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CT of Sebaceous Nevus Syndrome (Jadassohn Disease)

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The sebaceous nevus, a congenital skin disease, was first described in 1895 by Jadassohn [1] and later became known as the organoid nevus syndrome within the grouping of the phacomatoses [2]. In 1957, Schimmelpenninck [3] described its association with central nervous system, skeletal, and eye lesions. In 1962, Feuerstein and Mims [4] reported a case with epilepsy and mental retardation. In the last 10 years, 19 cases presenting with a wide variety of symptomatology have been reported in the literature [5-19]. We report a case of Jadassohn disease in which, to the best of our knowledge, both conventional axial and direct sagittal computed tomography (CT) were performed for the first time.

Case Report

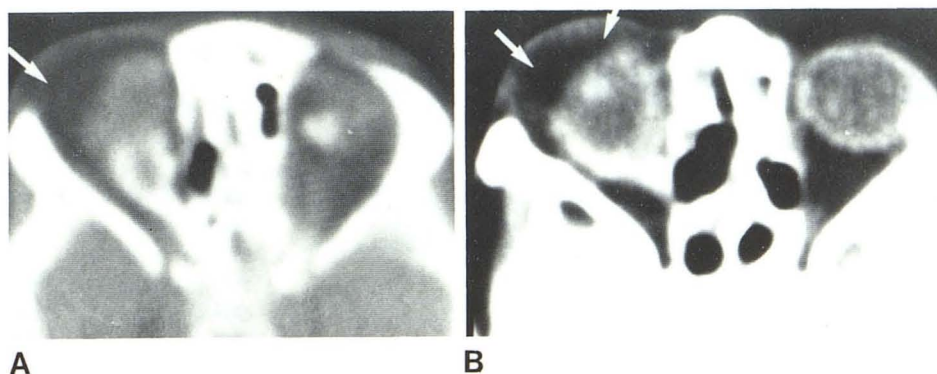
A 4-year-old boy had epileptic fits (infantile spasms), mental retardation, and amaurosis. Clinical examination showed multiple yellow papules on the right cheek and neck. The right eyelid was deformed by a soft-tissue mass (fig. 1). Lipodermoid deposits were embedded in both cornea. Macrophthalmia was noted on the right side. Apart from epilepsy and mental retardation, neurologic investigation showed no other abnormalities.

Conventional radiographs demonstrated an enlarged right orbit as well as calcifications at the apex of the orbit. CT demonstrated not only hypertelorism and thickening of the orbital walls, but also a large calcified mass involving the posterior wall of the right globe and the optic nerve (fig. 2A) and swelling of the eyelid (fig. 2B). A



Fig. 1.—Multiple papules on right cheek, eyelid, and upper lip.

Fig. 2.—A, Macrophthalmia of right eye. Soft-tissue lesion of eyelid (arrow). B, Calcifications in posterior wall of both globes and in right optic nerve. Lipoma (—56 H) of right eyelid (arrows).



smaller area of calcification was also present in the posterior wall of the left globe (fig. 2A). Conventional axial scans showed asymmetry of the ventricular system with an enlarged right lateral ventricle (fig. 3). These findings were confirmed by direct sagittal CT, which also demonstrated prominence of the basal ganglia (fig. 4B)

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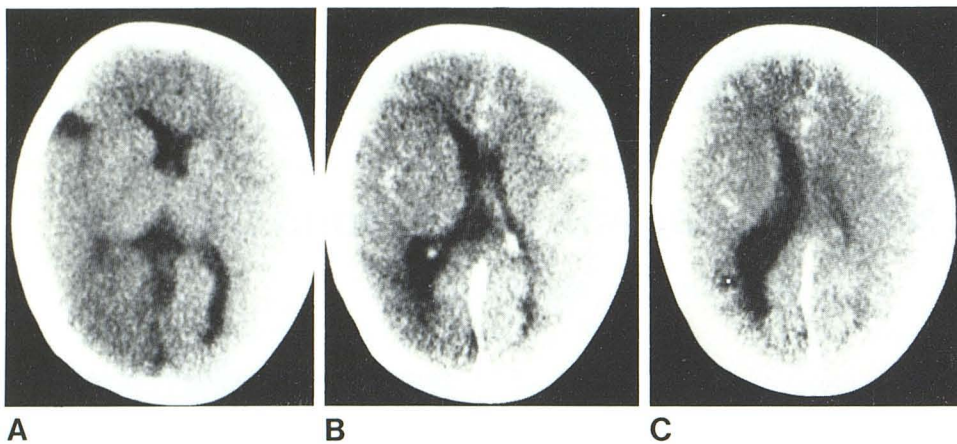


Fig. 3.—A, Without contrast. B and C, With contrast enhancement. Right ventricle is enlarged. Asymmetry of thalamic masses.

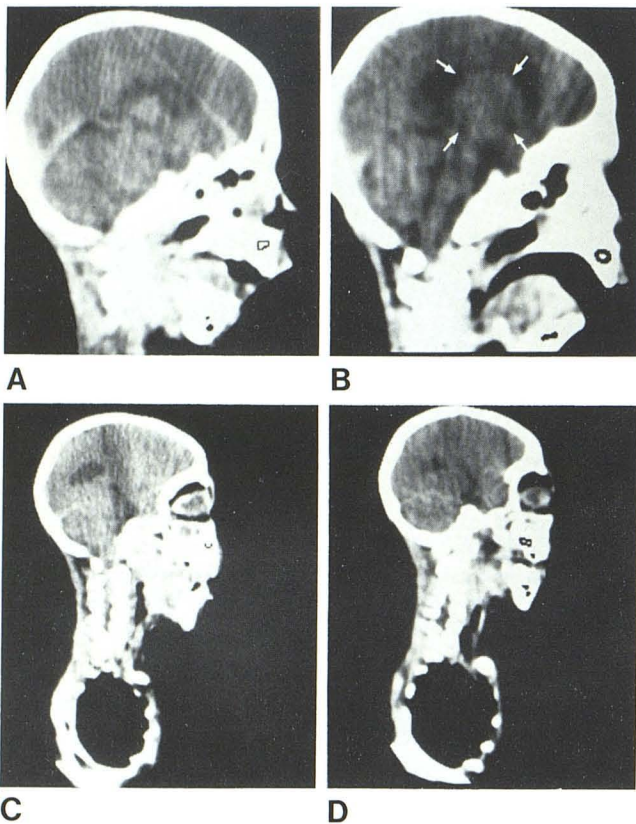


Fig. 4.—Direct sagittal CT of head and neck. A, Midline view. B, Right of midline. Enlarged lateral ventricle; prominence of ipsilateral basal ganglia (arrows). C, Further to right. Dilatation of occipital horn. Calcification of globe and optic nerve. D, Flattened posterior wall of eye socket and distribution of lipomatous tissue above and below globe.

and a deformed eye socket with a flattened posterior wall (figs. 4C and 4D) on the right side. The right-sided intraorbital calcifications were shown again in the posterior wall of the globe and in the optic nerve (fig. 4C). The actual spatial display of the deformed right eye socket and distribution of the lipomatous tissue (-56 Hounsfield units [H]) laterally and in the upper part of orbit were complemented

by the sagittal scans. A biopsy of the right eyelid demonstrated an angiolipoma.

Discussion

The CT appearances of the sebaceous nevus syndrome have been described in the conventional CT mode by van Balen [5], Bolthausen and Navratil [17], Leonidas et al. [18], and Kurokawa et al. [19]. Our patient was examined by both conventional and direct sagittal CT, which confirmed the presence of the abnormalities of the brain and eyes. Direct sagittal CT demonstrated the three-dimensional distribution of the pathologic changes of the brain and right eye. Furthermore, the basal ganglia were prominent on the right side.

Our patient had skin, ocular, and central nervous system abnormalities as well as psychomotor retardation and epilepsy. In the past 10 years, 19 other cases of sebaceous nevus syndrome have been described. Similar abnormalities were found in all the other reported cases. Not only were we able to demonstrate the degree of dilatation and asymmetry of the ventricular system, but we were also able to confirm that the atrophic cerebral hemisphere is on the same side as the skin lesions. In the literature, asymmetry of the ventricular system has been documented by pneumoencephalography (five cases), cerebral angiography (two cases), and CT (three cases). All of these cases except one had electroencephalographic epileptic abnormalities. The other cases were not completely investigated neurologically, but there were epileptic seizures in seven. Other cerebral abnormalities were cortical dysplasia (one case), cortical atrophy (one case), and an intracerebral lipoma (one case). In our case both eyes were involved (figs. 1, 2, and 4), though other reports indicate an apparently ipsilateral predominance. The most frequent orbital lesions, as in our case, were lipodermoid deposits (four cases) and microphthalmia (four cases). In one case there was only a coloboma and hemangioma, and in one case there was a brushfield spot of the iris. However, no previous reference to calcification of globe and optic nerves was found.

We believe that the sebaceous nevus syndrome is probably not genetically linked, but is developmental. Because

the condition is predominantly asymmetrical, it is suggested that it occurs very early in gestation (4–12 weeks) [20]. Neither in our case nor in the literature was the cause evident. The incidence of sebaceous nevus syndrome is very low considering that only 19 cases have been reported in the last 10 years. In comparison, however, the incidence of the most common forms of phacomatosis, such as neurofibromatosis, tuberous sclerosis, and Hippel-Lindau disease, are 1:3,000, 1:50,000, and 1:300,000 births, respectively. We believe that our case illustrates that multiplanar direct and axial CT scans are helpful and complementary in the diagnostic workup of sebaceous nevus syndrome.

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