LETTER TO EDITOR (VIEWER'S CHOICE)

HYPER IGE SYNDROME PRESENTED AS EXTENSIVE CELLULITIS AND SEVERE URTICARIA

Mukesh Vir Singh, K M Shukla

Keywords: Hyper-IgE syndrome, multiple abscess, urticaria

An 8-year-old boy presented with history of pain as well as redness of skin for 4 days. He developed high grade fever on the day of admission. Clinical examination revealed multiple abscesses, urticarial rashes, multiple old scar marks and hyperpyrexia. He had recurrent abscesses 2.5 years back for which patient was treated by local doctor with some oral medication and surgical drainage of abscesses. Laboratory evaluation revealed moderate anemia (hemoglobin 8.6 g/L). His total leukocyte count (8.6 × 103 cells/dL), platelet count (290 × 109/L), differential count (P58,L40,E02,), serum creatinine (0.77 mg/dL), blood urea (39 mg/dL), serum bilirubin (0.34 mg/dL), ASO titer (<150 i.u./L) and chest radiograph were within normal limits. Urine routine microscopy and culture did not show any abnormality. The ALT (55 i.u./L) and AST (50 i.u./L) were mildly raised. Serum IgE levels were raised (3000 i.u./mL) suggestive of Hyper IgE. Patient was treated with vancomycin and gentamicin combination and surgical drainage of abscess. Intravenous gamma-globulin was advised but patient's family could not afford. He was discharged on long term cotrimoxazole.

Hyper-IgE syndrome (HIES) was first described by Davis et al in 1966 in two girls with red hair, chronic dermatitis, and recurrent staphylococcal abscesses and pneumonias (1). They named the disease after the biblical character Job, whose body was covered with boils by Satan. In 1972, Buckley et al described two boys with similar symptoms as well as coarse facies, eosinophilia, and elevated serum IgE levels. These two syndromes are thought to be the same and are under the broad category of HIES (2). Abnormal neutrophil chemotaxis due to decreased production of interferon gamma is thought to cause the disease (3). Both autosomal dominant and recessive inheritance have been described. The disease was linked to mutations in the STAT3 gene after cytokine profiles indicated alterations in the STAT3 pathway (4).

Elevated IgE is the hallmark of HIES, usually > 10 times normal (5). However, patients younger than 6 months of age may have very low to non-detectable IgE levels. Eosinophilia is a common finding (5). IgE levels in HIES exceed 2000 IU/ml. However, IgE levels may decrease with age, may fall within the normal range (0.1-90 IU/ml) in about 20% of the cases (5,6).

HIES often appears early in life with recurrent staphylococcal and candidal infections, pneumonias and eczematoid skin. Finally, some patients have abnormal facial features and scoliosis and fragile bones (5). Most patients with HIES are treated with chronic antibiotics to help and protect them from staphylococcal infections. Good skin care is also important in patients with HIES. Intravenous gamma globulin has also been suggested for the treatment.

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LETTER TO EDITOR (VIEWER'S CHOICE)

RADIAL CLUB HAND

Ramesh R Pol, B.C.Yelamali, Mahesh Bhagawati

Key Words: Club Hand, Apical Ectodermal Ridge, Absent Thumb, Absent Radius

A 21 year old primigravida with severe anemia delivered a baby girl with abnormally short right forearm, absent right thumb with radial deviation of hand (Figure-1). Birth weight was 2.5 kg. General physical and systemic examination was normal, and no other malformation was noted clinically. Radiograph of right hand and forearm showed absent radius, absent thumb, absent 1st metacarpal bone, short ulna with radial deviation and no carpal bones (Figure-2). Other examination findings were normal. Chest x-ray, echocardiography, blood counts, ultrasound of abdmen were normal. There was no history of any drug intake during antenatal period. Child is being followed up since one and half years of age and her repeat blood counts are normal.

Figure-1: Short right forearm, absent right thumb with radial deviation of hand

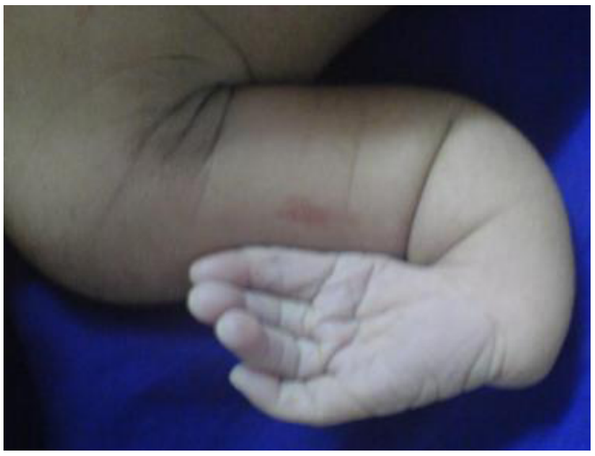
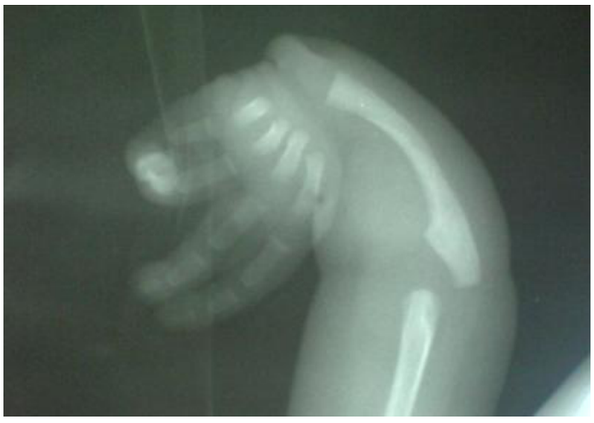


Figure-2: Radiograph of right hand and forearm showed absent radius, absent thumb, absent 1st metacarpal bone, short ulna with radial deviation and no carpal bone



Club hand deformitis are classified into two main categories radial and ulnar. Radial club hand includes a wide spectum of disorders that encompass absent thumb, thumb hypoplasia, thin first metacarpal and absent radius. Ulnar club hand is much less frequent than radial club hand and ranges from mild deviation of hand on the ulnar side of forearm to complete absence of ulna. Radial club hand is frequently syndromatic, where as ulnar club hand is usually isolated anomaly (1). Radial club hand is a deficiency along the preaxial or radial side of the extremity. Petit in 1733, described the first case of Radial club hand in an autopsy of neonate with bilateral club hands and absent radii. Although considerable forearm and hand anomalies are the classic findings, proximal deficiencies can occur throughout arm and shoulder girdle (2,3). Few cases of Radial club hand have been reported in Indian literature (4,5). Radial club hand develops early in pregnancy, with defect occurring during embryo development (between the 28th and 56th day of gestation), when the bones of the hand and forearm are being formed. Sometimes, it can be picked up on a prenatal ultrasound. Even if it is detected prenatally, the condition cannot be treated until after the baby is born. Most cases are sporadic without definable cause. The frequency of this anomaly is between 1:55000 to 1:100000 live births. Radial deficiency is bilateral in 50% of cases and is slightly more common in males than in females (3:2). The incidence of radial deficiency within the same family is 5 to 10%; it is most common in radial aplasia associated with cardiac abnormalities (3,6). Several theories have been raised, such as maternal drug exposure, compression of the uterus, vascular injury, but the Current theory relates the etiology of radial club hand to the Apical Ectodermal Ridge (AER). This structure is a thickened layer of ectoderm that directs differentiation of underlying mesenchymal tissue and limb formation. Removal of a portion of AER in chick embryos has produced anomalies similar to radial club hand. Therefore, a defect of AER is the most probable cause of radial club hand. The extent of deformity is related to the degree and extent of AER absence. Heikel, based on the amount of radius present, classifies radial club hand into four types. Type I: Short distal radius - mildest type, Type II: Hypoplastic radius, Type III: Partial absence of radius, Type IV: Total absence of radius - most common variant. Variable degrees of thumb deficiencies are frequently associated with all patterns (6). In Radial club hand, forearm is foreshortened, with marked curving of the forearm, stiffness of the elbow and fingers, the wrist is positioned in radial deviation, and the thumb will either be small or absent. Abnormalities of bone and joint, muscles and tendons, nerves and arteries are seen with this condition (7). Many anomalies can have association with Radial dysplasia. Cardiovascular anomalies include ventricular septal defect, patent ductus arteriosus, coarctation, Dextrocardia, and pulmonary stenosis. Genito-urinary anomalies include ectopic kidney, hypoplastic kidney,

urethral valve, horseshoe kidney and duplication. GIT anomalies seen are esophageal atresia, TE fistula, anal atresia, small bowel atresia and malrotation. Skeletal conditions like scoliosis, hemi vertebrae, Klein-filter syndrome, sacral agenesis and hip dislocation (8). Whenever a club hand is identified it is important to conduct a thorough examination of fetus and new born to delineate associated anomalies that may suggest a syndrome. Fetal blood sampling procedures and fetal echocardiography are recommended. A complete blood cell count including platelets is important to diagnose hematological conditions like Fanconi's anemia, TAR syndrome, Aase syndrome. Fetal karyotype is indicated because several chromosomal anomalies may be associated (Trisomy 18 and 21, deletion of long arm of chromosome 13 and ring formation of chromosome 4). Syndromes associated with absent or hypoplastic thumb with radial dysplasia include Holt-Oram, DeLange, Daune, Ives Houston, and Roberts, Rothmund Thompson and Shokeir syndromes.

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LETTER TO EDITOR (VIEWER'S CHOICE)

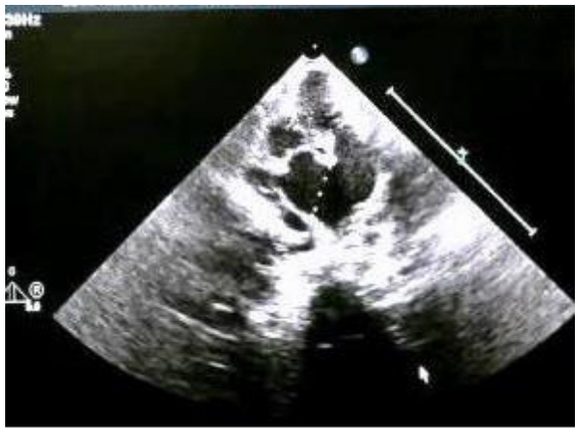
AORTO PULMONARY WINDOW IN INFANT

Amar Taksande, V Gouthami, Sumanta Padhi, Kinjal Bakshi

Keywords: Aortopulmonary window, Congenital heart disease, Infant

A 3 month old male infant, weighing 4.6kg was referred for the assessment of a heart murmur. History of breathlessness, noisy breathing since last 2 months and not feeding well since last 3 days was present. He was a full-term delivery born by emergency caesarean section for non progression of labour. On physical examination, heart rate was 130/min, respiratory rate was 48/min, blood pressure was 106/62 mmHg and oxygen saturation was 92%. There was no respiratory distress. All the peripheral pulses were well felt. On auscultation, first and second heart sounds were present with P2 louder than A2. Third heart sound was present. Ejection systolic murmur was best heard in the upper left parasternal area. On investigation, complete blood cell count showed hemoglobin of 9.3gm%, hematocrit of 30%, white cell count of 10,000/cumm, and platelet count of 3.8lakhs/cumm. Chest X-ray revealed an enlarged cardiac silhouette (CT>0.55) and increased pulmonary vasculature. Electrocardiography (EKG) showed sinus rhythm, normal QRS axis with biventricular hypertrophy. Echocardiography (fig1) showed large (13mm) distal aorto-pulmonary window involving the right pulmonary artery origin with bidirectional shunt with hyperkinetic pulmonary hypertension. There was mild tricuspid regurgitation with moderate pulmonary regurgitation (gradient=56mmHg). Left atrium and left ventricle was dilated. Subsequently, surgical closure of AP window was done. Post operative echo revealed no residual flow, normal left pulmonary artery & right pulmonary artery with good biventricular function. On follow-up after two month, child is doing well.

Figure 1: Echocardiography shows the large aorto-pulmonary window.



An Aortopulmonary window (APW) is usually wide, causing important symptoms in the first weeks or months of life, which result from the significant left-to-right shunt (1,2). In 1936, Abbot reported that the APW is a rare congenital cardiac anomaly accounting for about 0.1% of all cardiac defects in an autopsy study. This anomaly exists causing a communication between the ascending aorta (AA) and the pulmonary artery (PA) with normal separation of the aortic and pulmonary valves, which resulted from faulty embryogenesis of septation of the aortopulmonary trunk. John Elliotson first described it in 1830 and Gross performed the first successful surgery in 1948 (3,4). APW is classified into three types according to proximal, distal or total defects as per Mori's classification (5,6). Hemodynamic abnormalities of AP window mimic a large Ventricular Septal Defect (VSD) or Patent Ductus Arteriosus (PDA) with pulmonary hypertension. The heart murmur of this anomaly is often mistaken for the murmur of a high VSD, because although a continuous murmur may be present in patients with APW, more often there is only a systolic murmur that is generally heard along the upper left sternal border. Congestive heart failure (CHF) and pulmonary hypertension appear usually during early infancy. The clinical diagnosis of APW, although difficult to make, should be considered in any patient with congestive heart failure and low weight gain. EKG and chest X-ray are nonspecific, the echocardiogram is very important for diagnosing the APW (7,8). Garver et al. found that noninvasive imaging with echocardiography and/or MRI adequately defined the anatomical defects before surgery (9). Transcatheter closure of APW should be considered when anatomy is favorable in terms of location and size of the defect, in the absence of associated anomalies. In our case, the aorto pulmonary window was distal and large size, which was not possible to close by device. Hence, surgical closure of the APW was done. The Rashkind double umbrella device, the Amplatzer duct occluder, the Amplatzer septal occluder, muscular VSD occluder, and perimembranous VSD occluder have all been used to close small (type I) defects (10-12). Surgery is indicated as soon as the diagnosis is established, regardless of the patient's age when Eisemenger syndrome does not exist. The choice of the surgical technique is based upon the type and size of the AP window. The aorta and pulmonary artery may be divided, and the defects in the walls may be closed primarily or with patch material. Alternatively, the aorta or pulmonary artery may be opened and the defect patched using autologous, homologous, xenograft, or synthetic material. The prognosis of the patient with APW is excellent if surgical correction is performed early in life, before irreversible pulmonary vascular changes occur.

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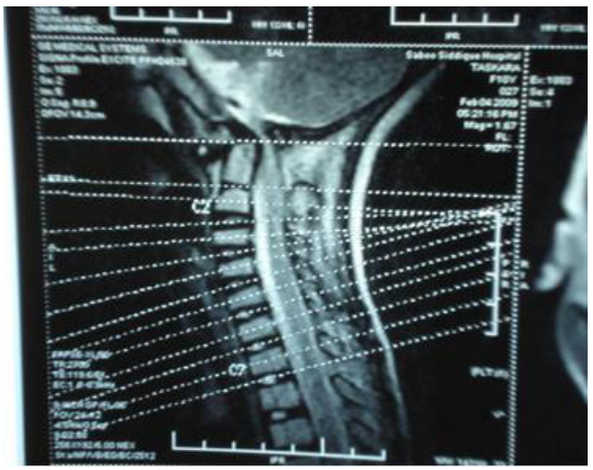
LETTER TO EDITOR (VIEWER'S CHOICE)

OS ODONTOIDEUM

Rajesh Kulkarni, Ashok D Rathod

A 10 year old female child was brought with history of weakness of left lower limb since 11 months, right lower limb since 10 ½ months and left upper limb since 8 months. Child was apparently alright one year back when she started having recurrent falls while walking and running. This was followed by weakness in left lower limb and, 15 days later, she started having weakness of right lower limb. The weakness gradually progressed over a period of 3 months such that she initially had difficulty in walking, later she could walk only with support and also could not stand up from sitting position. Since last 3 months patient is bed ridden & sits only with support. There was a history of weakness of proximal muscles in left upper limb since 8 months. Child was able to lift right hand above head but with difficulty. There was no history suggestive of cortical, cranial nerve, bladder or sensory involvement. On examination of motor system there was asymmetrical proximal muscle weakness (left more than right) in lower limbs as well as upper limbs with brisk deep tendon reflexes. Sensory system examination was normal .There was no abnormality on examination of the spine. MRI Cervical Spine and 3 D CT cervical spine revealed cranio vertebral junction anomaly with os - Odontoidium, atlanto-axial dislocation and congenital fusion of C2 and C3 vertebral bodies with cord compression by posterior superior portion of C2 with cord edema/myelomalacia at C1/C2 level and severe secondary canal stenosis at C2/C3 levels (Figure 1). Patient was given high dose steroid for spinal cord edema. Strict immobilization of spine with hard neck collar was done. The patient has been referred for traction & operative stabilization by wiring and screw fixation.

Figure 1: MRI spine showing os - Odontoidium, atlanto- axial dislocation and congenital fusion of C2 and C3 vertebral bodies with cord compression by posterior superior portion of C2.



In 1886, Giacomini coined the term os odontoideum. This entity is clinically important because the mobile or insufficient dens renders the transverse atlantal ligament (TAL) ineffective at restraining atlantoaxial motion. Translation of the atlas on the axis may lead to impingement of the upper cervical cord or vertebral artery. Os odontoideum is rare, but the exact prevalence and incidence are unknown. Many cases are either incidentally detected or are diagnosed when patients become symptomatic. The age at diagnosis varies significantly from the first to the sixth decades of life. With increased awareness, however, os odontoideum has been diagnosed in younger patients. While the etiology remains controversial an increased frequency of os odontoideum has been reported in patients with multiple epiphyseal dysplasia and Down's syndrome (1-3). Some authors speculate that os odontoideum represents a previous fracture of the odontoid synchondrosis before its closure at age 5-6 years. These authors describe os odontoideum in patients with previously normal cervical radiographs. For example, Schuler et al elegantly described the evolution of an os odontoideum following trauma in a child (4).

In patients with os odontoideum, management principles are clinical and radiologic surveillance in asymptomatic patients, high dose steroids for spinal cord edema, strict immobilization of spine by hard neck collar, traction and operative stabilization in the form of wiring and screw fixation if there is spinal instability, neurologic involvement or intractable pain.

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LETTER TO EDITOR (VIEWER'S CHOICE)

PALMOPLANTAR KERATOSIS

Kulkarni KP, Kaur S

A 4 year old male child presented with thickening of palmaer and plantaer surfaces of hands and feet respectively, since 2 years of age. He had painful fissures on the palmer surface with consequent limitation of activities of daily living. There was no history of any affected family member with similar complaints. The rest of the skin was unaffected. There was no evidence of any systemic involvement in the form of periodontitis, alopecia, neurological involvement, mental retardation, nail anomalies, polydactyly, syndactyly, clubbing of fingers or dwarfism. The rest of the systemic examination was within normal limits.

Thus, the child had isolated and sporadic palmoplantar keratosis. Palmoplantar keratoderma (PPK) constitutes a heterogeneous group of inherited as well as acquired disorders characterized by thickening of the palms and the soles of affected individuals (1,2). In diffuse PPK there is uniform involvement of the palmoplantar surface. This pattern is usually evident within the first few months of life. Keratin 1 and keratin 9 mutations have been reported in these patients (3). The diagnosis is largely clinical, especially in a resource limited setting. The solitary nature of presentation in the index patient precludes assessment of inheritance pattern and may have occurred due to sporadic mutations in the keratin genes.

Treatment includes salicylic acid, 50% propylene glycol in water under plastic occlusion several nights per week, and lactic acid- and urea-containing creams and lotions; all have been shown to be helpful. Patients with isolated PPK usually have a benign clinical course.

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LETTER TO EDITOR (VIEWER'S CHOICE)

LARYNGEAL CLEFT

Ashutosh Sudhakar Mangalgiri, Raza Razvi, Rajendra Saoji, Satish B Deopujari***

Keywords: Laryngeal cleft, Cleft, Congenital Anomaly, Larynx, Tracheo-esophageal cleft.

Larynx of an 8 month male foetal cadaver was removed by dissecting above the hyoid bone and below up to the 4th to 5th tracheal ring. An incidental finding of cleft was noted in the posterior part of larynx. The cleft was extending into the interarytenoid musculature. Cricoid cartilage was palpated and confirmed that the cleft was not extending up in to the cricoid cartilage. No other recognizable congenital malformations were observed in a foetal cadaver.

Laryngotracheoesophageal cleft is a rare entity. This congenital condition depending upon the extent of the cleft in the larynx and trachea may become life-threatening and lead to immediate death after the birth (1). The incidence of Laryngotracheoesophageal cleft (LTOC) is 0.3% out of the total congenital anomalies of the larynx (2). Overall 6% of cases of type I laryngeal clefts observed in total paediatric direct laryngoscopies (3). Minor conditions usually present with hoarseness and recurrent respiratory infections and therefore many a time diagnosis is delayed. Symptoms usually resembles with oesophageal atresia & tracheo-oesophageal fistula (1). One should suspect the condition when child presents with a triad of husky cry, feeding difficulty and aspiration pneumonia (1). The symptoms mostly aggravate during feeding.

The arrest of the cranial advancement of the tracheo-oesophageal septum is responsible for the non fusion of the cricoid lamina in the midline, leading to the development of the cleft (4,5,6). The clefts are functionally divided into 4 types. Type I involving only interarytenoid musculature, Type II involves cricoid only, Type III involves proximal laryngo-tracheo-esophagus & type IV involves thoracic tracheo-esophageal septum (7). LTOC may present singly or may present with the other anomalies like, harelip, cleft palate, oesophageal atresia, atresia ani, vulvo-vestibular fistula, sacral hypoplasia (5). Familial conditions of Laryngotracheoesophageal cleft (LTOC) have also been reported (8,9).

The diagnosis of laryngeal cleft can be confirmed by direct laryngoscopy as early as 3 weeks only (1). Barium esophagogram and bronchoscopy are also important investigations for diagnosis. Treatment consists of surgical repair, although some patients

with type I laryngeal clefts may be managed on a conservative trial (3). Endoscopic repair of cleft larynx is best used for type I clefts (10). Early surgical intervention would prevent further damage to lungs and oesophagus.

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