Cerebral Gigantism

Sir.—It was a pleasure to read the neuropathologic study of the hypothalamus and pituitary in cerebral gigantism by Whitaker and co-workers.[1] In their discussion they stated that there is no known explanation for this condition. However, I was involved in a series of investigations with two children who had this affliction and was able to make a number of novel observations.[2] Since this article appeared in a less well-read publication, I thought that it would be of benefit to summarize it and its results here.

In these two children, aged 10 and 19 months, who had characteristic features of cerebral gigantism, the levels of serum growth hormone, somatomedin C, nerve growth factor, and epidermal growth factor were unremarkable. Fibroblasts derived from a skin biopsy specimen from one child, when grown in confluent monolayer culture, demonstrated epidermal growth factor and somatomedin C receptor concentrations that did not differ from those of controls. Qualitative growth kinetics of these fibroblasts were also unremarkable.

Serum and plasma protein dialysate from one child significantly stimulated the growth of AKR-2B mouse fibroblasts in tissue culture when compared with adult and age-matched controls. This effect was reproducible.

Confirming the statements of Whitaker et al, these results indicated that the currently assayable growth factors and fibroblast receptors for epidermal growth factor and somatomedin C are unremarkable in cerebral gigantism. The serum and plasma protein stimulation of AKR-2B mouse fibroblast growth in one child may be a demonstration of a novel growth factor in this condition.

The reason one child demonstrated such a serum and plasma property and the other did not may be an age-related effect. Alternatively, cerebral gigantism may be a heterogeneous disease with different causes. This syndrome has been associated with a variety of disease processes including thyroid abnormalities, cerebral dysgenesis, macular degeneration, autonomic insufficiency, peripheral dysostosis, and intestinal polyposis. All of these diverse associations argue that this syndrome may arise from various causes, one of which may be related to this demonstrable growth effect.

The laboratory investigations summarized above were conducted in the Cell Biology Laboratories of the Mayo Clinic (Rochester, Minn) and at Stanford (Calif) University Medical Center.

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1. Whitaker MD, Scheithauer BW, Hayles AB, et al: The hypothalamus and pituitary in cerebral gigantism: A clinicopathologic and immunocytochemical study. *AJDC* 1985;139:679-682.

2. Plioplys AV, Childs CL, Rosenfeld RG, et al: Growth factors and fibroblast growth factor receptors in cerebral gigantism, in Berg JM (ed): *Perspectives and Progress in Mental Retardation*. Baltimore, University Park Press, 1984, vol 2: *Biomedical Aspects*, pp 257-264.