NEUROCUTANEOUS MELANOSIS: MRI FINDINGS IN THE BRAIN WITH REVIEW OF ARTICLES

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ABSTRACT

MRI findings in a 5 years-old boy with neurocutaneous melanosis was described. Melanocytic nodules were shown in the brain parenchyma, meninges and perivascular space. They were seen as hypersignal lesions on T1W1, iso to hyposignal lesions on T2W1. There was no significant enhancement of the nodules. Literatures were reviewed concerning this condition.

Key words Melanosis, brain, MRI

Neurocutaneous melanosis (NCM) is a rare neuroectodermal dysplasia that is characterized by multiple congenital pigmented or giant hairy cutaneous nevi in conjunction with leptomeningeal proliferation of melanin-producing cells (1). Forty percents of the patients develop primary malignant melanoma of the CNS (2). Its synonyms are Rokitansky van Bogaert syndrome and neurocutaneous syndrome (3). NCM was first described by Rokitansky (4) in 1861 and almost a century later in 1948 by Van Bogaert (5).

We report an outpatient case of this syndrome, investigated by MRI study.

CASE REPORT

A 5-months-old Thai boy, was sent from other hospital to the Ramathibodi hospital for an MRI study of the brain. He had generalized congenital giant hairy nevi at the skin. He developed convulsion without neurological deficit.

MRI study showed scattered nodules, size 2 mm to 2 cm, found at left cerebellar hemisphere, pons, both medial temporal lobes, roof of the third ventricle, grey matter of high parietal lobes on both sides and at midline frontal base. In addition, there were small plaque like lesions at the meninges of both supratentorial and infratentorial levels. These nodules and plaque like lesions were hyperintense on T1W1 sequence (TR 640, TE 11/Fr, G.E. Signa 1.5T)(Fig. 1). The signal of the lesions dropped to iso to hyposignal on T2W1 sequence (TR 2800, TE 85/Ef)(Fig.2). Gd-DTPA contrast enhancement on T1W1 (TR 640,TE 11/Fr) showed no significant enhancement of the lesions.(Fig.3) There were no other abnormality in the brain.

DISCUSSION

The mature melanocyte is one of a number of cell line that are derived from a common neural crest stem-cell pool (6). Melanocytes are normally found within the basal layer of the epidermis and also in the pia mater investing the brain and spinal cord.

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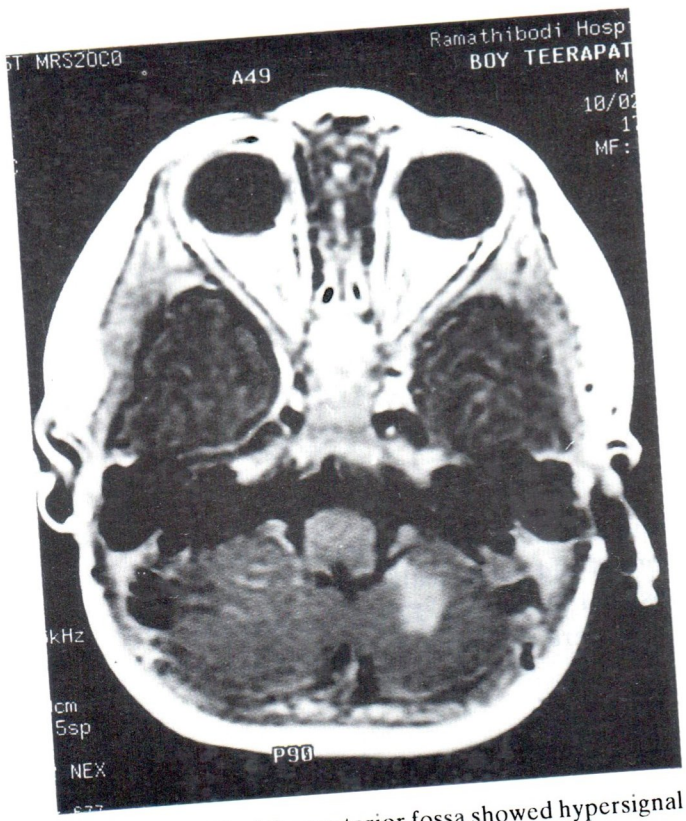


Fig. 1A. T_1WI of the posterior fossa showed hypersignal nodule at left cerebellum.

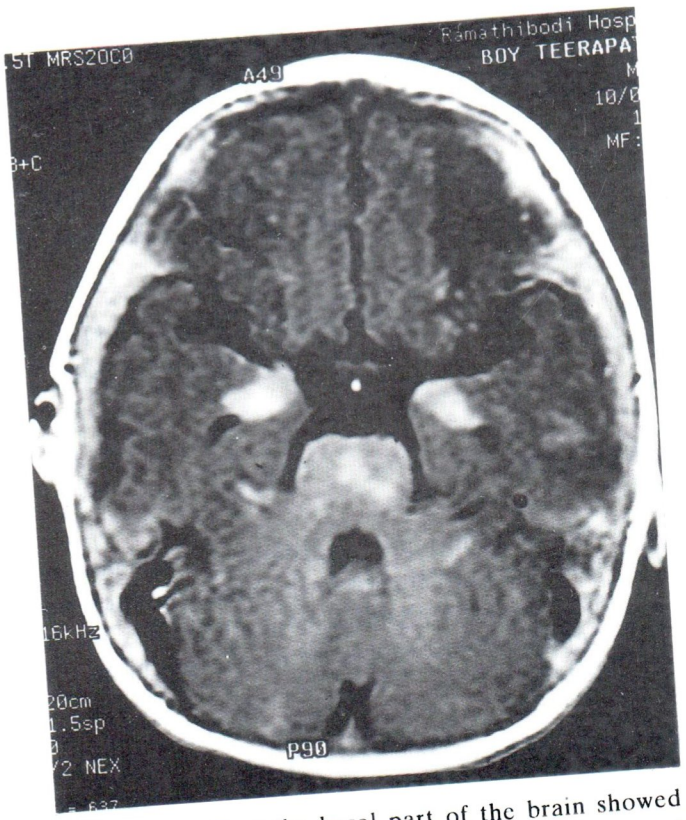


Fig. 1B. T_1WI of the basal part of the brain showed hypersignal lesions at both medial temporal lobes, pons, left cerebellum, vermian and peripheral aspect of left temporal lobe.

Neurocutaneous melanosis is thought to represent an embryonal neuroectodermal dysplasia in which excessive accumulation of melanin-producing cells occurs in a focal or diffuse distribution, within the skin and the leptomeninges (7). The pia and arachnoid membrane exhibit varying degrees of infiltration by melanotic cells. Melanotic cell infiltration may extend along the Virchow-Robin space, (8). These cells have been found also within the ventricular ependyma (9) and within the choroid plexus (10). It is frequently accompanied by a second cell type, called "melanophore". These represent melanin-laden macrophages, and it is these cells that are responsible for the parenchymal pigmentation which has been described in the basal ganglia (11), dentate nucleus (12), cerebellar hemisphere (13), pons (14), thalamus (15) and amygdaloid body (16).

The frequency of the CNS melanoma development remains unclear but was reported to be infrequent (17). Other clinical features that have been described in NCM include stillbirth (15), epilepsy (18), psychiatric disturbance (19), meningeal hemorrhage (19), cranial nerves palsies (20), subdural hemorrhage (19), and intracranial hemorrhage (22) and hydrocephalus due to obstruction of CSF circulation either at the fourth ventricle outlets or within the basal subarachnoid cisterns (7).

Involvement of the spinal cord and its coverings is variable. The cord and leptomeninges may be normal (17) or there may be varying degrees of leptomeningeal infiltration and thickening to the extent that the subarachnoid space is obliterated and the spinal cord distorted (23). Melanotic cells may infiltrate the Virchow-Robin space of the cord (11). A single malignant melanoma has been reported arising from the cord meninges (24). Associated syringomyelia was reported by Leaney (7).

Scattered abnormal areas of calcification and irregular contrast enhancement in the suprasellar cistern, in the right Sylvian fissure and the left occipital lobe were described by Leaney (7).

The lesions in our case were present both

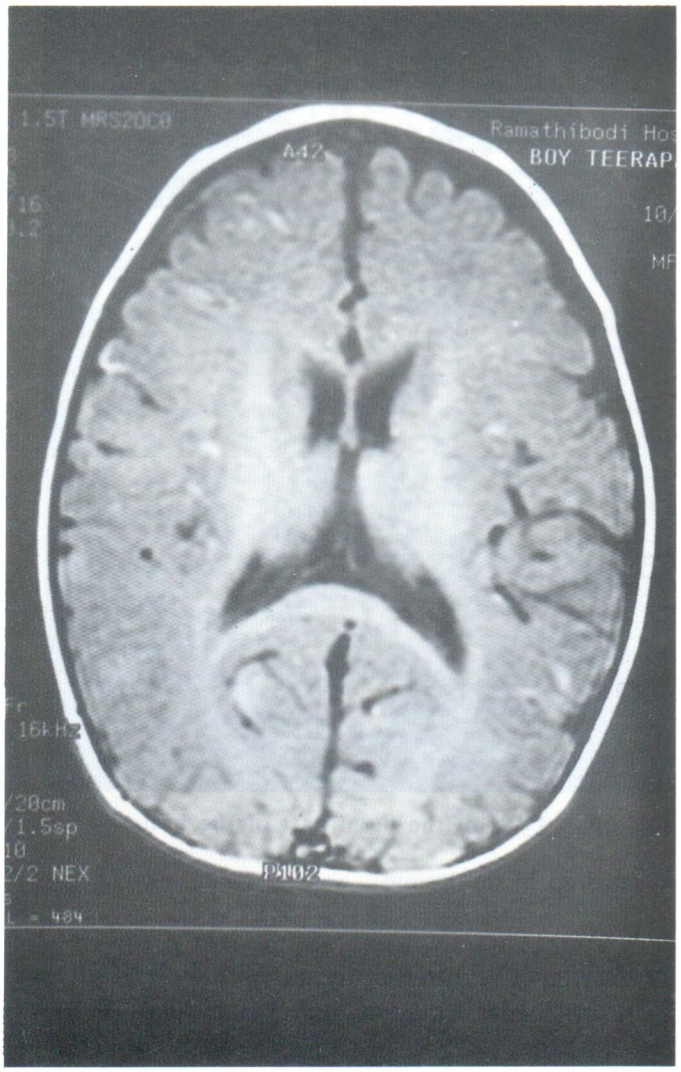


Fig. 1C. T_1WI of the brain at mid ventricular level showed hyper signal lesions scattering at convexity meninges and perivascular space.

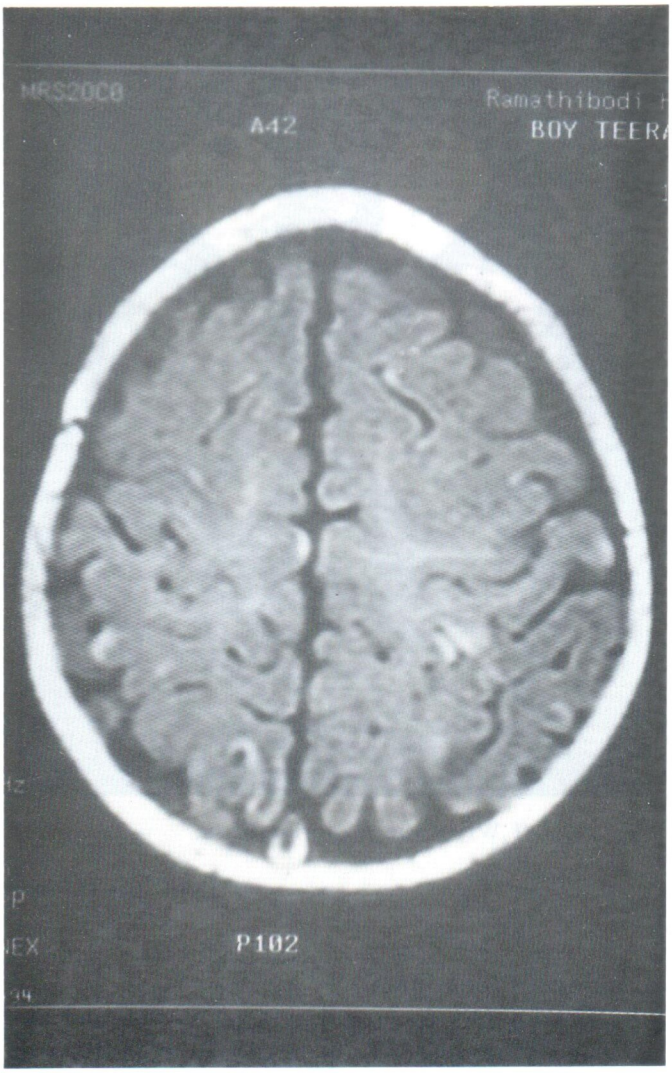


Fig. 1D. T_1WI of the brain near vertex showed bright signal lesions depositing at meninges.

in the brain parenchyma and leptomeninges, including perivascular space. The brain lesions in our case have both T1 and T2 shortening; it appears to be due to enhanced proton relaxation caused by a dipole interaction, secondary chelated metal ions within the melanin itself. Contrast enhancement showed no significant enhancement, either in the nodules or in the leptomeninges. A case report from Rhodes (1) showed strong leptomeningeal enhancement, howevere, no parenchymal nodule was present.

REFERENCES:

1. Rhodes RE, Friedman HS, Halter HP Jr et al: Contrast enhanced MR imaging of neurocutaneous melanosis. AJNR 1991;12:380-382.
2. Pont MS, Elster AD. Lesions of skin and brain. Modern imaging of the neurocutaneous syndromes. AJR 1992; 158:1193-1203.
3. Taybi H, Lachman RS: Neurocutaneous melanosis sequence;pp 329-310 in Radiology of Syndromes, Metabolic Disorders, and Skeletal Dysplasia; 3rd Ed, 1990, Chicago, Year book medical publishers, Inc
4. Rokitansky J: Ein ausgezeichneter Fall von Pigment-Mal mit ausgebreiteter Pigmentierung der inneren Hirnhaute und Ruecken-markshaeute. Allg Wien Med Ztg 1861;6:113-116.
5. Van Bogaert LC: La melanose neurocutanee diffuse heredofamiliale. Bull Acad Roy Med Belg 13:1948;13: 397-427.

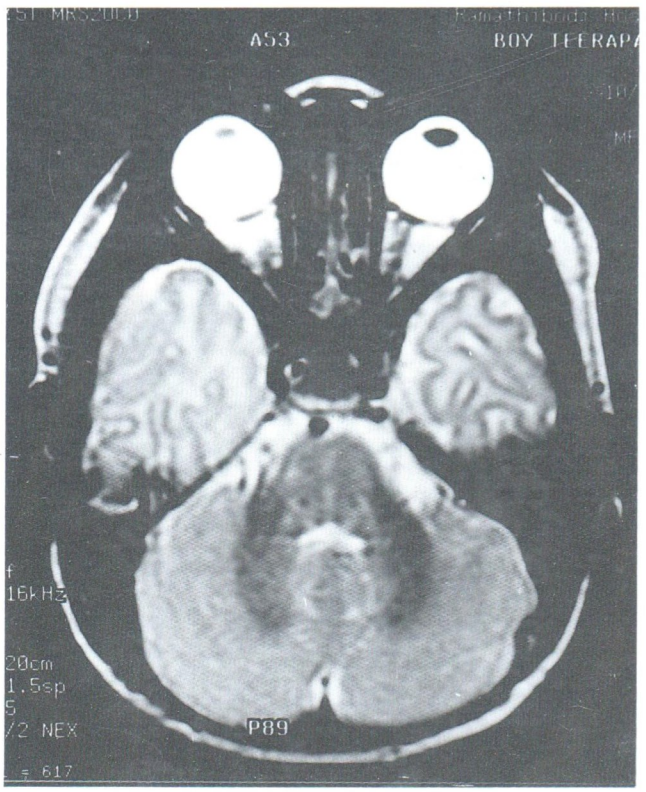


Fig. 2A. T_2WI of the posterior fossa.

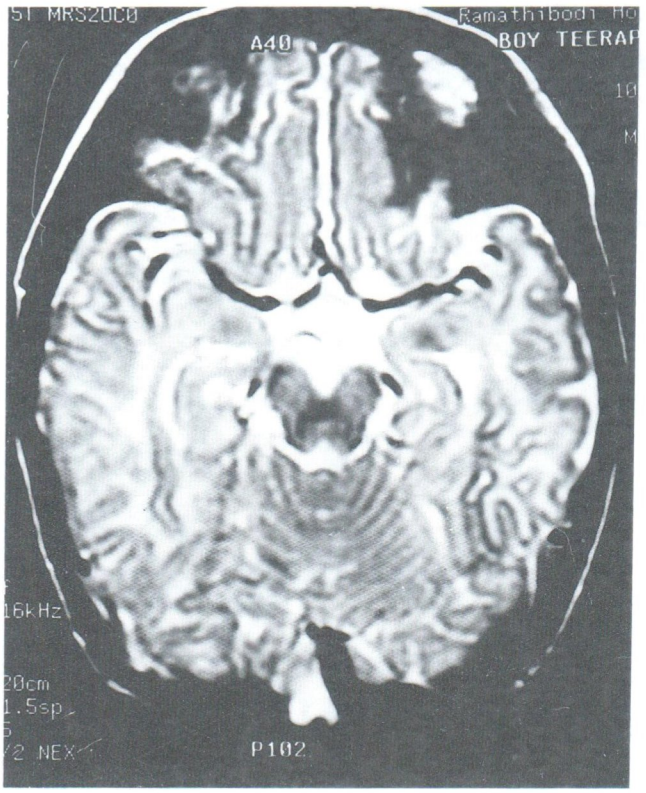
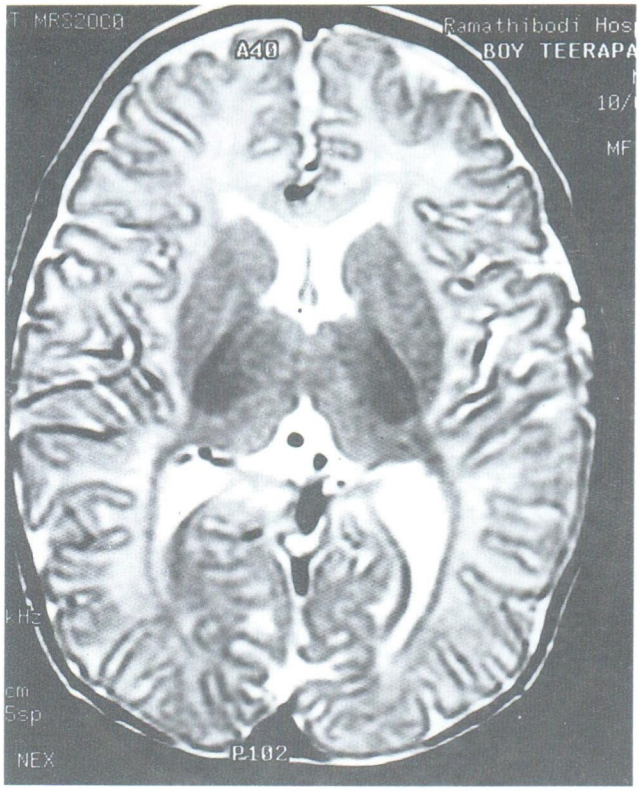


Fig. 2B. T_2WI of the basal part of the brain

Both figures showed signal drop of the lesions.

References (continued)

6. Weston JA: The regulation of normal and abnormal neural crest cell development, in Riccardi VM, Mulvihill JJ (eds): Neurofibromatosis (Von Recklinghausen Disease). Genetics, Cell Biology, and Biochemistry. Advances in Neurology, Vol 29. New York: Raven Press, 1981, pp 77-96.
7. Leaney BJ et: Neurocutaneous melanosis with hydro- cephalus and syringomyelia. J Neurosurg 1985;62: 148-152.
8. Bouton J: Primary melanoma of the loptomeninges. J Clin PATHO 1958;11:122-127.
9. Harper CG, Thomas DGT: Neurocutaneous melanosis. J Neurol Neurosurg Psychiatry 1974;37:760-763.
10. Fox H: Neurocutaneous melanosis, in Vinken PJ, Bruyn GW (eds): The Phakomatoses. Handbook of Clinical Neurology, Vol 14. Amsterdam: North-Holland, 1972, pp 414-428.
11. Slaughter JC, Hardman JM, Kempe LG, et al: Neurocutaneous melanosis and leptomeningeal melanomatosis in children. Arch Pathol 1969;88: 298-304.
12. Berlinger W: Ein Beitrag zur epithelialen Genese des Melanins. Virchows Arch (Pathol Anat) 1915;29: 328-365.
13. Fox H, Emery JL, Goodbody RA, et al: Neurocutaneous melanosis. Arch Dis Child 1964;39:508-516.
14. Farnell FJ, Globus JH: Primary melanoblastosis of the leptomeninges and brain. Arch Neurol Psychiatry 1931; 25:803-823.
15. Grahl F: Angeborener ausgedehnter Naevus pigmentosus in Verbindung mit Pigmentflecken im Gehirn. Beitr Pathol Anat 1906;39:66-81.
16. Ishikawa T, Nishi T, Shimada H, et al: Two cases of melanose neurocutanee with development of malignant melanoma: a microspectrophotometric and electron microscopic study. Gann 1975;66:277-289.
17. Bjoerneboe M: Primares Melanosarkom des Gehirns, massenhafte Naevi pigmentosi der Haut, ausgedehnte Neurofibromatose der Hautnerven. Frankfurt Z Pathol 1934;47:363-373.
18. Brandt RA, Schwartzman JS, Naspita C: (Neurocutaneous melanosis. Report of a case). Arq Neuropsoquiatr 1978 (Por); 36: 147-154.



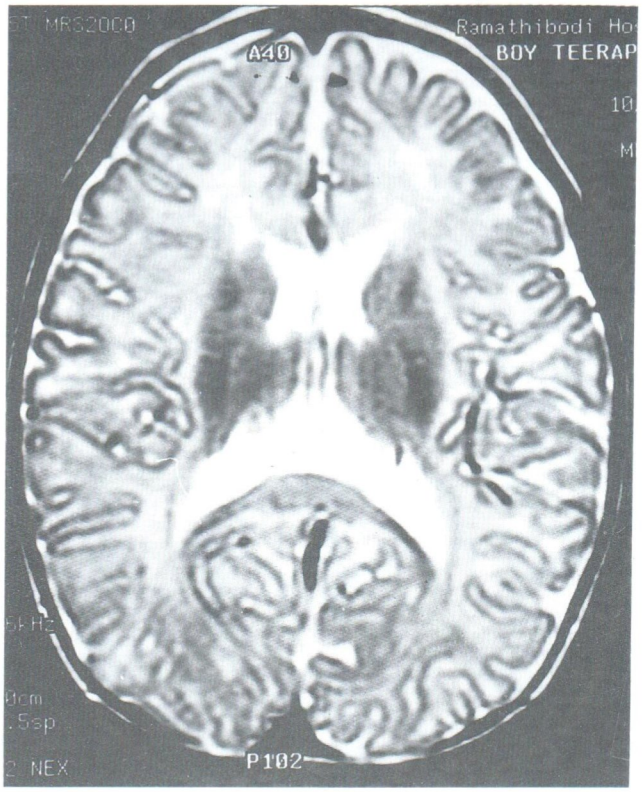


Fig. 2C. and Fig. 2D. showed T_2WI of the brain, the bright mass of the CSF obscured the bright signal deposits at the meninges.

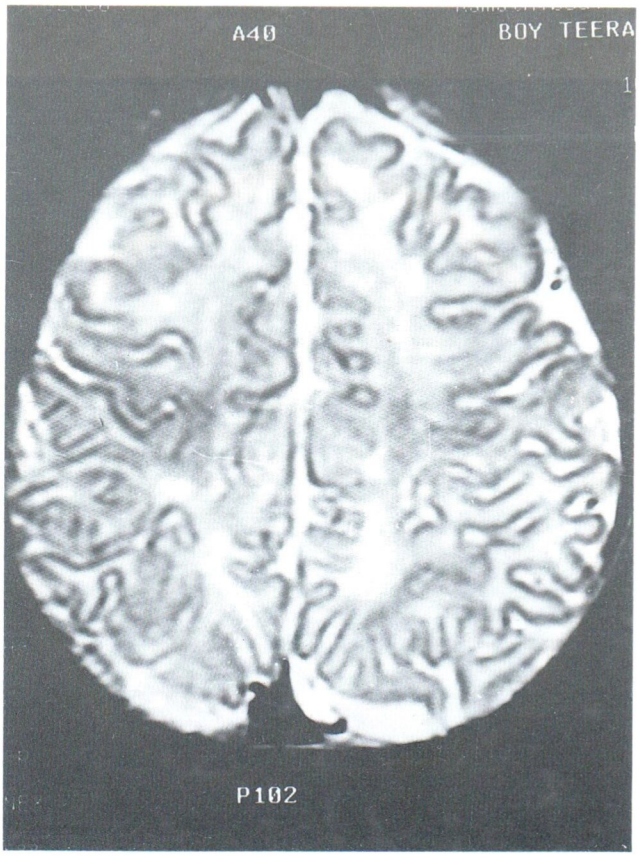


Fig. 2E. T_2WI of the brain at vertex showed obscuration of the meningeal deposits by the bright CSF signal.

References (continued)

19. Daily R, Forthomme J, Samson M, et al: Melanose neuro-cutanee a evolution tumorale. Une observation anatomo-clinique. Presse Med 1965;73:2867-2872.
20. Reed WB, Becker SW, Becker SW Jr, et al: Giant pigmented nevi, melanoma, and leptomeningeal melanocytosis. A clinical and histopathological study. Arch Dermatol 1965;91:100-119.
21. Komatus S,Takaku A, Suzuki J: (Neurocutaneous malanosis with chronic subdural hematoma on the left side and later on the right side-a case study). Clin Neurol 1974 (Jpn);14:855-860.
22. Medoc J, Folle JA: Malignant diffuse melanosis of the meninges of the neuro axis causing a hemorrhagic cerebrovascular type accident. Acta Neurol Lat Am 1969;15:106-114.
23. Hoffman HJ, Freeman A: Primary malignant leptomeningeal melanoma in association with giant hairy nevi. Report of two cases. J. Neurosurg 1967; 26:62-71.
24. Fasske E:Zur Genese der primaren Melanosarkome der Leptomeninx bei Saeuglingen. Zentrabl Allg Pathol 1958;98:113-121.

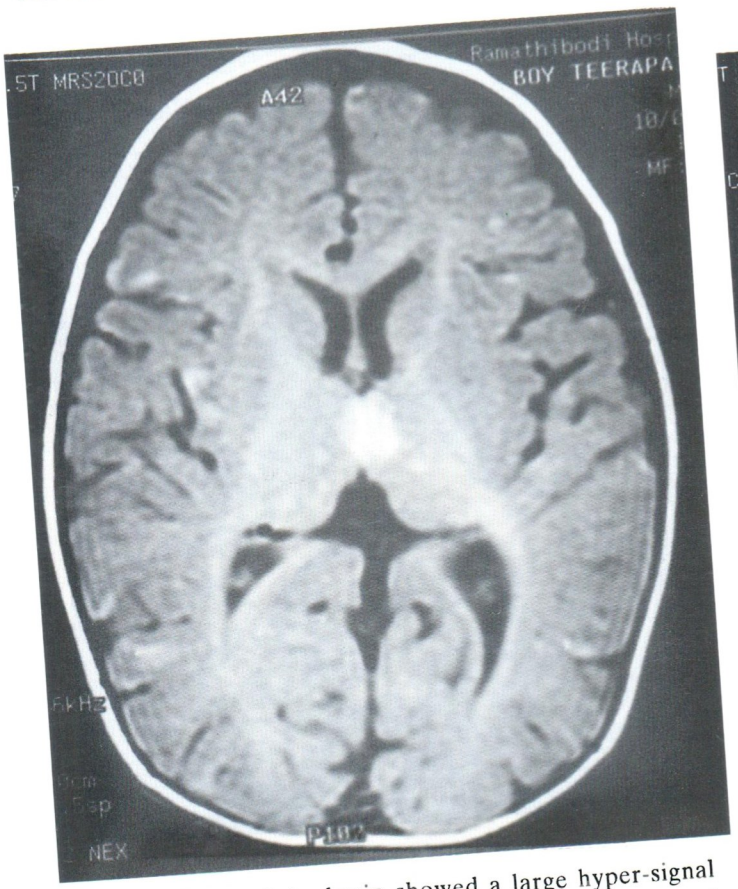


Fig.3A. T1WI of the brain showed a large hyper-signal nodule at 3rd ventricular roof and smaller meningeal deposits at convexity and at both thalami.

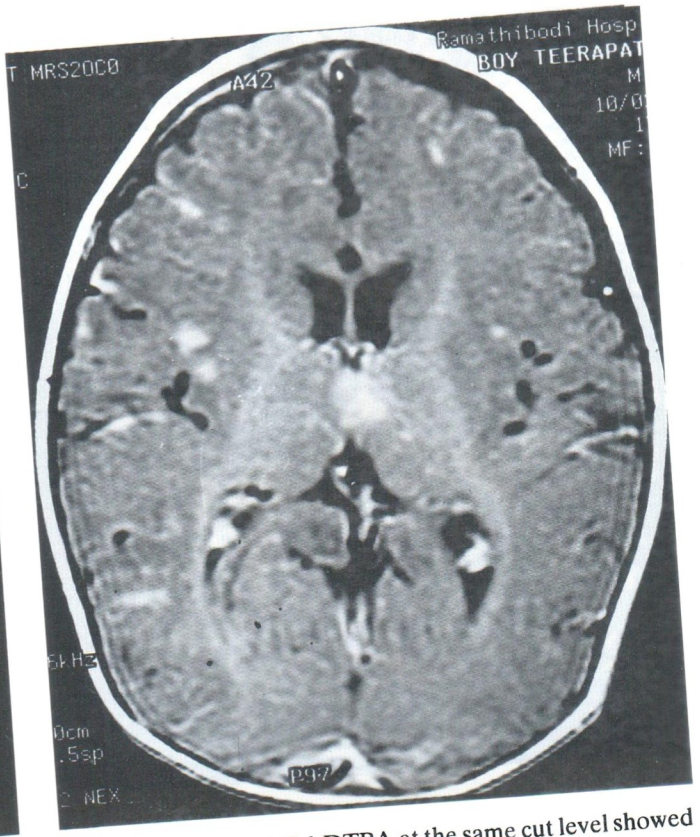


Fig. 3B. T1WI post Gd-DTPA at the same cut level showed no significant enhancement of the lesions.

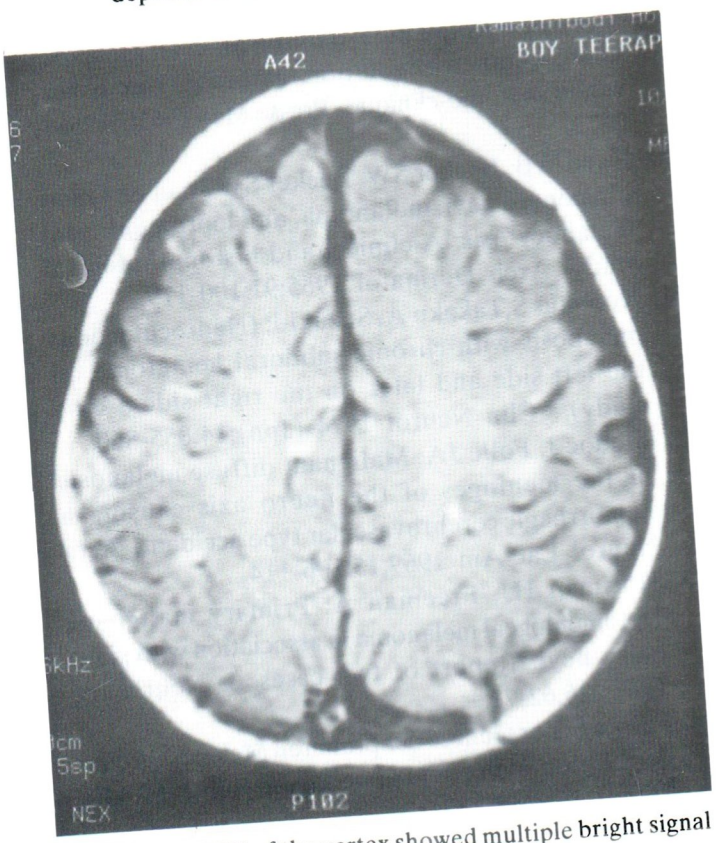


Fig. 3C. T1WI of the vertex showed multiple bright signal meningeal deposits

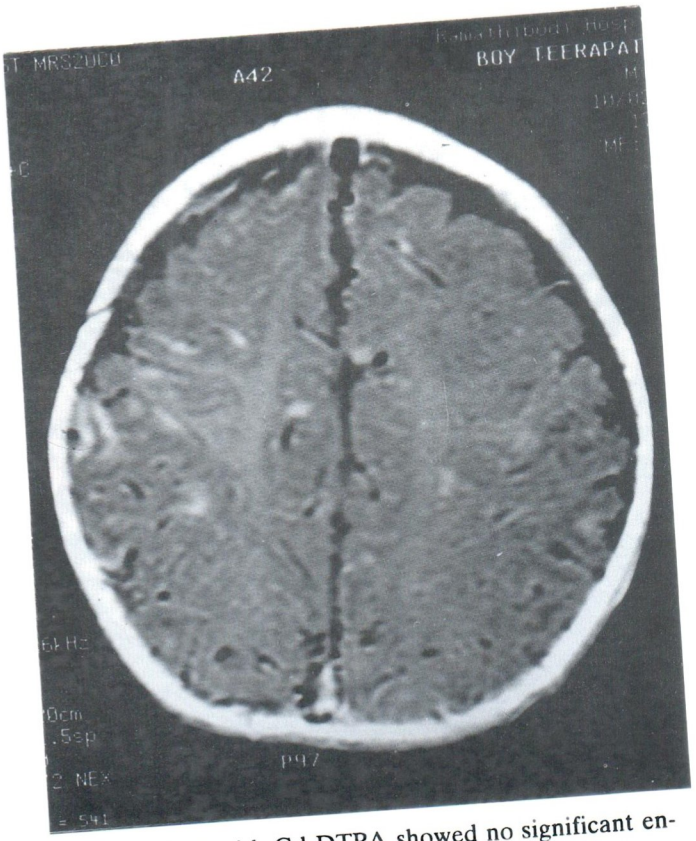


Fig. 3D. T1WI with Gd-DTPA showed no significant enhancement of the lesion .