Abstract

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A Case Report

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Between all specialties of neurosurgery pediatric and developmental surgery is most valuable one, it can make a difference and changing the destinies.

Among hundreds of ordinary cases there are some unusual cases which need more research and investigations for get most of benefit for the patients and the surgeons.

months old female patient was presented with inspective progressive enlarging of head and delayed in activities with poor feeding, her delivery was by CS (due to large head circumference)

At age of 1 month she was admitted to hospital for hydrocephalus and the decision was placement of VP- shunt, after preoperative work up others anomalies were revealed hydrocephalus ± Dandy walker malformation, cleft lip palate, 2.8 mm membranous VSD, hepatosplenomegaly, four limbs rigidity (stiffness), pediatric consultation (she was dehydrated and poor feeding status) and his opinion was to delay our intervention, Later patient examination found increasing in head circumference, and other signs of hydrocephalus were founded with declining in primitive refluxes (suckling, grasp, monro), CT scan Finding of ballooning signs and expansion of each lat, 3rd and 4th with missing of cerebellar vermian and carpus callosum. Patient also had cleft lip and palate, distended abdomen, limitation of active and passive movement in her 4 limbs. Surgery done by classical open way VP- shunt technique ventricle access smoothly with high pressure ICP. Day 2 Patient got intestinal perforation by device lower end, second surgery urgently was done with changing the device completely and repairing of the colon (by Para median laparotomy incision) with copious irrigation of the cavity with saline and garamycin. After complete the surgery patient was admitted to ICU with mechanical ventilation. Patient had cardiac arrest and complete apnea for few minutes and then she was came back by CPR. Patient died next day.

Probably the diagnosis is hydrocephalus-cleftpalate-joint contractures syndrome

(Aase-Smith syndrome I) is a rare genetic disorder characterized by a buildup of fluid in the brain (hydrocephalus) due to a brain abnormality called Dandy-Walker malformation, cleft palate, and stiff or "frozen" joints (contractures). Less than 20 cases of hydrocephalus-cleft palate-joint contractures syndrome have been reported. Other symptoms might include: thin fingers with absent knuckles and reduced creases over the joints, ear abnormalities, heart defects, and clubfoot. The cause of hydrocephalus-cleft palate-joint contractures syndrome is not known, but it is likely genetic due to reports of affected family members and likely autosomal dominant inheritance. Treatment is specific to the symptoms present in each individual and might include surgical correction of birth defects such as cleft palate and clubfoot. [1] [2] [3]

Keyword : hydrocephalus , VSD , aase-smith syndrome I

Biography

Nibras Alsumaidaee is a neurosurgeon from Iraq, he had his training courses during the requirement of the Arabic board in neurosurgery in Baghdad Medical Complex (Martyr Gazi Al-hariry for specialized surgical hospital),Now he is specialist neurosurgery senior in neurosurgery teaching hospital, his interest mainly toward neuro-oncology, functional neurosurgery, endoscopic neurosurgery, he will keep going in the way of science, researching and training in spite of the hard situation and practice obstacles in his country.

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