Xanthogranuloma of the third ventricle

Case report

IVAR SZPER, M.D., SHIZUO OI, M.D., JAN LEESTMA, M.D., KWAN SOO KIM, M.D., AND NICHOLAS E. WETZEL, M.D., PH.D.

Division of Neurological Surgery, Department of Neuropathology, and Department of Neuroradiology, Northwestern University, Chicago, Illinois

The case history of a patient with a xanthogranuloma of the third ventricle is presented. This type of lesion formed by desquamation of epithelium is usually noted as an incidental autopsy finding in the choroid plexus of the lateral ventricles. Total removal via a transcortical transventricular route led to complete recovery. These tumors should be suspected in the geriatric population. The computerized tomography findings and a review of the literature are included.

KEY WORDS • xanthoma • xanthogranuloma • intraventricular tumor • third ventricle tumor

Lesions that are localized completely within the third ventricle are rare entities, accounting for only 6% of all cerebral tumors. Clinically, they give rise to a ventricular syndrome and their neuroradiological diagnosis is difficult. Pathologically, most lesions of the third ventricle are neoplastic, or have a hamartomatous or developmental character. We are presenting a case that illustrates the diagnosis and treatment of one such tumor which was not a neoplasm, but rather a mass lesion probably produced by degeneration or an aging phenomenon.

Case Report

This 57-year-old man had been in good health until the spring of 1978. He presented to his neurologist in May, 1978, complaining of episodes of dizziness and tremor. Bradyphrenia, retropulsion, axial rigidity, and cogwheeling were noted. A diagnosis of Parkinsonism was made. He improved following treatment with Sinemet-10/100 (10 mg carbidopa and 100 mg levodopa) three times a day, and Antivert (meclizine hydrochloride), 25 mg, three times a day. He continued with his office work until 2 weeks before his admission, when his friends noted him to be unshaven, generally unkempt, forgetful, and stumbling. He was brought to the Veterans Administration Lakeside Hospital for a complete work-up in October, 1978.

Examination. General examination was negative; however, he admitted to an intake of one-third of a bottle of Scotch per day. Neurologically he was oriented to person and place, but calculated poorly, and could not make change or remember three objects after 15 minutes. Affect was flat and his intellect appeared to be much lower than demanded by his job. His deep-tendon reflexes were 2+ and equal, with bilateral Babinski signs. Positive suck, snout, and grasp responses were also elicited. No other abnormal neurological findings were noted.

A computerized tomogram (CT) was obtained before and after the infusion of contrast material. A round area of high density was demonstrated within the anterior and superior aspect of the third ventricle near the foramen of Monro (Fig. 1). The attenuation value of this area was slightly higher than that of brain parenchyma. There was faint calcification in a small portion of the rim of the lesion. The lateral ventricles were markedly dilated secondary to obstruction at the foramen. The postinfusion scan demonstrated the same lesion, which measured 2.5 cm in diameter, with ring-like contrast enhancement at the rim of the lesion. Angiography revealed only slight humping of the internal cerebral vein in association with a largely dilated ventricular system.

Operation. On November 6, 1978, a right frontal craniotomy was performed under the operating...
FIG. 1. Computerized tomography scan showing a xanthogranuloma of the third ventricle, without infusion (upper) and with infusion (lower). Note calcification in the preinfusion scan (arrow). Ventricular enlargement is also noted.

 FIG. 2. Low-power photomicrograph illustrating the variegated appearance of the lesion with many cholesterol clefts visible amid fibrotic inflammatory reaction. H & E, × 30.

microscope. After the dura was reflected, a core of cerebrum, 5 to 6 cm in diameter, was removed 2.5 cm from the midline, at the level of the coronal suture, down to the ventricle. The lateral ventricles were of generous size and the third ventricle was noted to be slightly dilated with a greenish, oblong, pedunculated mass attached to the fornix above. The septum pellucidum was noted to be perforated. Since a colloid cyst was suspected, multiple needle punctures were made, but no fluid could be aspirated. The tumor was incised after its surface had been coagulated, and a Love-Guenwald forceps was introduced into the tumor capsule with escape of thick, yellow, creamy, necrotic material. The tumor mass was almost completely removed except for a small portion attached to the choroid plexus posterosuperior to the fornix.

Histological Examination. The tissue mass contained dense fibrous connective tissue admixed with a chronic inflammatory infiltrate, foamy histocytes (xanthoma cells), siderophages, occasional foreign-body giant cells, calcific necrotic debris, and numerous cholesterol clefts (Fig. 2). In several portions, strips or small clumps of cuboidal epithelial cells were encountered about what may have been a cystic cavity. These epithelial cells contained prominent cilia at their surface. No squamous debris, colloid material, or other epithelial structures were observed.

Postoperative Course. The patient was disoriented and very talkative. Serum sodium was in the range of 118 to 124 mEq/liter with the most strict fluid restriction, while urinary sodium was 75 to 180 mEq/liter. Inappropriate antidiuretic syndrome was diagnosed and treated with declomycin, 600 mg/day, which was eventually increased to 1200 mg/day. Serum sodium levels returned to the normal range approximately 2 weeks postoperatively and the patient became more lucid and alert. After approximately 1 month he was thinking clearly. At the time of this writing he has returned to his former employment with a security firm.

Discussion

The pathological diagnosis was xanthomatous (xanthogranulomatous) foreign-body reaction of the choroid plexus. This entity has been noted by
Xanthogranuloma of the third ventricle

others, 1-5, 7-10, 12-17, 19 although different terms have been used, such as "xanthoma," 19 "cholesterol granuloma," 21 "cholesteatomes des plexus choroides," 7 and "xanthogranuloma." 14 This entity is generally found incidentally at autopsy, but is not the cause of death. By 1960, 102 cases found at autopsy had been reported in the literature. 1, 19

Blumer 2 first reported xanthogranuloma in 1900. Autopsy series have stated that the incidence of these lesions varies between 1.6% and 7%, 1, 19 most frequently in subjects in the seventh decade. There appears to be no relationship between these lesions and the sex or race of the individual, and they are seemingly not related to system diseases such as hypercholesterolemia, atherosclerosis, or diabetes. 5, 19

All autopsy specimens described to date have been found either unilaterally or bilaterally in the trigone of the lateral ventricles, 1, 2, 19 ranging in size from 2 mm to 12 cm. 1, 19, 19

The etiology of these intraventricular masses is debatable. The most recent explanation 16 postulates that the epithelium of the choroid plexus is in constant proliferation and desquamates epithelial cells into the interstitium at sites of naturally occurring disrupted basal laminae. Intracellular lipids increase as these epithelial cells degenerate, and the appearance of the cells becomes quite similar and practically indistinguishable from lipid-laden macrophages. The subepithelial portion of the choroid plexus becomes filled by aggregates of large, pale, foam cells with eccentric nuclei, among which can be seen empty clefts produced by extracellular cholesterol crystals. 13 Further disintegration of the epithelial cells releases lipid material into the stroma of the plexus. This incites a foreign-body reaction attended by proliferation and fusion of macrophages forming multinucleated giant cells. Changes in local circulation may lead to microscopic hemorrhages and the accumulation of siderophages.

Five clinical presentations of these lesions have been noted previously in the literature. 6, 9, 12, 10, 19 In three of these reports the lesion was located in the lateral ventricles, 6, 12, 19 and in two in the third ventricle. 6, 19 These lesions were all found in children under the age of 18 years, except for the case reported by Shuangshoti, et al., 19 which was a combined neuroepithelial cyst and xanthoma occurring in a 26-year-old woman.

Our case is the first clinical presentation of a xanthogranuloma of the choroid plexus in an older patient. It should also be noted that this is the first instance in which such a lesion in the third ventricle has been revealed on CT scanning. Terao, et al., 19 described a lesion with bilateral calcifications in the trigone shown on CT; after injection with 100 ml of 30% Conray, the scan showed marginal irregular enhancement. The CT scan in our patient showed a round area of high density which demonstrated enhancement in the margin; it also showed faint calcification in a small portion of the margin. The lesion could not be differentiated from a colloid cyst. The presence of calcification in this entity as seen in these two cases may be an important differential point, because calcification in a colloid cyst is rare.

Conclusions

Xanthogranulomas of the choroid plexus are benign, non-neoplastic lesions. If situated in the third ventricle, where xanthogranulomas can reach a clinically significant size, hydrocephalus can cause the appearance of a midline syndrome. Clinical differentiation from any other third ventricular lesion is difficult. Computerized tomography will demonstrate a regularly rounded mass with capsular enhancement and calcification which should be differentiated from a colloid cyst. Complete removal by a transventricular route is uncomplicated.

This benign lesion is found in a high incidence of autopsy series in the elderly and is thought to result from a normal aging process. It should be considered in evaluating elderly patients.

References


Address reprint requests to: Ivar Szper, M.D., 5415 N. Sheridan Road, Apt. 4706, Chicago, Illinois 60640.