Sphenopharyngeal Meningoencephalocele: Unusual Clinical and Radiologic Features

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AJNR Am J Neuroradiol 1989, 10 (5 suppl) S80
http://www.ajnr.org/content/10/5_suppl/S80.citation

This information is current as of August 17, 2024.
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A 42-year-old woman was admitted for further investigation of deficiency of growth hormone and gonadotropin first documented in 1969. On admission, the patient was mildly obese and of short stature (48 kg for 134 cm). There was no evidence of facial or palatal abnormality; fundoscopy was normal, and both visual fields and sense of smell were intact. Clinical examination also was normal.

The following radiologic examinations were carried out: radiography of the skull; frontal and lateral hypocycloidal tomography of the sphenoid bone; angio-CT of the sella turcica; metrizamide computed cisternography; and MR imaging. The radiologic findings included a sharply demarcated funnel-shaped bony defect stretching in a slightly anterior and inferior direction between the floor of the sella turcica and the pharyngeal surface of the basisphenoid. A round structure was seen hanging beneath this defect, in the nasopharynx. This structure appeared to contain the structures of the group, Currarino et al. [3] described a classification based on the size of the bony defect.

Regardless of the group, Currarino et al. [3] propose the same underlying defect in embryonic development, explaining the frequent associated dysraphic anomalies. This case presents several unusual features. The means of presentation as isolated endocrine deficiencies is exceptional. It involves a 13-mm wide bony defect and thus belongs to group 3 (classification of Currarino et al. [3]), although we did not find a dilated third ventricle, as has been reported in all previous cases. No associated anomalies were found; there was no facial abnormality, even occult, and ophthalmologic examination was normal. It is therefore interesting to recall another theory of the pathogenesis proposed by Danoff et al. [4] and McCoy [5].

In our patient, the site of the bony defect is in the medial part of the postphenoid. Indeed, endochondral ossification of the postphenoid originates from two lateral nuclei, allowing speculation that the defect may have resulted from inadequate medial fusion of these elements. This would explain the formed lateral walls of the basisphenoid and the absence of other associated dysraphic abnormalities. Whatever the underlying pathogenesis, the herniation must be considered secondary and aided by the absence of a dural covering in the floor of the sella at the level of the bony defect, and by an incompetent diaphragm sellae. Thus, under the influence of CSF pressure, the parasellar and sellar structures descend, which explains the endocrine findings.

In summary, the isolated endocrine findings, the anatomic configuration, and the absence of associated dysraphic anomaly characterize this exceptional case. In addition, the possibility of another developmental abnormality (bony fusion) must not be forgotten in the explanation of isolated cases without other facial abnormality.

ACKNOWLEDGMENT

We are grateful to J. Crabbe for referring this patient.

REFERENCES


Fig. 1.—A, Coronal CT section of sella turcica shows liquid nature of mass in rhinopharynx (meningocele) (arrow) and invagination of optic nerves (arrowheads) into bony defect. B, Coronal CT section of sella turcica during metrizamide cisternography confirms the continuity between sphenopharyngeal meningocele (arrow) and optochiasmatic cistern. C, Midsagittal T1-weighted MR image (470/30) of sella turcica after gadolinium IV contrast shows locations of pituitary gland (black arrow), which lies posterior to bony defect, and infundibulum (white arrow). Arrowhead: meningocele pouch.