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Diffuse meningeal melanosis
(An unusual presentation)

by

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Abstract

An unusual presentation of diffuse meningeal melanosis with involvement of the temporal bones, the spheno-ethmoidal complex, the pharynx and the viscera is reported.

Introduction

Melanosis of the temporal bones may occur in the leptomeningeal type of melanosis (Babin and Benjamin, 1976). Henschen, until 1955, had reported 70 patients with melanosis of the brain; 33.33 per cent had presented with pigmented nevi. Hydrocephalus was the cause of death in five cases before their first year of life. Netherton (1936), Wilcox (1939) and Fanconi (1956) considered cerebral melanocytosis to be nevoid in character; others, on the basis of the cytology and infiltrative ability, are inclined to regard it as neoplastic. We have found no case of diffuse meningeal melanosis reported with melanotic nodules and involvement of both temporal bones, spheno-ethmoidal complex, pharynx and the viscera.

Case report

B.R., a 25-year-old man, presented to the ENT department of the All India Institute of Medical Sciences, in September, 1984. He complained of painless, progressive bilateral diminished vision for the last three months. There was no history of diplopia or field cut. For the same duration he had had diffuse headache, most marked in the vartex and occipital region. The headache was not associated with vomiting and the intensity of pain had increased over the previous two months. The headache was relieved by cerebral decongestants. There was no history of fits, unconsciousness, cranial nerve palsy, motor or sensory deficit, secondary endocrinological or hypothalamic dysfunction, spiky fever, tinnitus, vertigo, café-au-lait spots or any subcutaneous swellings on the body. The past history revealed chronic suppurative otitis media in both ears during childhood. A family history of diabetes mellitus was elicited.

General physical examination revealed a well-built and well nourished young man who was blind. There was no anemia, jaundice, significant lymphadenopathy, cafe-au-lait spots or subcutaneous swellings over the body. Respiratory, cardiovascular and abdominal examination was unremarkable. The central nervous system was normal except that there was no perception of light in either eye. Fundoscopy revealed bilateral secondary optic atrophy.

ENT Examination: There was bluish-black pigmentation on the posterior pharyngeal wall opposite the soft palate (Fig. 1c). On posterior rhinoscopy there was pigmentation of the nasopharynx. There was no mass or pigmentation on anterior rhinoscopy.

EARS: Diffuse bluish-black discoloration of both drums was seen (Fig. 1a,b). Tuning fork tests and audiometry revealed a false Rinne negative response on both sides. The Weber was not lateralised. ABC was shortened on both sides. Recruitment was positive on both sides. Tone decay was negative. Lateral X-rays of the skull showed enlargement of the sella. The X-rays of the mastoids and internal auditory meati were normal. CT scan showed evidence of a high attenuation mass on the surface of the right cerebellum, also posterior to the fourth ventricle (paramedian).

Biopsy from the posterior pharyngeal wall, nasopharynx, both the middle ears, sphenoid and ethmoid sinuses were reported as melanosis. Cerebrospinal fluid, routine blood and urine examination were within normal limits.

A trans-sphenoidal biopsy was attempted follow-

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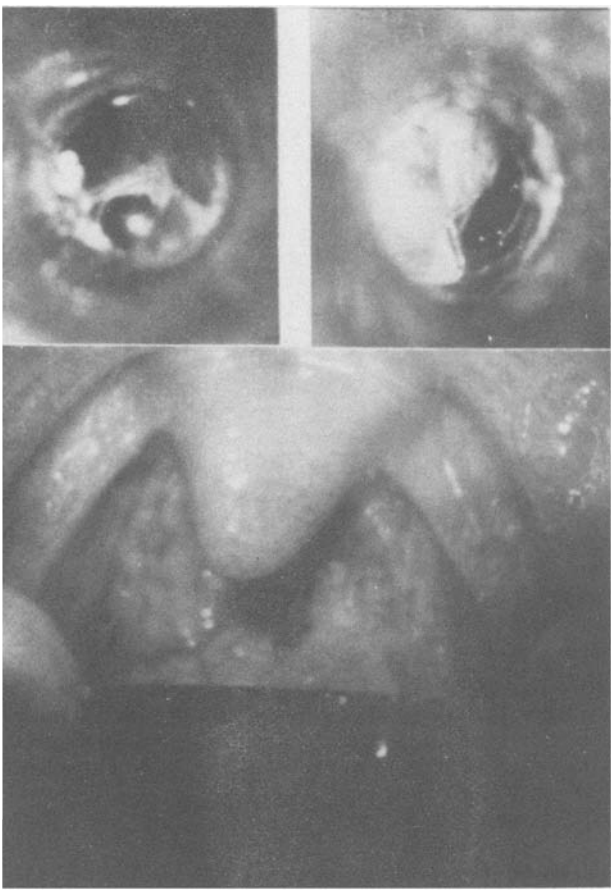


FIG. 1

Bluish-black pigmentation of the right tympanic membrane (a), left middle ear cavity (b), and posterior pharyngeal wall (c).

ing which he developed acute meningitis; and despite heavy antibiotics and cerebral decongestants he succumbed on 16 November 1984. The clinical diagnosis entertained at the time of death was diffuse meningeal melanosis, malignant melanoma with involvement of the sphenoid and ethmoid sinuses, ears and the pharynx.

Histopathology

Biopsies from the black areas in the ears and posterior pharyngeal wall showed melanin-laden cells in the submucosa with mild pleomorphism evident after bleaching the section. No mitoses were seen (Fig. 2).

At necropsy diffuse black pigmentation of the dura mater and leptomeninges of the brain and spinal cord, inner table of occipital, sphenoid bones, cerebellar folia and pituitary was seen. A well circumscribed, sessile, black dural nodule measuring 2.5 cm. in diameter was attached to the tentorium cerebelli, in relation to the cerebellum to the right of the midline (Fig. 3). The right cerebellar hemisphere in relation to the nodule was compressed and indented. There was bilateral tonsillar herniaton and left-sided uncal herniation. The sulci on the superior surface of the brain were full of purulent exudate. The cut surface of the brain showed fresh intracerebellar haemorrhage adjacent to the dural nodule in addition to fresh subarachnoid and focal intracerebral haemorrhages.

On microscopic examination the black nodule consisted of closely packed spindle cells arranged in interlacing fascicles and bundles. These cells had a

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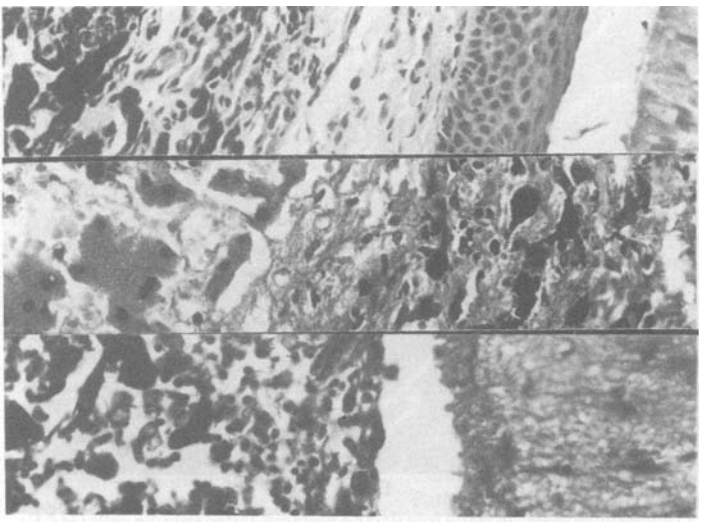


FIG. 2

Melanin-laden cells in the pharynx (top), liver (middle) and leptomeninges (bottom). (H&E: ×300).

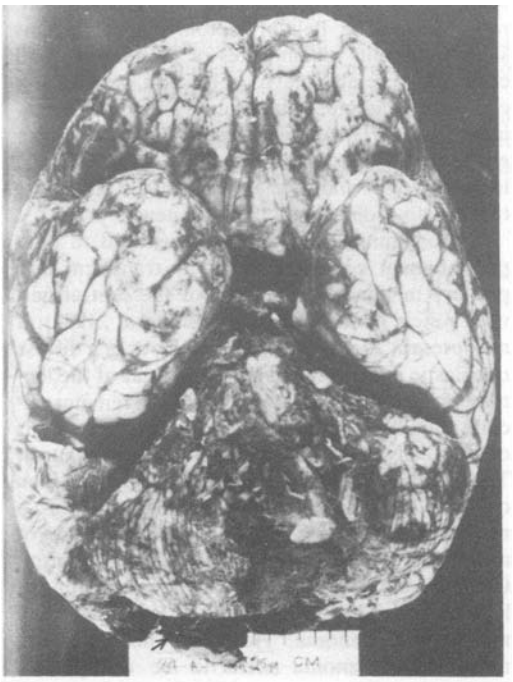


FIG. 3

Diffuse meningeal melanosis with a pigmented nodule.

moderate amount of cytoplasm loaded with melanin (Fig. 4). The nuclei were round-to-oval and vesicular, with a prominent single eosinophilic nucleolus. There was mild pleomorphism but an extensive search failed to show any mitoses. The morphologic appearance was closely akin to that of a pigmented cellular blue naevus. The leptomeninges and dura mater revealed macrophages and spindle cells laden with melanin (Fig. 2). The section from the posterior pituitary was infiltrated heavily by pigment-laden spindle cells similar to the dural nodule. There was an area of ischaemic necrosis in the anterior pituitary. The gross findings of intracerebral, intracerebellar and subarachnoid haemorrhages along with pyogenic meningitis were confirmed on microscopic examination.

The external and cut surface of the liver showed multiple, black pigmented areas varying from 0.2 to 1.5 cm. in diameter with intervening normal liver tissue. Microscopically the pigmented cells were confined to the portal tracts and were seen to spill into the adjacent sinusoids (Fig. 2). Some of the Kupfer cells phagocytosed melanin pigment. The lungs were grossly unremarkable except for multiple black, discrete areas on the pleural surface measuring 0.2–0.4 cms. in diameter. On microscopic examination, spindle cells packed with melanin were seen distributed either subpleurally or around bronchial blood vessels. Foci or bronchopneumonia were also observed. No mitoses were found in the lungs and liver as in the brain. The rest of the organs were unremarkable.

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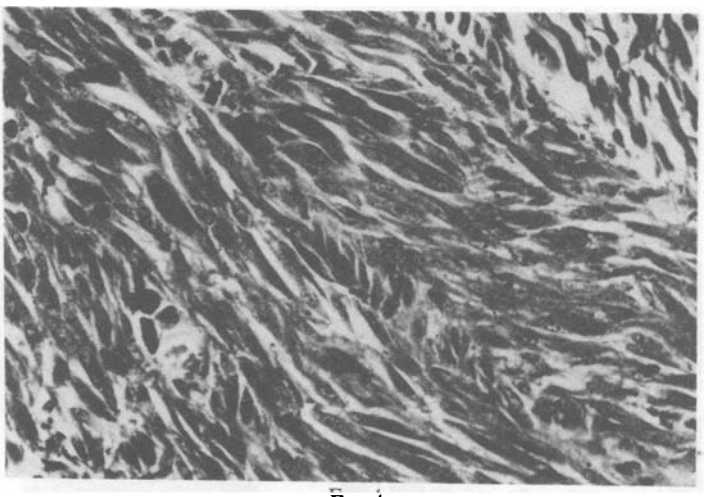


FIG. 4

Section from nodule showing interlacing melanin-laden spindle cells (H&E: ×300)

Discussion

Melanosis of the head and neck is far less common than malignant melanoma (Babin and Benjamin, 1976). A case report of a naevus arising in the middle ear is available, but primary malignant melanoma has not been reported till recently (McKenna *et al.*, 1984). Thus, pigmentation of the middle ear may occur in the benign form of meningeal melanocytosis (Reed *et al.*, 1965). Melanin-bearing cells are normally found in the leptomeninges in adults and the melanoblasts are most numerous on the ventral aspect of the lower medulla, upper part of cervical spinal cord and inter-lobular septa crossing the pineal gland (Russel and Rubinstein, 1977). It is postulated that the melanin-producing cells are capable of producing melanotic tumours which may be benign or malignant. These tumours often spread diffusely along the leptomeninges, mainly over the basal surfaces, or they form meningeal or parenchymatous collections (Deshpande *et al.*, 1970), as also experienced in our case.

Meningeal melanosis is usually associated with giant cutaneous nevi and collectively is known as the neuro-cutaneous melanosis syndrome (Harkin and Reed, 1969). It is a non-familial disorder in which diffuse and often frankly invasive proliferation of melanin-containing cells occurs throughout the leptominges (Hoffman and Freeman, 1967; Slaughter *et al.*, 1969). It is usually found at birth or in infants under the age of two years. Excessive basal leptomeningeal involvement may lead to internal hydrocephalus. This was also evident in our case with signs of raised intracranial tension. Despite an extensive search on admission and at autopsy, no cutaneous nevi could be spotted. In neuro-cutaneous melanosis the benign cutaneous lesions may show features of the ordinary cellular blue nevus, neurofibroma and melanocytic neurofibromas. The benign leptomeningeal lesions show numerous melanocytes or clusters of small cells that resemble nevus cells, and perivascular collections of pigment-laden cells are often a prominent feature (Harkin and Reed, 1969). In the present case, histologically the cells in the leptomeninges and in the dural nodule show features of an ordinary cellular blue nevus.

Leptomeningeal melanosis is believed to be a premalignant condition and may transform into primary malignant melanoma of the central nervous system (Bouton, 1958; Gibson *et al.*, 1954). It may present as a circumscribed tumour or a diffuse neoplastic process involving wide areas of the leptomeninges. Occasionally, the growth remains circumscribed and relatively benign; more often it is cytologically malignant with many mitotic figures. These patients usually do not survive more than one year. In the present case, the feature against a malignant melanoma is the absolute absence of mitotic figures, with only mild pleomorphism. Cytologically, therefore, it was more in favour of leptomeningeal melanosis than of leptomeningeal malignant melanoma. However, melanosis and malignant melanoma seem to be at opposite extremes of a spectrum rather than two distinct entities; transitional forms may thus be seen quite often (Rubinstein, 1972).

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In the lungs and liver there were similar nests of melanin-containing cells. These were not believed to be metastatic foci from a primary melanoma of the brain, for two reasons: firstly, primary malignant melanoma of the central nervous system very rarely metastasizes outside the latter (McKenna *et al.*, 1984; Russel and Rubinstein, 1977); secondly, the melanin-containing cells with the liver and the lungs appeared to be histocytes with phagocytosed melanin pigment. There was no destruction of the parenchyma. Thus, these melanophages were considered to be a benign process being involved as a part of the diffuse leptomeningeal melanosis.

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