of high fever in warm environment, dry skin, sparse hypopigmented hair, frontal bossing, flattened nasal bridge are some of other features. Poor development of mucous glands in the respiratory tract may result in increased susceptibility to respiratory tract infections. Treatment of these children include protecting them from high ambient temperatures and early dental evaluation so that prostheses can be provided. Artificial tears should be used to prevent the damage to cornea. Wigs may be advised to improve appearance. (1) Both the brothers reported here had the classical triad along with other clinical features of hypohidrotic (anhidrotic) ectodermal dysplasia which was confirmed by skin biopsy. The mode of inheritance of hypohidrotic ectodermal dysplasia is X-linked recessive with full expression only in males; however, an autosomal recessive mode of inheritance may be operative in some families (1,5).

REFERENCES

1. Gray LD. Ectodermal Dysplasia. In: Nelson Textbook of Pediatrics. Behrman RE, Kliegman RM, Jenson HB, editors. 16th edn. W.B.Saunders Company 2000; 1974-1975.
2. Smagrupps Centrum - the Swedish Information Center for Rare Diseases- produced and edited this information material. Date of publication: 2005-05-03 Version: 2.1 Article number: 2002-110-10
3. Priolo M, Lagana C. Ectodermal dysplasias: a new clinical-genetic classification. J Med Genet. 2001; 38: 579-585
4. Sepulveda W, Sandoval R, Carstens E, Gutierrez J, Vasquez P. Hypohidrotic ectodermal dysplasia: prenatal diagnosis by three-dimensional ultrasonography. J Ultrasound Med. 2003; 22: 731-735.
5. Munoz F, Lestringant G, Sybert V, Frydman M, Alswaini A, Frossard PM, Jorgenson R, Zonana J. Definitive evidence for an autosomal recessive form of hypohidrotic ectodermal dysplasia clinically indistinguishable from the more common X-linked disorder. Am J Hum Genet. 1997; 61: 94-100.

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LETTER TO EDITOR (VIEWERS CHOICE)

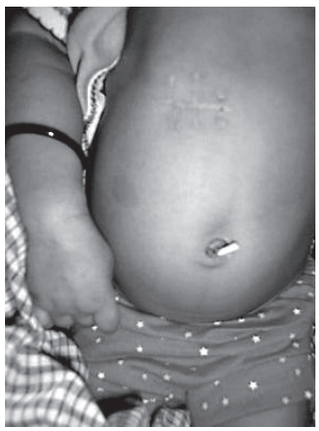
VENTRICULOPERITONEAL SHUNT CATHETER MIGRATION THROUGH UMBILICUS - A RARE COMPLICATION

Dipankar Sarkar, Shruti Sarkar

A 9 month old baby was brought to our outpatient department with the complaint of a plastic tube protruding out of abdomen through umbilicus for a few days. The baby had undergone Ventriculoperitoneal shunt surgery nearly 4 months back for congenital hydrocephalus in another hospital. He was born at term by caesarean section and was diagnosed to have congenital hydrocephalus soon after birth. The baby had recovered well from surgery, was feeding well on demand and thriving. On examination the baby looked well and the VP shunt catheter was seen protruding out of umbilicus (Figure 1). On pressing the shunt reservoir behind the ear, fluid started dribbling out of the tube protruding through the umbilicus confirming the protruding end to be the distal end of the VP shunt. Systemic examination apart from this did not reveal any other abnormality. The infant along with his parents had come from a distant village. The parents were explained in detail the occurrence of this complication and the need for correction by surgery. They declined corrective surgery and were lost to subsequent follow up.

Migration or protrusion of the ventriculoperitoneal shunt catheter to various sites is not very uncommon and has been reported in several case reports. Cases of shunt migration into stomach (1), bowel (2), liver (3), chest (4), jugular vein (5), anus (6), vagina (7), and scrotum (8) have been reported. In 1973 Adeloye et al reported a 9 month old baby with shunt migration through umbilicus within 2 months after surgery (9). Another case has been reported from Srinagar, India in 2000 where an 18 month old child presented with low grade fever, irritability, abdominal pain along with the distal end of VP shunt coming out through the umbilicus (10) .This particular complication can be explained on anatomical basis. Umbilicus is a centrally situated scar

Figure 1: Distal end of VP shunt protruding from the umbilicus



and it becomes an area of anatomical weakness when there is a foreign body inside the abdomen. There are five ridges in the peritoneum that line the lower part of the anterior abdominal wall, which converge in the midline at the umbilical region. Peristaltic activities can thus direct foreign bodies in the peritoneal cavity toward the umbilicus and push it to come out (9). Migration of the lower end of the shunt catheter is an infrequent problem, which occurs without any recognizable cause. The reported incidence of distal shunt migration is 10% (11). Of all the sites of distal end migration reported so far umbilicus is very rare.

References

1. Alonso-Vanegas M, Alvarez JL, Delgado L, Mendizabal R, Jimenez JL, Sanchez-Cabrera JM. Gastric perforation due to ventriculo-peritoneal shunt. Pediatr Neurosurg. 1994; 21: 192-194
2. Wilson CB, Bertan V. Perforation of the bowel complicating peritoneal shunt for hydrocephalus. Report of two cases. Am Surg. 1966; 32: 601-603.
3. Touho H, Nakauchi M, Tasawa T, Nakagawa J, Karasawa J. Intrahepatic migration of a peritoneal shunt catheter: case report. Neurosurgery. 1987; 21: 258-259
4. Cooper JR. Migration of ventriculoperitoneal shunt into the chest. Case report. J Neurosurg. 1978; 48: 146-147
5. Cowman MA, Allen MB Retrograde migration of venous catheter as a complication of ventriculoatrial shunt in adults. Case report. J Neurosurg . 1971; 35: 348
6. Schulhof LA, Worth RM, Kalsbeck JE. Bowel perforation due to peritoneal shunt. A report of seven cases and a review of the literature. Surg Neurol. 1975; 3: 265-269
7. Patel CD, Matloub H. Vaginal perforation as a complication of ventriculoperitoneal shunt. Case report. J Neurosurg. 1973; 38: 761-762
8. Ramani PS. Extrusion of abdominal catheter of ventriculoperitoneal shunt into the scrotum. Case report. J Neurosurg. 1974; 40: 772-773
9. Adeloye A. Spontaneous extrusion of the abdominal tube through the umbilicus complicating peritoneal shunt for hydrocephalus. Case report. J Neurosurg. 1973; 38: 758-760
10. Wani AA, Ramzan A, Wani MA. Protrusion of a peritoneal catheter through the umbilicus: an unusual complication of a ventriculoperitoneal shunt. Pediatr Surg Int. 2002; 18: 171-172
11. Kanojia R, Sinha SK, Rawat J, Wakhlu A, Kureel S, Tandon R. Unusual ventriculoperitoneal shunt extrusion: experience with 5 cases and review of the literature. Pediatr Neurosurg. 2008; 44: 49-51

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LETTER TO EDITOR (VIEWERS CHOICE)

CONGENITAL ADRENAL HYPERPLASIA IN A MALE CHILD

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An one and half month old male baby was brought in cardio respiratory arrest with a history of noisy breathing for 3 days and several episodes of vomiting and watery loose motions for 1 day. Baby was immediately resuscitated. His birth weight was 2.5 kg and perinatal course was uneventful. There was one previous hospitalization at day 21 of life with acute gastroenteritis and sepsis. Physical examination revealed an emaciated baby of 1.75 Kg (< 3 rd centile) with normal systemic examination and genitalia. Investigations revealed normal hemogram, hypoglycemia (Blood sugar = 24mg/dl), positive CRP, hyponatremia (serum sodium of 118 mEq/dl) and hyperkalemia (Serum potassium = 6.5 mEq/L). These investigations lead us to suspicion of Congenital Adrenal hyperplasia (CAH). Steroid levels were done which showed Serum Cortisol to be low (25.13, normal range = 28-662 nmol/l) and Serum 17 Hydroxy Progesterone to be high (8800.0 ng/dl, normal: <100 ng/dl) thus confirming the diagnosis of 21 hydroxylase deficiency. The child was managed with antibiotics, Fludrocortisone and hydrocortisone. Child improved, gained a weight of 500 grams over 10 days of hospital stay. Serum electrolytes normalized (Serum sodium of 140mEq/L and serum potassium of 5.5mEq/L) and child was discharged. At 3 months of follow-up he weighs 3.2 kg and electrolytes were normal.

Congenital adrenal hyperplasia is a group of autosomal recessive disorders resulting from the deficiency of one of the five enzymes required for the synthesis of cortisol (1). This disease is easily clinched in female newborns where the ambiguous genitalia are present, but the diagnosis in male newborns is often overlooked as was in our case. He was admitted at 21 days of life and treated as acute gastroenteritis with sepsis and diagnosis of CAH was missed. There are three types of CAH due to 21-hydroxylase deficiency: classical salt wasting disease which is most severe, both cortisol and aldosterone are deficient, classical simple virilizing disease in which adequate levels of aldosterone are synthesized but adrenal androgens are elevated and non-classical disease in which adrenal androgens are mildly elevated leading to signs of androgen excess after birth. Progressive weight loss, anorexia, vomiting,