tion. IgM does not reliably persist for more than six weeks after a primary infection, let alone a reinfection where only small amounts of IgM may be produced. The pregnancy proceeded uneventfully thereafter.

The baby weighed 2010 g at birth (below the third centile); his length was also below the third centile, and the head circumference just above it. At 2 months of age he had a dense right sided cataract associated with a right microphthalmia, signs of a small ventricular septal defect, hepatosplenomegaly, and profound sensorineural deafness confirmed by electrocochleography and brainstem evoked response audiometry. He was hypertonic with marked head lag and showed signs of delayed development. His head circumference had fallen below the third centile.

A blood sample from the baby at 10 weeks was strongly positive for rubella IgM antibody, and negative for cytomegalovirus and toxoplasma antibody.

Discussion

Both the cases described above represent proved subclinical rubella infections in previously immune pregnant women, which resulted in adverse outcomes. This confirms the work of others,[1 2] that such infections do not have to be clinically apparent to harm the fetus. In neither mother were we able to establish whether their infection occurred after immunity that had been acquired naturally or after vaccination. We did have a report of vaccination in 1979 for the first mother, but no knowledge of any previous vaccination or infection.

If these cases were reinfections it is possible that the quality of antibody produced in women in whom seroconversion has occurred after vaccination may be inadequate for total protection compared with that induced by natural infection. If this is the case we can expect more reinfections with rubella that will produce fetal damage while natural rubella virus is still circulating, despite the recent introduction of the measles, mumps, rubella (MMR) vaccine. Harcourt *et al*,[3] for example, found significant differences in rubella specific IgG, IgA, and IgM responses on subsequent challenge with rubella vaccine between volunteers in whom previous immunity had been induced by vaccine and those who had natural immunity.

Although O'Shea *et al* found that only one of 19 volunteers with low concentrations (<15 IU) of pre-existing immunity induced by rubella vaccine, and none of 12 with previous low natural immunity, had detectable viraemia after intranasal challenge with RA27/3 rubella virus vaccine,[4] it is possible that this low incidence of viraemia might increase when the challenge is with natural virus.

Prospective surveillance of congenital rubella infections and their association with previous maternal immunisation will therefore need to be continued for many years to come.

Further studies are also indicated to determine the risks to the fetus, however small, in infections where there is previous antibody resulting from vaccination. We must, however, stress that rare events such as these should not detract from vigorous advocacy of the MMR vaccine.

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Sturge-Weber and Klippel-Trenauny syndromes with absence of inferior vena cava

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Abstract

A baby girl born at 33 weeks' gestation weighing 2250 g presented with Sturge-Weber syndrome, features of the Klippel-Trenauny syndrome, and absence of the inferior vena cava. We suggest that aplasia of the vena cava may be a feature of Klippel-Trenauny syndrome when the capillary malformation affects the trunk.

We report a case of Sturge-Weber syndrome with extensive capillary malformation over the trunk and features of Klippel-Trenauny syndrome, together with congenital absence of the inferior vena cava. There is to our knowledge only one previous report of Sturge-Weber syndrome (associated with an anomalous (double) inferior vena cava) and in this instance there were also features of Klippel-Trenauny syndrome.[1]

The Klippel-Trenauny syndrome can be associated with anomalies of the deep venous drainage of the limbs, including aplasia and duplication.[2] Servelle, in his extensive experience of 786 cases with Klippel-Trenauny syndrome and venous malformations, encountered four cases with aplasia of the lower portion of the inferior vena cava.[3]

The patient showing port wine capillary malformation.

Case report

A baby girl was born at 33 weeks' gestation weighing 2250 g after spontaneous onset of labour. Pregnancy had been complicated by bleeding in the 13th week, and polyhydramnios diagnosed clinically and by ultrasound scan at 31 weeks. Labour and delivery were normal.

She had a particularly widespread port wine capillary malformation (figure), virtually the whole face below the supraorbital ridges being affected. The lesion also extended onto the left frontal region, and backwards over both sides of the head to the occiput. The right arm and the back were completely covered, and the lesion extended forwards over the greater part of the trunk. There were extensive patches on the left arm and right leg, but the left lower limb was largely unaffected. Several distended veins crossed the abdomen, the largest extending from the right groin to the xiphisternum. Blood flowed towards the head. There was no bony hypertrophy of the extremities, but the diameters of the right arm and leg were slightly greater than the left.

The platelet count was normal. Ultrasound scan and computed tomography with contrast enhancement showed that the abdominal organs were normally positioned. A thin tubular structure ran behind the liver as far as the renal pedicles, which was consistent with either a small segment of inferior vena cava or with a hypertrophied portion of azygous vein.

Postnatal progress was unremarkable, and she was discharged after three weeks. At follow up the skin lesion remained unchanged. At the age of 11 months, and three days after being given the third dose of diphtheria, tetanus, pertussus, and polio vaccines, she was readmitted to hospital with a left sided convulsion complicated by Todd's paralysis. Over the next few days several further seizures, some of which were prolonged, were successfully treated with intravenous diazepam, and maintainance treatment with sodium valproate was started. Despite an intermittent fever during this period, bacterial and viral cultures grew no pathogens. Skull radiographs were normal. Computed tomography of the brain with contrast revealed dramatic gyral enhancement in the right parietal and occipital region consistent with a large superficial vascular malformation.

On review at the age of 12 months, function in the left side had recovered, although she remained right handed as she had been throughout life. There was no spasticity, and hand-eye coordination was normal. She was not yet crawling or cruising, but could bottom shuffle with reasonable efficiency. Vocalisation was normal for age, and she could use three words appropriately. Ophthalmic examination was normal.

Discussion

This case fulfils the criteria for Sturge-Weber syndrome. In addition, the concurrence of superficial varicosities with extensive capillary malformation suggests the Klippel-Trenauny syndrome.

Klippel and Trenauny considered skeletal hyperplasia (not present in the present case) to be an indispensable diagnostic feature of their syndrome.[4] Other authors have been less fastidious.[2] Servelle argued that bony hyperplasia is a secondary phenomenon occurring as a result of venous hypertension, which is in turn caused by abnormalities in the deep veins[3]: if that is so, this case corresponds to the Klippel-Trenauny syndrome in essential features.

The superficial varicosities (and deep venous anomalies) found in Klippel-Trenauny syndrome generally occur in the limb affected by the capillary malformation. When the capillary malformation is truncal, absence of the vena cava may be an analogous phenomenon. We support the view of Schofield *et al* that, where such a malformation exists, it may be wise to visualise the venous drainage before embarking on a major abdominal operation.[1]

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