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Craniocervical Neuroepithelial Cyst (Colloid Cyst)

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Colloid cysts represent between 0.25–1% of intracranial tumors [1–3]. Although most frequently located in the third ventricle, they are also found throughout the ventricular system [4]. Extraventricular location is rare [5]. We describe a case of a neuroepithelial cyst located in the craniocervical region, which was examined by CT and vertebral angiography, with subsequent biochemical study of the cyst contents and histologic confirmation.

Case Report

The patient was a 14-year-old girl with hypoacusia of the right ear and right-sided headaches, which had intensified over the 2 preceding months, accompanied by nausea and vomiting. Physical examination revealed right facial paralysis, horizontal bilateral nystagmus, absent nausea reflex, hypotonia, right hyporeflexia, and bilateral papilledema.

Plain skull and cervical spine radiographs were normal. CT scan showed a round, well-defined high-density area (97 HU) extending from C1 through the foramen magnum and into the posterior fossa. The right cerebellopontine angle was occupied by the mass that passed in front of the cerebral trunk, displacing the fourth ventricle to the left. Active hydrocephalus was present. There was no contrast enhancement (Fig. 1). Vertebral angiography revealed an expansive avascular lesion. Resection of the posterior arc of the atlas was performed together with hemirrhinotomy of the posterior fossa, during which a cystic lesion covered by a fine membrane and containing rapidly coagulating brownish material was removed. Microscopic study of the cyst wall revealed that this was formed by columnar epithelium on the interior wall and an external layer of lax connective tissue (Fig. 2). The epithelium was flattened and tense in places, and the acellular content appeared densely proteinaceous. Biochemical analysis of the contents revealed highly viscous material: one gram of material contained 910 mg of protein. Electrophoresis showed that 64% corresponded to albumin while the remaining 36% was globulin (26% $\alpha_1 + \alpha_2 + \beta$ and 10% gammaglobulin).

Discussion

Colloid cysts are most frequently found in the anterior half of the third ventricle [6, 7], although they may also be found in the fourth and lateral ventricles [8]. Location outside the ventricular system, in the leptomeninges of the quadrigeminal

lamina, was described by Hamby and Gardner [9]. Challa and Markesbery [5] reported a case of a neuroepithelial cyst in a 14-year-old boy who died from tetra-ethyl lead poisoning. Autopsy revealed a 2-cm cyst in the left paravermal region of the cerebellum.

Although our case is similar to that described by Challa and Markesbery, we believe it to be exceptional due to the symptomatology and CT appearance. It is well known that on CT colloid cysts produce high attenuation in 80% of cases [10, 11]. However, the fact that this lesion was located in the posterior fossa and high cervical region outside the ventricular system led us away from a diagnosis of neuroepithelial cyst and to that of dense dermoid or epidermoid cyst (the density of which ranges from 68–132 HU) [12, 13]. The nonexistence of dermoid sinus led us to the diagnosis of an epidermoid cyst.

Different theories attempt to explain the high attenuation of colloid cysts on CT. Isherwood et al. [14], carrying out biochemical studies of cyst contents, found that 88% was water and that there was no element of high atomic number. These authors state that the high density is due to the high concentration of electrolytes. Ganti et al. [7] believe the high density is due to desquamative secretory products from the cyst wall, hemosiderin, and possibly microscopic calcified foci. In our case, biochemical study failed to show any element of high atomic number, and we believe the high content of protein material is responsible for the high attenuation. Angiography showed only a nonspecific avascular mass and did not contribute to diagnosis.

Shuangshoti and Netsky [8] believe that neuroepithelial cysts can originate at any level of the ventricular system and that they may be formed from a fold in the neuroepithelium either inside or outside the ventricular system. In the third ventricle, colloid cysts may originate from the paraphysis [15]. Palma [16] proposes that neuroepithelial cysts form from primary ependymal ectopia. Popoff and Feigin [17] call attention to the ependyma-lined channels in heterotopic glial tissue in the subarachnoid space. Challa and Markesbery [5], in their case description, suggest that the cyst arises from an ependymal recess at the rhombencephalic isthmus.

In conclusion, we believe that when faced with an extra-

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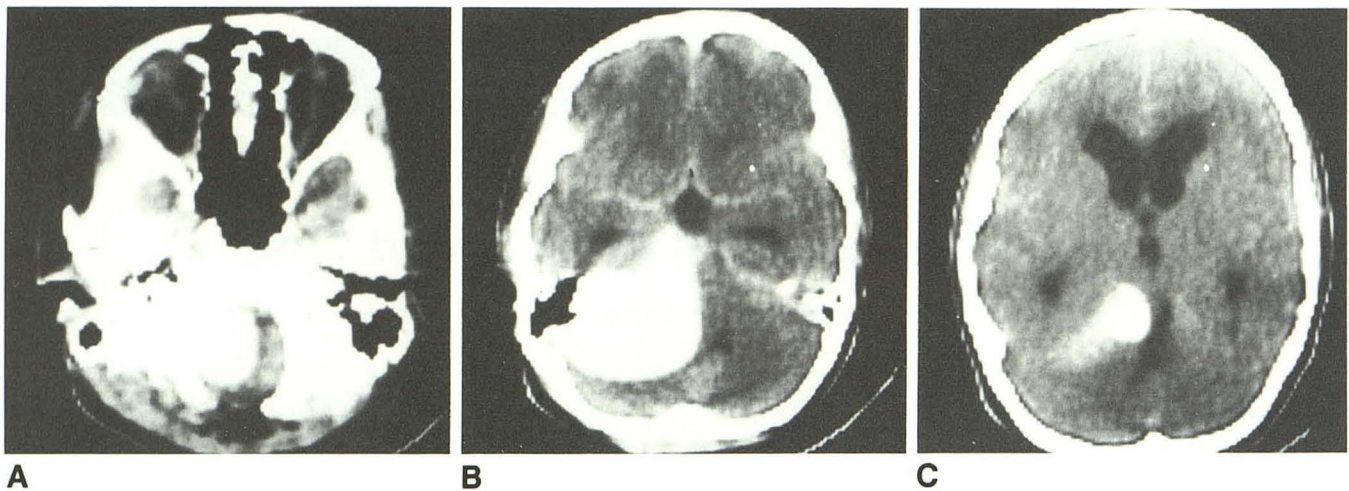


Fig. 1.—A, High-density rounded lesion at level of foramen magnum.
B, High-density lesion occupying the right cerebellopontine angle, displacing 4th ventricle. No contrast enhancement.
C, Higher slice, showing upper portion of lesion and ventricular dilatation.

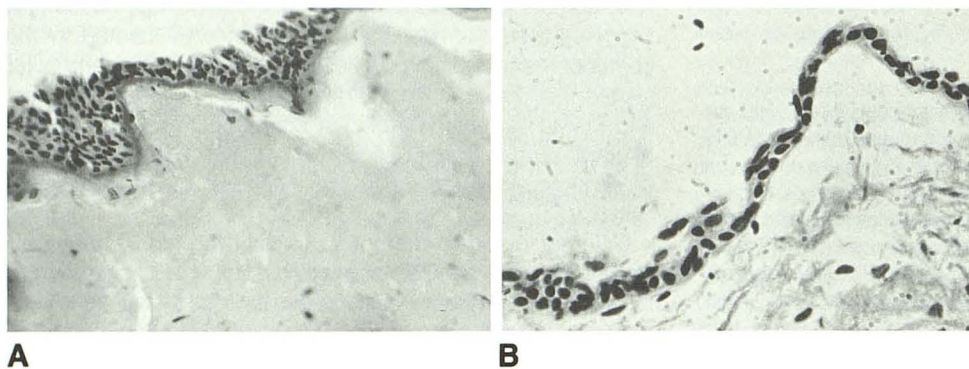


Fig. 2.—A, Columnar epithelium lining the cyst wall. Subjacent amorphous acellular contents (H and E $\times 250$).
B, Cuboid epithelium flattened on a delicate fibrous capsule in another area of the same specimen (H and E $\times 400$).

ventricular hyperdense lesion with no contrast enhancement, colloid cysts should be included in the differential diagnosis.

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