1 in 500000 to 1 in 1000000 newborns[1]. The pathognomonic feature is bilateral absence of radii with presence of thumbs. Although most cases are bilateral, absence of only one radius has also been described[2]. Associated cardiac, renal and other skeletal malformations also may occur.

Infants with major structural anomalies are more commonly associated with intrauterine growth restriction[3]. Our baby also suffered from primary hypothyroidism. It is found that severe growth restricted fetuses may have a low T4 and high TSH value[4].

In our case, there were two risk factors, congenital anomaly (TAR syndrome) and primary hypothyroidism, both of which are probably responsible for severe intrauterine growth restriction.

References

1. Weidemann HR, Kunje J eds. Clinical Syndromes. 3rd Edn. Italy. Times Mirror International Publishers Limited. *1997 pg 430.*

2. Alter BP, Young NS. The Bone Marrow Failure Syndromes. In Nathan DJ and Orkin SH eds. Nathan and Oski's Hematology of Infancy and Childhood. 5th Edn. USA W.B. Saunders Company. *1998 pg 307.*

3. Khoury MJ, Erickson JD, Cordero JF et al. Congenital malformation and intrauterine growth retardation. A Population Study. *Pediatrics 1988; 82:83.*

4. Thrope-Beeston JG, Nicolaides KH, Snijders RJM et al. Thyroid Function in small for gestational age fetuses. *Obstet and Gynecol 1991; 77:701-6.*