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Tc-99m HMPAO brain SPECT in linear nevus sebaceous syndrome

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The authors present a case of linear nevus sebaceous syndrome (LNSS) with atrophy of bilateral frontotemporal areas of the brain, left cerebellum and hippocampus, and bilateral uncal dysplasia demonstrated by magnetic resonance imaging (MRI). Magnetic resonance angiography revealed bilateral internal carotid artery hypoplasia with absence of flow in the anterior and middle cerebral arteries. Tc-99m HMPAO brain SPECT demonstrated more areas of perfusion defects while MRI detected volume loss and gliosis in affected areas. Tc-99m HMPAO brain SPECT may be more useful for revealing absent or decreased perfusion areas of brain lesions than MRI in LNSS.

Key words: linear nevus sebaceous syndrome, Tc-99m HMPAO brain SPECT, MRI, MR angiography

INTRODUCTION

Linear nevus sebaceous syndrome (LNSS) or nevus sebaceous of Jadassohn was first defined by Feuerstein and Mims.[1] This syndrome is characterized by nevus sebaceous, mental retardation, seizures and ocular abnormalities. There appears to be a broad spectrum of manifestations of LNSS. Dilated lateral ventricles, atrophy of some areas of the brain and hemimegalencephaly have been reported on conventional brain scans.[2–8] However, there are few reports regarding the scintigraphic pattern with Tc-99m HMPAO in patients with LNSS.[7,8]

In this report, a case of LNSS studied by Tc-99m HMPAO brain SPECT was presented. We discussed the relation between our results and the abnormalities detected by magnetic resonance imaging of the brain.

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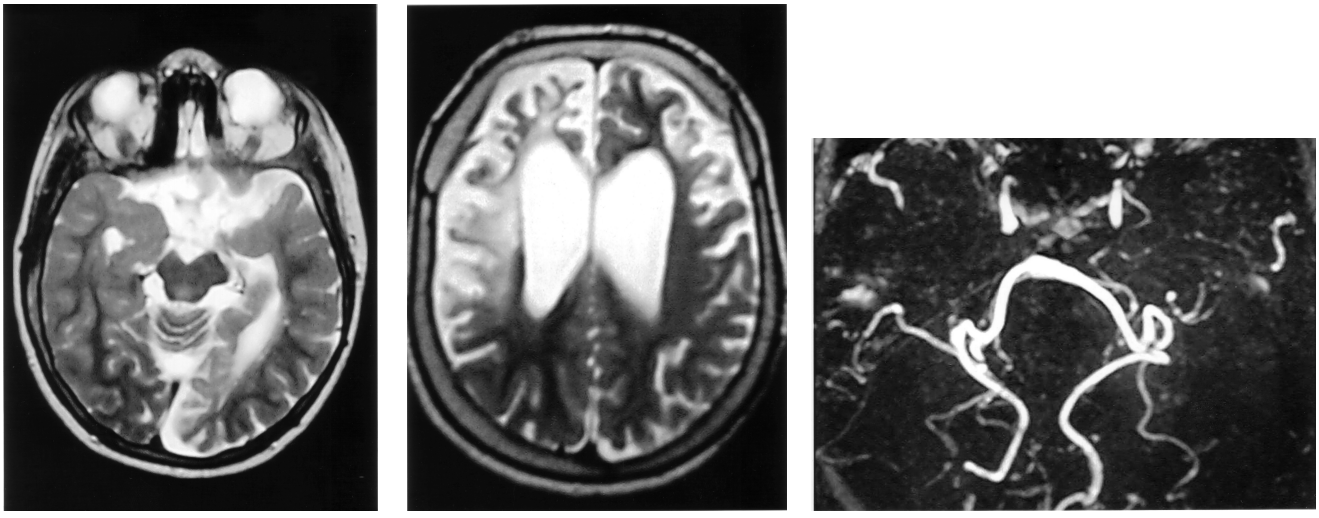
CASE REPORT

The patient was a male product of a full-term pregnancy. Delivery was vaginal and uncomplicated. There was no family history of consanguinity or birth defects. Shortly after birth; it was noted that the baby had bilateral epibulbar masses with a linear nevus on his scalp. At 9 months of age, he was referred to our pediatric ophthalmology service for evaluation and treatment of his bilateral epibulbar tumors. Histopathologic examination of the excised tumors revealed complex choristomas. Skin findings included linear nevus and patchy alopecia. The scalp lesion was also excised and demonstrated keratinizing stratified squamous epithelium covering a connective tissue that contained sebaceous glands. At the age of 17 months, the child presented with seizures, hypotonia, developmental delay and hemiparesis of the right side.

The patient underwent magnetic resonance imaging (MRI), magnetic resonance angiography (MRA) and single photon emission computed tomography (SPECT) with Tc-99m HMPAO. Cranial MRI and MRA were performed with a 1.5 T clinical MRI scanner (Philips Gyroscan, Best, the Netherlands) under sedation. Axial and sagittal T1-weighted (W) spin-echo (SE) (TR: 550 msec; TE: 14 msec) and axial T2-W turbo SE (TR: 2000



A B C

Fig. 1 Axial T2-weighted MR image of the patient. A: Bilateral uncal dysplasia with Wallerian degeneration of right cerebral peduncle and left hippocampal atrophy. Flow voids of middle and anterior cerebral arteries were absent. B: Bilateral frontal gliosis with marked cortical atrophy and bilaterally dilated lateral ventricles.

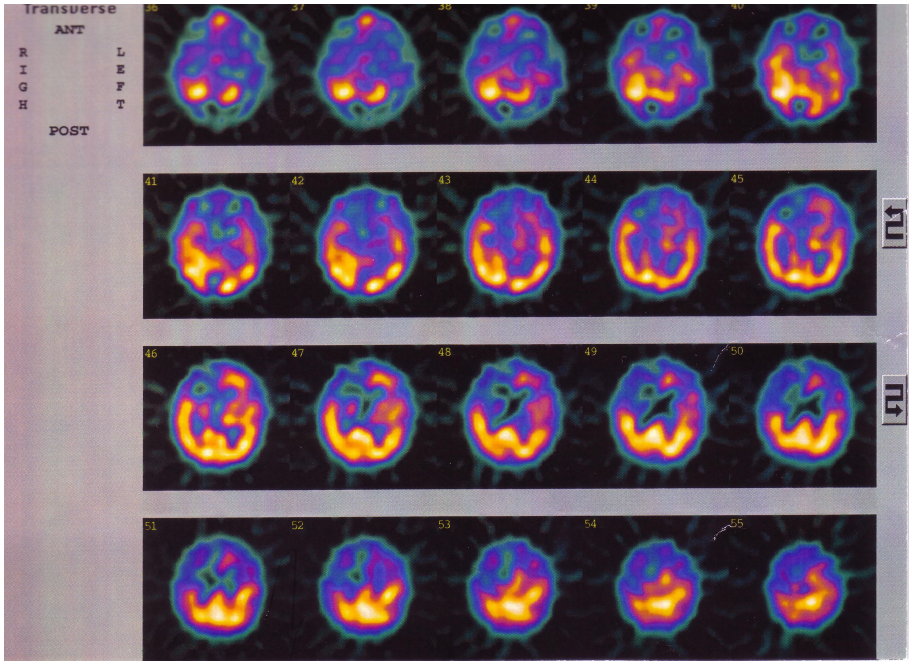


Fig. 2 Tc-99m HMPAO brain SPECT showing perfusion defects and areas of marked hypoperfusion.

msec; TE: 100 msec) and coronal FLAIR images (TR: 4700 msec; TE: 100 msec; IT: 2000) were obtained. MR angiography images were obtained with 3D time of flight (TOF) technique.

Tc-99m HMPAO brain study was studied in the supine position with eyes closed in a silent and darkened room at 10.00 a.m. Tc-99m HMPAO (Ceretec, Amersham, International plc, UK) was prepared according to the manufacturer's instructions and used within 20 minutes after labeling. The SPECT was performed by using a 360° rotating dual head gamma camera system (Siemens, E-cam), equipped with high resolution collimators, after 20 minutes following the injection of 350 MBq of Tc-99m HMPAO. The patient's head was held in a plastic head holder during scanning to prevent movements. Data were obtained in 64 × 64 pixel matrices through 360° rotation at 3° intervals, for 30 s per arc interval. No zoom was used and the corresponding pixel size was 6.4 mm. Reconstruc-

tion was performed by filtered back-projection by using a Butterworth filter (cut-off frequency 0.5, power factor 8). Slice thickness was one pixel, and no attenuation or scatter correction was done.

MRI of the brain revealed bilateral frontal gliosis with marked cortical atrophy, atrophy of right thalamus, basal ganglion, corpus callosum, left hippocampus and cerebellum. There were cortical dysplasia including unci of bilateral temporal lobes, Wallerian degeneration of right brainstem, arachnoidal cyst in the left posterior fossae, bilaterally dilated lateral ventricles and absence of flow voids in the anterior and middle cerebral arteries (Fig. 1A, 1B). MRA with 3D TOF technique showed bilateral internal carotid artery hypoplasia with lack of flow in the anterior and middle cerebral arteries. The vertebrobasillary system arteries were normal (Fig. 1C).

Tc-99m HMPAO SPECT brain imaging demonstrated left cerebellar atrophy, perfusion defects in the right occipital lobe, bilateral frontotemporal regions of the brain, basal ganglion, right thalamus and corpus callosum. Marked hypoperfusion was seen especially in the anterior parts of the right hemisphere (frontal, parietal and temporoparietal regions) (Fig. 2).

DISCUSSION

Linear nevus sebaceous syndrome is a rare systemic hamartomatosis involving structures of ectodermal and mesodermal origin.[1,3,4] The etiology of LNSS is unknown and there is an absence of genetic abnormalities, sexual predilection or identifiable Mendelian inheritance pattern. External factors such as viral infections, radiation, or drug ingestion during pregnancy have not been associated with LNSS either.[5,6,9] LNSS may be caused by anomalous development of the neuroectoderm before the fourth week of gestation, which results in brain, skull and ocular abnormalities.[10]

Patients with LNSS have linear nevus sebaceous lesions of the face, seizure disorder, and mental retardation, in addition to numerous ocular, cardiac, central nervous system (CNS), urogenital and skeletal abnormalities.[3,6] Neurologic involvement is a common finding in LNSS and a number of neurologic abnormalities have been reported related to the abnormal central nervous system development. Grebe et al.[11] reviewed 74 cases and found the most common CNS abnormalities to be seizures (50%) and mental retardation (49%). Other commonly reported disorders include hemiparesis, quadriparesis, hypotonia, gait disorders, microcephaly, reflex abnormalities and diencephalic syndrome.[3,11–13] Our case had some neurological abnormalities including seizures, hemiparesis, hypotonia, deafness, mental retardation and developmental delay.

Previously reported MRI results include cortical atrophy, dilated ventricles, hemimegalencephaly, hypoplasia of optic radiation, unilateral lissencephaly, white matter hypoplasia, excessive and heterotopic gray matter, deficient sulcation and focal areas of pachygria.[8,12,14,15] As different findings, cranial MRI of our case revealed atrophy of right thalamus, basal ganglion, left hippocampus and corpus callosum. MRA showed very narrow internal carotid arteries with absence of flow in the anterior and middle cerebral arteries. Some vascular abnormalities in LNSS such as distortion of the main cerebral arteries,[8] dolichomegalic artery,[16] and dilatation and tortuosity of the middle cerebral artery[17] have been reported but, to our knowledge, bilateral internal carotid artery hypoplasia with lack of flow is a newly reported finding.

Tc-99m HMPAO is a very useful method for SPECT imaging of regional cerebral blood flow.[18] Studies of localization patterns of HMPAO have correlated well with other studies of regional cerebral blood flow.[19] Although findings of MRI in patients with LNSS have been frequently reported, there are only a few reports studied with Tc-99m HMPAO SPECT. Tc-99m HMPAO SPECT may be more beneficial than MRI to show the extent of brain defects. In our case, atrophic areas of the brain observed by MRI were compatible with the findings of Tc-99m HMPAO brain SPECT. However, more perfusion defects were revealed by Tc-99m HMPAO brain SPECT than by MRI. Therefore, we suggest that Tc-99m HMPAO brain SPECT should be used to show all areas of abnormal perfusion in patients with LNSS. Demonstrating all abnormal areas by SPECT may play a role in the treatment of this rare syndrome.

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