Linear nevus sebaceous syndrome

Leema Pauline, Balakrishnan Kannan, Viveka Saravannan, Venkatesan Prasanna Eswaradass[1]

Departments of Neurology, Institute of Child Health, Egmore, Chennai, [1]Neurology, PSG Institute of Medical Sciences and Research, Coimbatore, Tamil Nadu, India

For correspondence:
Dr. Venkatesan Prasanna Eswaradass, 3, Vijayarahavachari Road, Gandhi Road, Salem - 636 007, Tamil Nadu, India.
E-mail: eprasanna2k1@gmail.com

Ann Indian Acad Neurol 2014;17:468-9

Case Report

A 8-year-old male child born of consanguineous marriage presented with history of recurrent right focal seizures becoming generalized since childhood. He also had delayed milestones and mental retardation. He was admitted several times for his seizures since infancy. On examination, he had characteristic nevus over his forehead extending into the scalp along midline oriented vertically [Figure 1]. He also had right hemiplegia. We made a diagnosis of Linear nevus sebaceous syndrome (LNSS) and magnetic resonance imaging (MRI) showed left hemimegalencephaly (HME) [Figure 2].

Discussion

LNSS, also called epidermal nevus syndrome, is a rare, sporadic neurocutaneous syndrome characterized by a linear sebaceous nevus of Jadassohn, mental retardation, and, seizures. The syndrome was originally described by Schimmelpenning and further defined by Feuerstein and Mims.[1,2] The skin lesions are characteristically found in scalp, face, or neck. A paramidline location of the nevus is typical with the lesion often vertically oriented from the middle of the forehead along the nose to the upper lip just lateral to midline. Hemangiomas, lipomas, hypopigmented lesions, and rarely *cafe´ au lait* spot have also been described.

Among central nervous system (CNS) abnormalities, unilateral HME is the most common finding and is typically ipsilateral to the skin lesions. In addition, other neuronal migration abnormalities (pacygyria, agyria, heterotopia, etc.), vascular malformations, agenesis of the corpus callosum, Dandy-Walker syndrome, myelomeningocele, Arnold-Chiari malformation, and tumors have been reported.[3-5] HME is characterized by unilateral enlargement of a cerebral hemisphere and a normal contralateral hemisphere, cerebellum, and brainstem. It occurs in about 50% of LNSS. The most common MRI findings in HME are ventricular enlargement, increased signal intensity in white matter of the affected hemisphere on T2-weighted images, loss of delineation between white and gray matter and agyria.[6,7] HME is clinically characterized by seizures, mental retardation, and contralateral hemiparesis. Seizures are focal and may present as infantile spasms initially. Usually they are refractory and require hemispherectomy. Pathologically, HME is characterized by complete disorganization of the cortical cytoarchitecture without normal cortical layers and the area of the cortex is smaller than the contralateral side, suggesting abnormal proliferation of the white matter as the mechanism of hemispheric enlargement.[8]

Hemihypertrophy of the face occurs in about 50% of patients ipsilateral to the nevus and hemiatrophy contralateral to the nevus is occasionally seen. Eye involvement occurs in about 33% of patients and includes lipodermoid scleral tumors, microphthalmia, corneal opacities, focal globe calcifications, and colobomas. Cardiac anomalies include coarctation aorta and ventricular septal defect. With increasing age, there is an

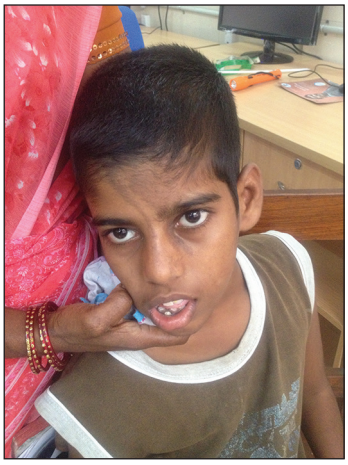

Figure 1: Vertically oriented forehead naevus along midline

Access this article online

Quick Response Code:

Website: www.annalsofian.org

DOI: 10.4103/0972-2327.144042

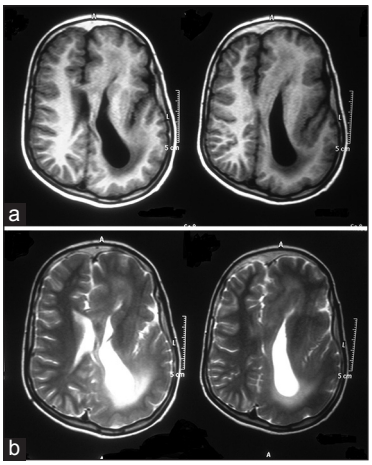

Figure 2: (a) T 1 weighted image shows left hemimegalencephaly with agyria-pachygyria complex (b) T 2 weighted image shows left ventricle colpocephaly

increased incidence of skin, breast, salivary gland, stomach, esophageal, and bladder cancer.

Conclusion

LNSS is a rare neurocutaneous syndrome. Most common CNS abnormality associated with it is HME. It typically presents with seizures, mental retardation, hemiparesis, and linear nevus.

References

1. Lansky LL, Funderburk SF, Cuppage FE, Schimke N, Diehl AM. Linear sebaceous nevus syndrome. Am J Dis Child 1972;123:587-90.
2. Sarwar M, Schafer ME. Brain malformations in linear nevus sebaceous syndrome: An MR study. J Comput Assist Tomogr 1988;12:338-40.
3. Hennekam RC, Kwa VI, van Amerongen A. Arteriovenous and lymphatic malformations, linear verrucous epidermal nevus and mild overgrowth: Another hamartoneoplastic syndrome? Clin Dysmorphol 1999;8:111-5.
4. Chatkupt S, Ruzicka PO, Lastra CR. Myelomeningocele, spinal arteriovenous malformations and epidermal nevi syndrome: A possible rare association? Dev Med Child Neurol 1993;35:737-41.
5. Dodge NN, Dobyns WB. Agenesis of the corpus callosum and Dandy-Walker malformation associated with hemimegalencephaly in the sebaceous nevus syndrome. Am J Med Genet 1995;56:147-50.
6. Carter BS, Hurst DL. Magnetic resonance imaging of the brain in the linear sebaceous nevus syndrome. J Child Neurol 1990;5:68-9.
7. Cavenagh EC, Hart BL, Rose D. Association of linear sebaceous nevus syndrome and unilateral megalencephaly. AJNR Am J Neuroradiol 1993;14:405-8.
8. Kato M, Mizuguchi M, Sakuta R, Takashima S. Hypertrophy of the cerebral white matter in hemimegalencephaly. Pediatr Neurol 1996;14:335-8.

How to cite this article: Pauline L, Kannan B, Saravannan V, Eswaradass VP. Linear nevus sebaceous syndrome. Ann Indian Acad Neurol 2014;17:468-9.
Received: 15-01-14, **Revised:** 27-05-14, **Accepted:** 08-05-14
Source of Support: Nil, **Conflict of Interest:** None declared.