Correspondence

History Taking: Still a Valuable Tool in Today’s Era of Modern Medicine

Sir,

A 20 years old male was referred to our institute for sudden onset of unconsciousness not preceded by headache, fever or convulsions. There was no history of head injury or alcohol consumption. He had not suffered from any major illness in past and never had seizures before. Clinical examination revealed an unconscious patient. The vital parameters were normal except tachycardia 116/minute and tachypnoea 24/minute. The skin colour was normal. There was no evidence of cyanosis. There was no local neurodeficit. All routine laboratory parameters including blood sugar level and arterial blood gases were normal. Electrocardiogram (ECG), magnetic resonance imaging of brain (MRI), CSF examination, 24 hours Holter monitoring and serum cholinesterase levels done subsequently were normal. Electroencephalogram (EEG) done was within normal limits. Patient responded to supportive measures and supplemental oxygen.

Later on again after taking indepth history from relatives, it was revealed that patient had taken prolonged bath for more than 30 minutes in a stuffy bathroom in which a gas heater was recently installed. On the basis of history and ruling out other structural lesions or metabolic causes, he was diagnosed as most likely case of “Gas Heaters Encephalopathy”. It could have occurred because of taking prolonged bath in a poorly ventilated bathroom where combustion of gas from on Liquefied Petroleum gas (LPG) unit, which itself is a combination of butane and propane producing CO and H2O resulting in secondary hypoxia due to carboxyhaemoglobin formation.1

Recently in 40 such cases observed so far in Pune City all had similar history of taking bath for at least average 7 minutes and regaining consciousness after being brought out of bathroom and were asymptomatic later. All had a gas heater with cylinder and burner fitted inside their bathroom. There was no injury, tongue bite, neuroimaging and EEGs were normal and symptoms did not recur after removing geyser from bathrooms.

Awareness of Gas heater encephalopathy in treating doctors would help to minimize unnecessary investigations and antiepileptic treatment, it would also help in educating others that though cost effective, but if burnt in insufficient air, gas heater may be risky by producing carbon monoxide (CO) and it should better be fitted in well ventilated bathrooms with good CO sensors.2

References

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Klippel-Feil Syndrome with Unilateral Renal Agenesis and Renal Failure

Sir,

The triad of short neck, low posterior hairline and severe restriction of cervical motion is a classic definition of Klippel-Feil syndrome. Klippel-Feil syndrome (KFS) is characterized by congenital fusion of two or more cervical vertebrae and is believed to result from faulty segmentation along the embryo’s developing axis during the second to eighth week of gestation. This is commonly associated with many congenital malformations. The anatomic and clinical expressions of this syndrome vary widely, ranging from mild cosmetic deformity to severe disability and end stage organ diseases such as renal failure. Hereby, we report a patient with KFS who had unilateral renal agenesis and presented with end stage renal disease.

Diagnosis of KFS should stimulate careful search for other associated abnormalities.

A 27 year old male, unmarried from rural background was admitted with complaints of progressive generalized weakness, dyspnea on exertion, puffiness of face for one month and severe anorexia, hiccoughs and decreased urine output for about a week. When his history was traced back, he had short neck, slight facial dysmorphism and restriction of neck movements since childhood. There had been no major trauma, surgery, systemic illness or prolonged treatment in the past. His parents were alive with no significant illnesses. On examination, his height was 146 cms. There was severe pallor, anasarca, limitation of neck extension and rotation, low hair line, scoliosis and Sprengel’s deformity of the left scapula. His blood pressure was 180/90 mmHg. Abdomen examination revealed presence of ascites and auscultation of chest showed presence of few basal

Fig. 1: A case with Klippel-Feil syndrome. Clavicles are not at the same level
crepitations. He had ejection systolic murmur at pulmonary area. Neurological examination was unremarkable.

Biochemical investigations showed that he had advanced renal failure and hyperkalemia. On the basis of history, clinical examination, laboratory investigations and imaging he was diagnosed to have KFS (Figures 1 and 2) with end-stage renal disease and unilateral agenesis of left kidney (Figure 3). Since he had serum creatinine 8mg/dl and urea 226 mg/dl, contrast studies could not be performed. He was treated conservatively with renal replacement therapy.

KFS occurs in a heterogeneous group of patients unified only by the presence of a congenital defect in the formation or segmentation of the cervical spine. Three types of KFS have been described. Type 1: Cervical spine fusion in which elements of many vertebrae are incorporated into a single block. Type 2: Cervical spine fusion in which there is failure of complete segmentation at only one or two cervical levels and may include an occipito-atlantal fusion. Type 3: Type 1 or type 2 fusion with co-existing segmentation errors in the lower dorsal or lumbar spine.

The importance of recognizing KFS lies in the fact that there is a strong association of this anomaly with significant abnormalities of other systems in the body. These include skeletal system abnormalities, such as, scoliosis and/or kyphosis (60%), Sprengel deformity of the scapula (30%), and torticollis, urinary system abnormalities (35%), loss of hearing (30%), facial asymmetry and flattening of neck (20%), synkinesis or mirror movements (20%), congenital heart diseases (4.2 - 14%). Brain stem anomalies, congenital cervical stenosis, adrenal aplasia, ptosis, lateral rectus muscle paralysis, facial nerve paralysis, syndactilia, and diffuse or focal hypoplasia in the upper extremities may also be seen.

Our patient had Sprengel’s deformity and unilateral agenesis of left kidney. Existing embryologic data suggest that the blastoma of the cervical vertebrae, scapulae, and the genitourinary system have an intimate spatial relationship at the end of the 4th or beginning of the 5th week of fetal life. An alteration in this region can affect the cervical vertebrae and scapulae directly, and the genitourologic changes are mediated indirectly through the inductive capacity of the pronephric duct. The high incidence of genitourinary anomalies with Klippel-Feil syndrome has been described in the literature. Grise et al have demonstrated 52% incidence in their series of 35 cases with Klippel-Feil syndrome. More than half had unilateral defects. Most of these patients remain asymptomatic but risk of renal failure always exists.

In conclusion, there is high incidence of congenital anomalies of the genito-urinary tract in patients with KFS so they should be made aware of the potential complications. Detection of KFS should stimulate careful search for other associated systemic abnormalities.

References

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