LETTERS TO THE EDITOR

A compound heterozygous state result in moderate to severe hypercalcemia in the neonatal period labeled as NSHPT. Most neonates with NSHPT require subtotal or complete parathyroidectomy.

In view of presentation in the neonatal period, symptomatic hypercalcemia and low to normal urinary calcium excretion, we believe that an inactivating mutation of the CaR was responsible in both the above patients. A primary adenoma of the parathyroid gland, as proposed by Shah and Shah(1), would on the other hand, be associated with increased urinary calcium excretion (proportionate to the degree of hypercalcemia). Careful assessment of urinary calcium excretion (on one or more occasions) is thus useful in differentiating a primary adenoma of the parathyroid gland from an inactivating receptor mutation. Estimation of blood and urinary levels of calcium in parents and siblings may also provide important information.

We emphasize that most cases of primary hyperparathyroidism in neonates and infants are due to a defect in CaR receptor rather than a primary adenoma(5). An extensive radiological evaluation of parathyroid glands is rarely necessary. The management strategy in such patients chiefly depends on the severity of the metabolic abnormalities.

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A Rare Cause of Congestive Heart Failure in Newborn

Arteriovenous malformations (AVMs) are rarely seen in neonates and most often present with congestive heart failure (CHF). We report a newborn with congestive heart failure due to AVM in the posterior fossa.

Arteriovenous malformations (AVMs) are rarely seen in neonates and most often present with congestive heart failure (CHF). We report a newborn with congestive heart failure due to AVM in the posterior fossa.

A term male baby was admitted to neonatology department with respiratory distress at 5 hour of life. On physical examination perioral cyanosis, dyspnea, tachypnea and a grade 3/6 systolic ejection murmur at left lower sternal border were noticed. These factors lead to the diagnosis of CHF. On echocardiography only right atrial, ventricular and superior vena cava
dilatation was observed with no other cardiac anomaly. Cranial auscultation revealed a bruit over right frontoparietal area which necessitated further evaluation with cranial ultrasound and doppler. An AVM was detected and diagnostic angiography confirmed the presence of an AVM supplied by many arteries and draining to the lateral mesencephalic and inferior petrosal sinuses on the anterior surface of the right cerebellar hemisphere and pontomesencephalic corner. Because of its location over the brainstem and huge dimensions it was regarded inappropriate for endovascular surgery and the patient was managed conservatively. He was ventilated mechanically, received inotropic agents and diuretics for CHF but died on 19th day of life.

AVMs which can occur congenitally or can be acquired are non-neoplastic conglomerate vascular abnormality. They are formed by joining of the arteries and veins without lamina elastica by thin channels. AVMs are most commonly seen in cerebral hemispheres (65-85%). According to a published series, 10-18% of AVMs are located in the posterior fossa(1). Clinical manifestations of AVMs are CHF, hemorrhage, convulsions, focal neurological signs, hydrocephaly and macrocephaly(1). AVM’s are rare cause of CHF in the newborn(2-4). Doppler ultrasonography, computed tomography, MRI and MR angiography are useful tools in the diagnosis(1). However, angiography is the most important imaging technique that aids planning the appropriate management(1). Treatment of AVM may be surgical, endovascular therapy, radiosurgery or a combination of all. Although many cases treated by endovascular approach has been reported, this is not the standard therapy for AVMs(5).

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