lowed by prepubertal children (31%). This observation is similar to that observed in other places in India(2,4,5). However lymphoma occurred more (75%) in children above 5 years.

Although there was an overall male preponderance (1.9 : 1), the nervous system tumors showed definite female preponderance (1 : 1.8) which has not been reported earlier(2-5). Wilms' tumor involving the left kidney has also been reported by other workers(2,4). Hodgkin's disease formed 2/3rds of malignant lymphoma cases. All other studies have reported a higher incidence of non-Hodgkin's lymphoma(2,4).

The possibility of carcinoma in young age is usually not thought of and these cases are liable to be missed clinically. We noted it in 5.4% cases, similar to what has been observed from Bombay(1).

To conclude the prevalence of solid pediatric tumors in Kashmir is not significantly different from the rest of the country. It is tempting to postulate that environmental carcinogens do not have sufficient time to act and have thus no role in pediatric tumors. The role of genetic factors thus appears to be more important.

REFERENCES

1. Talvalkar GV. Tumors in Children. Published by International Academy of Pathology 1982, pp 1-5.
2. Venugopal KV, Joseph JP, Varma KK. Malignant tumors of infancy and childhood: A clinicopathological study. Indian Pediatr 1981, 18: 365-368.
3. Mallya M, Madhaban KK, Rama Rao BR. Malignant abdominal tumors in children. Indian Pediatr 1975, 12: 499-502.
4. Panda BK, Dandapat MC, Parida N. Pattern of pediatric solid malignant tumors in Southern Orissa. J Indian Med Assoc 1989, 87: 136-137.
5. Banerjee CK, Walia BNS, Pathak IC. Pattern of neoplasms in children. Indian Pediatr 1986, 23: 93-97.

Congenital Sensory Neuropathy with Anhidrosis

L.C. Thakur
V. Chandran
K.S. Anand

Congenital sensory neuropathy with anhidrosis (CSNA) is a rare hereditary disease with autosomal recessive mode of inheritance. The condition has also been referred to as congenital insensitivity to pain with anhidrosis in an earlier report(1). Subsequently, other reports have also appeared in literature(2,3). Because of its extreme rarity only nine cases have been described in world literature(4). We report a case diagnosed clinically as congenital sensory neuropathy with anhidrosis in a 12-year-old boy from Pondicherry.

Case Report

A 12-year-old boy, was brought in May 1989 with the complaints that he repeatedly injures himself, is indifferent to pain following injuries and has a low intelligence. The patient was the product of a consanguinous marriage and was born prematurely after 8½ months of gestation. The patient is the eldest among three siblings. His younger brother is normal, while the

From the Departments of Neurology, Jawaharlal Institute of Postgraduate Medical Education and Research, Pondicherry 605 006.

Received for publication: March 20, 1991;
Accepted: February 6, 1992

youngest–a female child–has congenital heart disease. The patient's developmental milestones were delayed grossly.

The patient was first noticed to be abnormal at 6 months of age when he had a cut with a broken bottle and did not cry. Since then, the child had several self-mutilating behaviors leading to serious injuries without any pain such as crushed fingers in the door, banged head against the wall leading to skull fracture, hitting jaw repeatedly with a hard object and pulled out the loosened teeth, jumped from heights leading to injuries, with a dislocated right hip joint in one such attempt. Since then he has been unable to walk. He has also been noticed not to sweat like others do. His body becomes very hot if the temperature of the external environment is high with normal lacrimation. On examination, the patient showed subnormal intelligence with IQ of 40-50 on clinical assessment. Pain sensation to pin prick was absent all over the body and temperature sensations were impaired. Sensation of touch was intact. Deep tendon reflexes were diminished and abdominal and plantar reflexes could not be elicited. The family was counselled regarding nature and prognosis of the illness and the importance of measures to prevent further injuries.

Discussion

A diagnosis of CSNA, as reported in literature, is based on the absence of pain and impairment of temperature sensations all over the body, onset of illness at birth, decreased tendon reflexes, anhidrosis and mental retardation(3-5). All these features were seen in this patient reported. The self-mutilating behavior exhibited by the patient has also been described in 8 out of 9 cases reported in literature, while a history of consanguinous marriage of the parents was found in a third of them(4).

Autopsy findings in one of the cases had shown an absence of Lissaver's tract and dorsal root axons and a reduction in size of spinal tract of the trigeminal nerve with paucity of small fibres in it(6). Myelinated fibres in peripheral nerves and dermal networks are intact(3). A genetically determined defect in differentiation and migration of neural crest elements early in embryogenesis has been postulated(6). Despite the anhidrosis, dermal sweat glands are normal(2).

The differential diagnosis of CSNA rests primarily is separating it from other mutilating neuropathies. Congenital sensory neuropathy may be differentiated by the absence of all modalities of sensation including touch, normal sweating, and an absence of myelinated nerve fibres and dermal nerve endings on biopsy(7). Hereditary sensory radicular neuropathy has an onset in later childhood, with absence of all modalities of sensations, greater involvement in distal parts of extremities and a loss of myelinated nerve figures on biopsy(8). In congenital insensitivity to pain, there is no abnormality of temperature sensation, tendon reflexes or sweating(9). Familial dysautonamia shows increasing sweating and impaired lacrimation, while the loss of pain sensation is not total(5).

The short term management of CSNA involves treatment of the injuries, correction of fractures sustained and correction of the resultant deformities. Long term management involves counselling the family members for prevention of self mutilating behavior which in the presence of mental retardation and in the absence of protective reflexes, makes the management of such children very difficult, even in the most advanced countries with modern

facilities. The skin in these patients shows normal sweat glands but their intervention by the post-ganglionic sympathetic fibres is defective. When external temperature is raised, as happens in hot summer months in Pondicherry, patients have inability to sweat, leading to alteration in thermoregulation and spells of high grade fever requiring lowering of external temperature by physical means like cold/ice sponging or air conditioning. This method proved successful in preventing heat exhaustion in the long term management of our patient.

REFERENCES

1. Swanson AG. Congenital insensitivity to pain with anhidrosis: a unique syndrome in two male siblings. Arch Neurol 1963, 8: 299-306.
2. Pinsky L, Digeorge AM. Clinical familial sensory neuropathy with anhidrosis. J Pediatr 1966, 68: 1-13.
3. Vassella F, Emrich HN, Kraus-ruppeat R, Aufdermaur F, Tonzo. Congenital sensory neuropathy with anhidrosis. Arch Dis Child 1968, 43: 124-130.
4. Mazar A, Herold HZ, Vardy PA. Congenital sensory neuropathy with anhidrosis: Orthopedic complications and management. Clin Orthop 1976, 118: 184-187.
5. Gillespie JB, Peucca LG. Congenital generalized indifference to pain. Am J Dis Child 1960, 100: 124-128.
6. Swanson AG, Buchan GC, Alvrod EC. Anatomic changes in congenital insensitivity to pain. Arch Neurol 1965, 12: 12-18.
7. Murray TJ. Congenital sensory neuropathy. Brain 973, 96: 294-387.
8. Denny-Brown D. Hereditary sensory radicular neuropathy. J Neurol Neurosurg Psychiatry 1951, 14: 237-252.
9. Thrush DC. Congenital insensitivity to pain: A clinical, genetic and neurphysiological study of four children from the same family. Brain 1973, 96: 369-386.

Priapism in Children with Sickle-Cell Disease

E.O. Enwerem
E.M.L. Endeley
C. Holcombe
R.V. Patel

Priapism is a painful and persistent penile erection. Majority of cases are primary (idiopathic), present in middle age and require surgical treatment(1). We wish to report two pediatric cases of secondary priapism due to hematological disorder of sickle-cell disease leading to slugging of blood within the corpora cavernosa which were treated successfully with conservative measures.

Case Reports

Case 1: A 9-year-old male child was admitted with a history of painful and persistent erection of penis associated with yellow sclera and high grade of fever with rigors of 2 days duration. He was a known case of sickle-cell disease (HbSS) diagnosed at the age of 2 years. The patient was in severe pain, febrile, mildly icteric and had pallor. Hepatosplenomegaly with tender erect penis was observed. Peripheral smear showed *P. falciparum*, features of sickle-cell anemia and polymorphonuclear

From the Department of Surgery, College of Medical Sciences and University of Maiduguri Teaching Hospital, Maiduguri, Nigeria, West Africa.

Reprint requests: Dr. R.V. Patel, 210, A.G. Office Staff Quarters, Near Saurashtra University, Rajkot 360 005.

Received for publication: September 25, 1991; Accepted: February 6, 1992