AJNR

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AJNR Am J Neuroradiol 1989, 10 (5) 1007-1010
http://www.ajnr.org/content/10/5/1007

MR Imaging of Middle Cranial Fossa Arachnoid Cysts: Temporal Lobe Agenesis Syndrome Revisited

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MR studies in eight patients with extraaxial arachnoid cysts in the middle cranial fossa were reviewed in order to identify any associated structural defect in the ipsilateral temporal lobe. The study was prompted by the original theory that agenesis of the temporal lobe is the primary factor in the development of these cysts. Authors of subsequent studies proposed that the cysts are a consequence of embryological malformation of the meninges only and that the adjacent temporal lobe is compressed.

Our findings suggest that middle cranial fossa cysts are associated with temporal lobe hypogenesis, and also that compression of the temporal lobe is an infrequent accompaniment.

AJNR **10:1007–1010, September/October 1989**

Arachnoid cysts are congenital CSF-filled collections that occur in specific locations such as over the convexity of the brain, in the middle cranial fossa, behind the cerebellum, or in the perimesencephalic cisterns. MR imaging has been established as an excellent method for demonstrating CNS structural abnormalities. The effect of cystic lesions on adjacent neural elements is particularly well seen [1]. In reviewing the MR changes found in patients with middle cranial fossa arachnoid cysts we noted that hypogenesis of the temporal lobe was present in all our cases. An analysis of the MR appearances of these arachnoid cysts forms the basis of this article.

Materials and Methods

Eight middle cranial fossa arachnoid cysts were found in seven patients during a review of a consecutive series of 24 intracranial cysts identified by MR. The patients consisted of four males and three females, 16 to 53 years old. All were examined with a 1.0-T Siemens imaging system. Imaging parameters included T1- and T2-weighted SE pulse sequences with 5- or 8-mm axial, coronal, and/or sagittal planes, and a 256 × 256 matrix. The interscan gap and number of acquisitions varied according to the type of coil and software package in place. Each patient was examined for hypogenesis or compression of the ipsilateral temporal lobe. Hypogenesis was considered to be present when there was (1) undulation of the margins of the temporal lobe adjacent to the cyst, together with loss of temporal lobe parenchymal tissue; (2) no displacement of the temporal horn or the adjacent brain structures, e.g., midbrain and perimesencephalic cisterns; and (3) no thinning of the squamous temporal bone or wing of the sphenoid bone. Thinning of the cortex of the temporal lobe was looked for and was not found to be a feature of hypogenesis. Thickening of the bone was also not an associated feature.

Compression was considered to be present when (1) the margins of the temporal lobe adjacent to the cyst were concave and (2) the temporal horn and/or the adjacent brain structures were displaced. Thinning or bulging of the adjacent bone was only present in the more severe cases, and therefore was not considered essential to diagnose compression.

Received October 14, 1988; revision requested December 19, 1988; revision received January 17, 1989; accepted January 18, 1989.

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0195-6108/89/1005–1007

Results

In five middle cranial fossa arachnoid cysts, temporal lobe hypogenesis alone was present (Figs. 1 and 2). The anterolateral aspect of the temporal lobe, most commonly the middle temporal lobe gyrus, was most frequently involved. In three cases, both compression and hypogenesis were present (Figs. 3 and 4). In one of the three patients, bulging of the squamous temporal bone was present. Abnormal signals in the compressed or hypogenetic temporal lobes were not seen in any of the cases.

Surgery was not carried out in any of the cases, since in none was the cyst considered to be causing increasing intracranial pressure. All patients were investigated for headaches or seizures, and the cysts were considered to be incidental findings.

Discussion

Arachnoid cysts represent 1% of all atraumatic intracranial masses with 50–66% occurring within the middle cranial fossa in most series. Ten percent occur in the suprasellar and quadrigeminal regions and approximately 5% each in the posterior fossa and over the frontal convexities [2–5]. Most investigators report that 60–80% of all arachnoid cysts are symptomatic, with the most common symptoms initiating MR imaging being seizures, headaches, and focal neurologic signs. Asymptomatic patients were in the minority in most series [3–7]. All patients in our series were imaged because of headaches or seizures except for one patient with posttraumatic rupture of bilateral cysts and the subsequent formation of subdural hygromas.

Two types of congenital supratentorial arachnoid cysts have been described [8]. The more common type is that of a dilated, circumscribed, expanded, subarachnoid space lined by the arachnoid membrane and external arachnoid cells on the external surface, and usually lined by pia and internal arachnoid cells on the inner surface. Embryologically, the subarachnoid space is considered to be formed by expansion of the intercellular space of the meninx primitiva, which surrounds the neural tube. The space is formed by clearing out of the cellular elements of the meninx [9]. The outer layer of the space condenses into a compact layer that becomes the dura and the outer layer of the arachnoid. The loose mesenchyme of the inner layer forms the pia and the inner layer of the arachnoid. Naidich et al. [10] reviewed the anatomy of the arachnoid and postulated that arachnoid cysts may arise as a minor aberration in the formation of the subarachnoid space. Variations in the condensation of the embryological meninx primitiva and/or slight variations in the flow of CSF into the

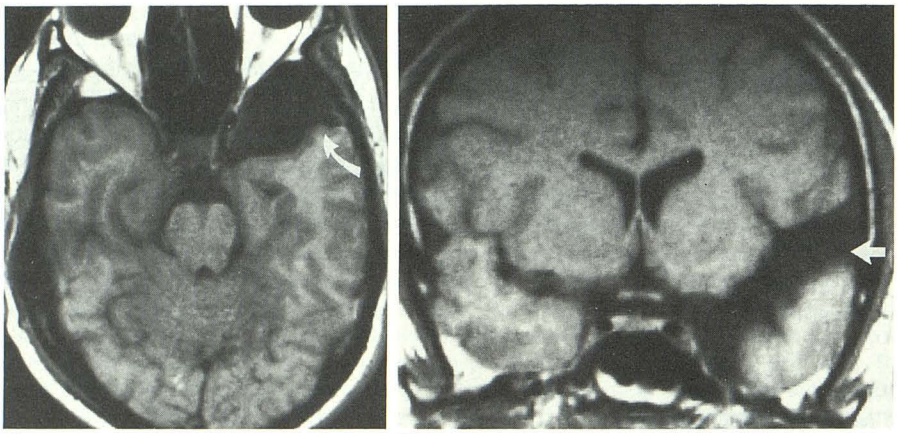

Fig. 1.—38-year-old man with large left middle cranial fossa cyst.

***A*, Axial T1-weighted image (650/20). Note that anterior temporal lobe is absent at this level and that residual margin is undulating (*curved arrow*). Left temporal horn is not displaced.**

***B*, Coronal T1-weighted image (650/20). Sylvian fissure is widened and superior temporal gyrus is absent (*straight arrow*). Note that there is no displacement of suprasellar structures or ventricles.**

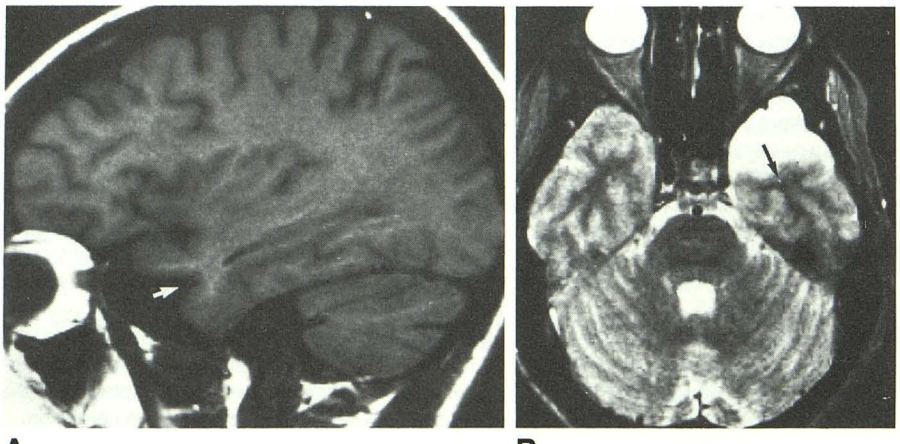

Fig. 2.—33-year-old woman with left middle cranial fossa arachnoid cyst.

***A*, Sagittal T1-weighted image (650/20) demonstrates hypogenesis of left middle temporal gyrus (*arrow*).**

***B*, Axial T2-weighted image (3000/90). Margin of temporal lobe adjacent to middle cranial fossa arachnoid cyst is undulating (*arrow*) and there is no expansion of middle cranial fossa. Middle cerebral artery (not shown) was not displaced. Coronal orientation of white matter in left temporal lobe is considered to be developmental and not due to expansion of adjacent cyst.**

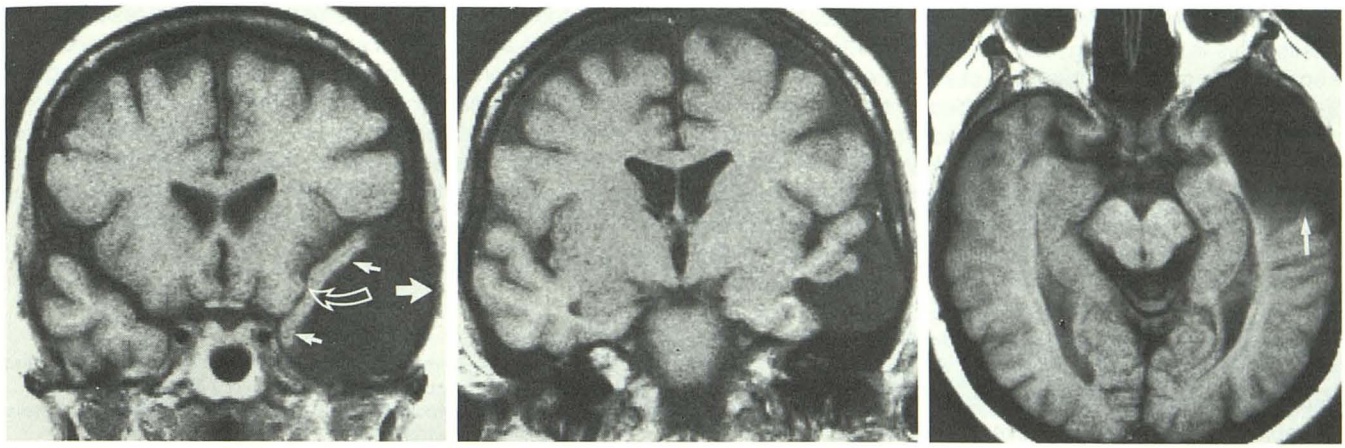

Fig. 3.—51-year-old woman with large left middle cranial fossa cyst.
***A*, Coronal T1-weighted image (650/20). Squamous temporal bone is bulging (*large straight arrow*). Sylvian fissure (*open arrow*) is medially displaced, as is part of temporal lobe (*small arrows*).**
***B*, Coronal T1-weighted image (650/20). More posteriorly, lateral margin of temporal lobe is undulating.**
***C*, Axial T1-weighted image (650/20). Note absence of anterior temporal lobe (*arrow*) with slight medial displacement of tip of temporal horn.**

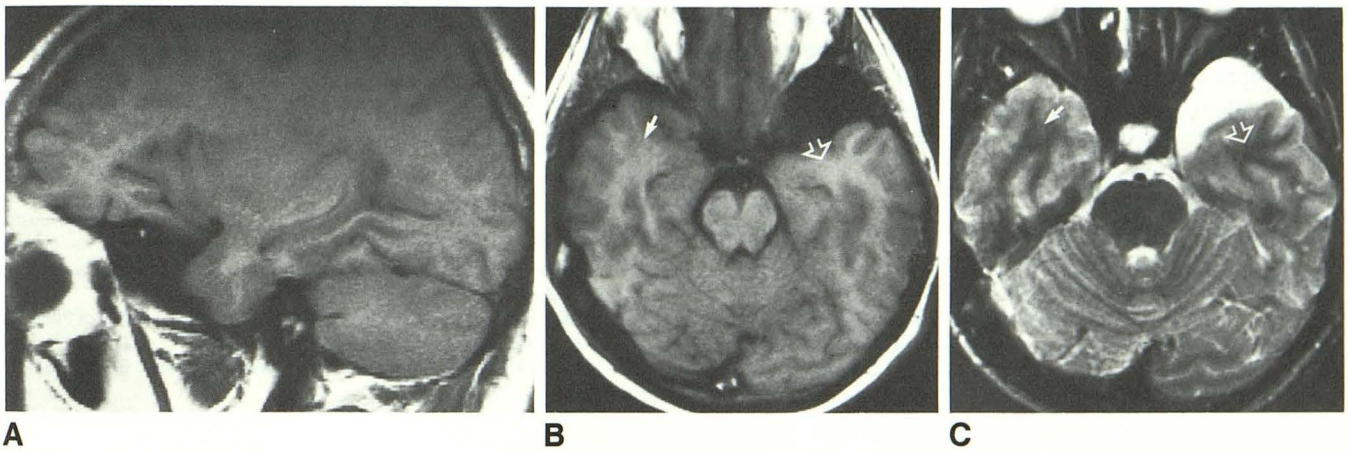

Fig. 4.—16-year-old boy with left middle cranial fossa cyst.
***A*, Sagittal T1-weighted image (650/20). Note absence of part of superior temporal gyrus of left temporal lobe, which has relaxed, undulating margins.**
***B*, T1-weighted image (650/20).**
***C*, T2-weighted image (3000/90).**
Fig. 4B is 1 cm craniad to Fig. 4C. As in Fig. 3B, white matter in left temporal lobe is coronally oriented as compared with that in right temporal lobe (*arrows*). Again, this is considered to be developmental in origin. However, slight posterior displacement of left temporal horn indicates a minor degree of associated compression.

forming pia-arachnoid during the earliest stages of embryogenesis could lead to formation of the cyst [10].

The less common type of arachnoid cyst is an intraarachnoid cyst—a concept introduced by Starkman et al. [11], who considered that some cysts are formed by splitting and duplication of the arachnoid membrane.

Robinson [12, 13] originally proposed that middle cranial fossa cysts were related to temporal lobe agenesis and that the adjacent CSF spaces represented a passive collection. In a later review, Robinson [14] reconsidered his earlier opinion and stated that arachnoid cysts are related to primary embryological malformation of the meninges and that the underlying temporal lobe is merely compressed. Angiographic studies of patients with middle cranial fossa cysts may often show venous anomalies [3]. The rate of occurrence with which the superficial middle cerebral vein drains into the cavernous sinus is decreased and that of nonvisualization of the vein is increased when angiographic studies of patients with middle cranial fossa cysts are compared with normal controls [15]. According to Giudicelli et al. [15], these angiographic changes date the malformation to between the sixth and eighth week of intrauterine life.

Our analysis of middle cranial fossa arachnoid cysts demonstrated only hypogenesis of the ipsilateral temporal lobe without compression in five of the cases and both hypogenesis and compression in three cases. The cortex of the

hypogenetic temporal lobes was of normal thickness in all cases. This is in keeping with Robinson's [13] autopsy observations in one of his reported cases. The cysts were not as large as some of those described in the literature, and therefore thickening of the skull, which may be expected in cases of hypogenesis without cyst expansion, was not seen in any case. That the two entities, the arachnoid cyst and the temporal lobe agenesis, are present together does not necessarily reflect a causal relationship. The expansion of the intercellular space of the meninx primitiva with clearing out of the cellular elements of the meninx occurs between fetal ages 6.5 and 8 weeks [9]. The neocortex is present by the eighth fetal week with the waves of ventriculocortical cellular migration occuring between fetal weeks 7 and 15 [16]. It is conceivable, therefore, that whatever fetal insult occurs embryologically to effect development of the arachnoid membrane, a similar event could effect development of the temporal lobe. In some patients compressive effects on the temporal lobe would result from cyst expansion.

Several authors have addressed mechanisms by which these cysts may enlarge and thus create symptoms. The common theories include diffusion of fluid into the cyst due to osmotic gradients between the cyst and adjacent subarachnoid space, a ball valve mechanism in which fluid can enter but not exit the cyst, and active cyst wall secretion by ependymallike cells lining the inner cyst wall. Go et al. [17] and Rengachary and Watanabe [18] discussed cyst ultrastructure and enzyme ultracytochemical evidence to support the theory that the cyst walls secrete fluid, since the cells lining the cyst contain structural features of fluid secretion and have enzymatic properties of fluid transport. Kumagai et al. [19] favor the ball valve mechanism as the most likely explanation but cannot exclude the osmotic gradient theory. In the series reported by these authors there was no histologic evidence of secretory lining within the cyst wall. Williams and Guthkelch [20] stated that the fluid pulsation of the cyst is an important factor in cyst expansion.

Handa et al. [21] and Wolpert and Scott [22] discussed metrizamide CT findings in patients with arachnoid cysts and described two patterns. The first is free flow into a CSF space, which may signify no true cyst but a passive CSF space. The second pattern is delayed filling of the cyst, confirming a separate space that is not filled by free communication. In cases with massive cysts, Handa et al. [21] suggest that the cyst may be large enough to exclude contrast material from the subarachnoid space. Sato et al. [3] reviewed 18 cases with either ^{111}In-DTPA or metrizamide CT cisternography and demonstrated delayed, progressive concentration of these agents within the cysts, confirming a separate structure. These data have surgical implications, as direct cyst shunting will be necessary if a separate expanding cyst is to be effectively managed [3, 21]. Some physicians [6] have advocated surgical treatment for the majority of lesions; however, others advise surgery only when there is evidence of an expansile lesion, since a cyst that is nonexpansile suggests that it is in equilibrium or communicates with the subarachnoid space in some fashion [7]. We would add that if the MR study demonstrates hypogenesis of the temporal lobe without compression, or if the compression is mild and not causing elevated intracranial pressure, surgery is not indicated.

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