Hydrocephalus due to xanthogranuloma

Case report

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Xanthogranulomas and cholesterol granulomas of the choroid plexus in the lateral ventricles are usually incidental findings at autopsies. This report presents a case with hydrocephalus due to a xanthogranuloma that obstructed the foramina of Monro and was successfully treated by surgical intervention.

KEY WORDS  xanthogranuloma  cholesterol granuloma
- hydrocephalus - intraventricular tumor

Xanthogranulomas of the lateral ventricles are occasionally found as incidental findings at autopsies.1-4,11,14 The case presented here exhibited clinical symptoms and signs and suggests that these lesions can expand, causing obstruction of the foramen of Monro.

Case Report

This 18-year-old woman started having headaches, nausea, and vomiting 3 months before she was admitted to the hospital following an acute exacerbation of these symptoms.

Examination. The patient was mentally alert but unable to stand or walk unsupported. She had bilateral papilledema. No other neurological signs were found. Plain skull films and brain scan (99Tc pertechnetate) were both normal. Bilateral carotid and vertebral angiographic studies suggested supratentorial hydrocephalus. During ventriculography through a right-sided frontal burr hole, the Silastic catheter diverted unexpectedly into the left frontal horn. The air filled an enlarged left lateral ventricle, but no air passed to the third or to the right lateral ventricles. Dimer-X* (3 ml) was then injected through the Silastic tube and outlined an expanding lesion in the left lateral ventricle, near the foramen of Monro. No contrast medium passed to the right lateral ventricle, and only traces to the third.

Operation. A right-sided frontoparietal transcortical approach was made to the lateral ventricle. The smooth surface of a tumor in the third ventricle was seen through the enlarged right foramen of Monro. Upon surgical fenestration of the septum pellucidum, a plum-sized tumor was seen rising from the left foramen of Monro into the left lateral ventricle, so that half of

*Dimer-X is made by Laboratoires Andre-Guerbet, 16-24 rue Jean-Chaptal, F 93 Aulnay/Bois, France.
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Postoperative Course. On the third postoperative day the patient developed signs of increased intracranial pressure and meningitis. External drainage of the right lateral ventricle was established and maintained for 6 days, and antibiotics were given. The symptoms and signs gradually subsided, and the further course was uneventful. On follow-up examination 5 months later she had no complaints, no neurological deficits, and had resumed her previous occupation.

Histological Examination. The specimen had a fibrous cystic wall; the contents were amorphous, eosinophilic-staining, and included red cells, fibrin, and numerous large, partly necrotic foam cells. In addition there was fibrous granulation tissue with cholesterol crystal spicules surrounded by foreign body macrophages (Fig. 1). Only a few of the choroid plexus cells were preserved. The histological structure was typical of a cholesterol granuloma in the choroid plexus.

Discussion

The first report on xanthogranuloma of the choroid plexus in man was made by Blumer in 1900 under the name “cholesteatomatous endothelioma.” He showed that this was the same lesion as the “plexus-cholesteatoma,” which for some time had been known to occur in horses.

Later reports have used a variety of names for this lesion: “cholesterinhaltige Geschwulste,” “xanthoma,” “cholesteatome des plexus choroides,” “cholesterinose des plexus choroides,” “cholesterol granuloma,” and “xanthogranuloma.” Shuangshoti and Netsky suggested that the latter term be used, because they felt that xanthomas and cholesterol granulomas simply were different stages of the development of the same fundamental process.

Most of these reports discuss incidental findings at autopsies, and neurological symptoms or signs due to the presence of xanthogranulomas are not mentioned. The xanthogranuloma is not included in the extensive list of intraventricular tumors recently recorded by Pecker, et al. Manlove...
Hydrocephalus due to xanthogranuloma

and McLean\(^7\) tried to find a connection between small intraventricular calcifications seen on skull films in some patients with headache and xanthogranulomas, which occasionally calcify. However, they did not refer to any particular case in which clinical symptoms could be definitely related to the calcification. Wolf, \textit{et al.},\(^14\) found xanthogranulomas in the remarkable high frequency of 1.6% in 1181 consecutive routine autopsies. Even higher frequencies have been reported from other autopsy series.\(^1,13,14\) It is therefore somewhat surprising that clinical symptoms never seem to have been convincingly related to this lesion.\(^1,14\) The present case, however, shows beyond doubt that the xanthogranuloma can cause symptoms and signs in man, and that surgical treatment is possible.

There have been many theories as to the origin of xanthoma cells.\(^1,2,4,6,18,14\) Shuangshoti and Netsky\(^11\) believe they are derived from choroidal epithelial cells that normally contain some lipid material and are known to have phagocytic properties.\(^9,10\) The high cholesterol content of the lesion indicates that an active storage mechanism is involved.\(^5\) Xanthogranulomas are unrelated to age, sex, or race,\(^1\) or to such systemic diseases as hypercholesterolemia, atherosclerosis, and diabetes mellitus.\(^1,14\)

The histological picture is probably due to a necrosis of foam cells with release of lipids into the stroma, which gives rise to the formation of cholesterol crystals and granulomatous reaction with foreign body giant cells and occasionally to calcium deposits.\(^1,14\) Disturbance of the local circulation may add hemorrhage and cyst formation to the original lesion.\(^1,11\) Most of the reported xanthogranulomas have been attached to the choroidal plexus of the lateral ventricles, mainly to the glomus choroideum, although they have occasionally been found in the third ventricle.\(^12\) According to Shuangshoti and Netsky,\(^9,11\) the histogenesis of the choroid plexus offers an explanation for this phenomenon.

From the surgical point of view, it should be stressed that xanthogranulomas are circumscribed, non-neoplastic expanding lesions which are technically easy to remove in spite of their location.

References


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