COMBINED RETINO-CEREBELLAR ANGIOMATOSIS AND DEEP CERVICAL ANGIOMAS

CASE REPORT

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I have been unable to find, after a thorough perusal of the literature, any case of retino-cerebellar angiomatosis associated with deep cervical angiomas. Worthy of note therefore is a case of such a syndrome which I have had occasion to study from a roentgenological point of view.

CASE REPORT

M.M. was a 26-year-old male with unremarkable familial anamnesis. At the age of 16 he noticed gradual and progressive dimness of vision, first of one eye and later of the other eye. In approximately 3 years the patient became blind. At the age of 24, i.e. 2 years before ente-
ing the Neurological Institute of the University of Naples, subjective sensorial disturbances, characterized by tingling and numbness of the tips of the fingers, while reading by the Braille system, were noticed. These disturbances were then followed by severe pain, of radicular type, radiating from the nuchal region down into both upper limbs, much more so into the left one. Furthermore, symptoms and signs of increased intracranial pressure developed and the gait became impaired.

At the Neurological Institute the diagnosis of typical bilateral retinal angiomatosis was reached and the presence of a space-occupying lesion in the posterior fossa was strongly suspected. However, no satisfactory explanation for the radicular pains was given.

Plain roentgenograms of the skull showed no clear-cut signs of increased intracranial pressure. The calcified pineal gland was not displaced. In each orbit a linear, circular calcification was present, the size of which exactly reproduced the posterior and middle portions of the eyeballs (Fig. 1).

The left vertebral angiograms (Figs. 2 and 3) revealed the presence of at least two posterior fossa and two deep cervical angiomatous formations. The two intracranial angiomatous formations had the appearance of two homogeneous rounded sacs, the size of these being that of a bean, fed by branches of the posterior inferior cerebellar artery; no definite displacement of small vessels, surrounding these sacs, pointing to a possibly associated cystic tumor, was seen. The inferior portion of the basilar artery was pressed against the clivus. The anterior loop of the posterior inferior cerebellar artery did not descend beyond the foramen magnum. The two deep cervical formations were located at the level of and behind the posterior border of the body of the 3rd cervical vertebra. The upper one had irregular contours and was not homogeneously dense. Its size was that of a hazelnut. The lower formation, which more closely resembled the intracranial ones, was as large as a bean. Both structures were fed by branches of the vertebral artery and were drained, as clearly seen in the venous phase (Fig. 3), by a very peculiar bead-shaped vessel, directed upward, toward the base of the skull.