JIHS The Journal of Integrated Health Sciences

Available online at www.jihs.in

Case Report

Xanthogranuloma of Choroid plexus:Case report and Review

Bhalodiya DH*[1], Lakhani JD[2], Shah S[1], Kumar S[3], Pathak KJ[2]

[1]Resident, [2]Professor, [3]Assistant Professor; Department of Medicine, Smt. B. K. Shah Medical Institute and Research Center, Sumandeep Vidyapeeth, Piparia, Waghodia, Vadodara, Gujarat, India

ABSTRACT

Xanthogranuloma (XG), a condition of obscure pathogenesis and varied terminology, can occur in choroid plexus. We present here xanthogranuloma of the choroid plexus of lateral ventricle, who presented with headache, mild cognitive impairment and hemiplegia. It was difficult to determine whether bilateral small xanthogranuloma found on MRI was responsible for patient's presenting symptoms or not. Appraisal of clinical and pathological findings related to intracranial and intraventricular choroid plexus XG, is done from the available literature. Furthermore, review in topic of "relation of XG with arthrosclerosis" as well as "Dyslipidemia" is also done.

Key Words: Xanthogranuloma (XG), juvenile Xanthogranuloma (JXG), choroid plexus, foamy cells, Hemiplegia, Symptomatic Xanthogranuloma

INTRODUCTION

Xanthogranuloma (XG) occurs as tumefactions/benign tumors intracranially especially in the choroid plexus. This condition bears various names and reported as xanthoma, cholesterol granuloma, cholesteatoma xanthosis and cholesterinosis[1]. Blumer in 1900, (cited by Shuangshoti S, Netsky MG) reported first case of xanthogranuloma as "cholesteatomatous endothelioma" of the choroid plexus.[1] Most reviews suggest that XG are benign tumors which are asymptomatic and accidentally detected. They are of uncertain etiology. They remain symptom free as they do not cause "mass effect". Such lesions can be in form of tiny spec or a very large mass which microscopically contains lipid/cholesterol containing cells. Thus it is also known as cholesterol-containing cell granuloma or tumor.[1] Various symptomatic cases are described in literature in relation to third ventricle, lateral ventricle and fourth ventricle. XG of third ventricle may present as hydrocephalus due to obstruction of the Foramen of Monro.[2] Similarly symptomatic XG involving the choroid plexus of the lateral ventricle and fourth ventricle are also described.[3,4] Such lesion could be found incidentally at autopsy in 1.6–7% of cases.[1,5-7] The small size lesions may not be visible on imaging.

CASE HISTORY

50 years old female patient residing at Badwani, Madhya Pradesh, who is a housewife belonging to lower socio-economic status presented with chief complaint of headache since two months and right upper and lower limb weakness since one month. Patient was complaining of headache since two months. Headache was initially of mild intensity, not hampering her routine activities. It

***Correspondence:**
E-mail: dharmesh16490@gmail.com

initially lasted for 20-30 minutes; mainly at occipital area and was on & off. Gradually intensity and duration of each episode increased to such an extent that patient had to take some oral medication to get relief. With period of time, headache became generalised; however, more at the occipital area and it almost lasted the whole day. Headache was not associated with nausea, vomiting, fever or any visual disturbances. One day, in the morning at 11 am when patient was cooking; she felt weakness in right lower limb such that patient was unable to stand without support. For these complaints, she took treatment from village's "Hadvaid"(Bone-doctor). Subsequently after 14-15 days, when patient woke up from the sleep she noticed weakness in right upper limb also. The weakness of right upper limb was initially more in proximal muscles that she felt difficulties in combing hair and lifting objects over head. Weakness was gradually progressive in nature, such that within the next 8-10 days, it involved distal group of muscles also, such that she could not grip objects. This weakness was not associated with deviation of mouth, diplopia, dysphagia, bowel bladder dysfunction or any sensory loss. Patient had no similar complaints in past and also no past history of hypertension, diabetes, tuberculosis, blood transfusion or recent vaccination. There was no significant family or personal history. On examination patient was conscious, co-operative and well oriented to time place and person. She was afebrile, pulse was 86 per minute in right radial artery and blood pressure was 110/74 mm Hg in right brachial artery in supine position. Mini-mental score of the patient was 24. In systemic examination of central nervous system, higher function and cranial nerve examination was normal. Clasp-knife spasticity was present in right upper limb and lower limb while in left upper and lower limb tone was normal. Power in the affected limbs was 2/5 while on the unaffected side it was 5/5. Superficial reflexes were normal. Deep tendon reflexes on right side were brisk and on left side normal. Plantar was extensor on right side and flexor on left side. Cerebellar and Autonomic functions, peripheral nerve, spine and cranium were normal. Rest all systemic examination was within normal limit.

Table 1: Laboratory findings

Haemoglobin (gm/dl)	10.6
Total White Blood Cell /cu mm	8100
Differential Count	83/10/3/4
Platelets /cu mm	2.57
Random Blood Sugar (mg/dl)	93
Blood Urea Nitrogen (mg/dl)	33
Serum Creatinine (mg/dl)	1.1
Total Bilirubin	0.7
Direct Bilirubin	0.3
Indirect Bilirubin	0.4
SGOT (I U)	12
SGPT(I U)	12
Serum Sodium (mmol/L)	137
Serum Potassium (mmol/L)	3.8
Serum Cholesterol (mg%)	254
Triglycerides (mg%)	168
HDL (mg%)	48
LDL (mg%)	116
VLDL (mg%)	43.6
HIV, HBsAg, HCV : Negative	
ECG: Normal sinus Rhythm	USG ABDOMEN & PELVIS: NAD
Bilateral Carotid/ Vertebral Doppler	Normal

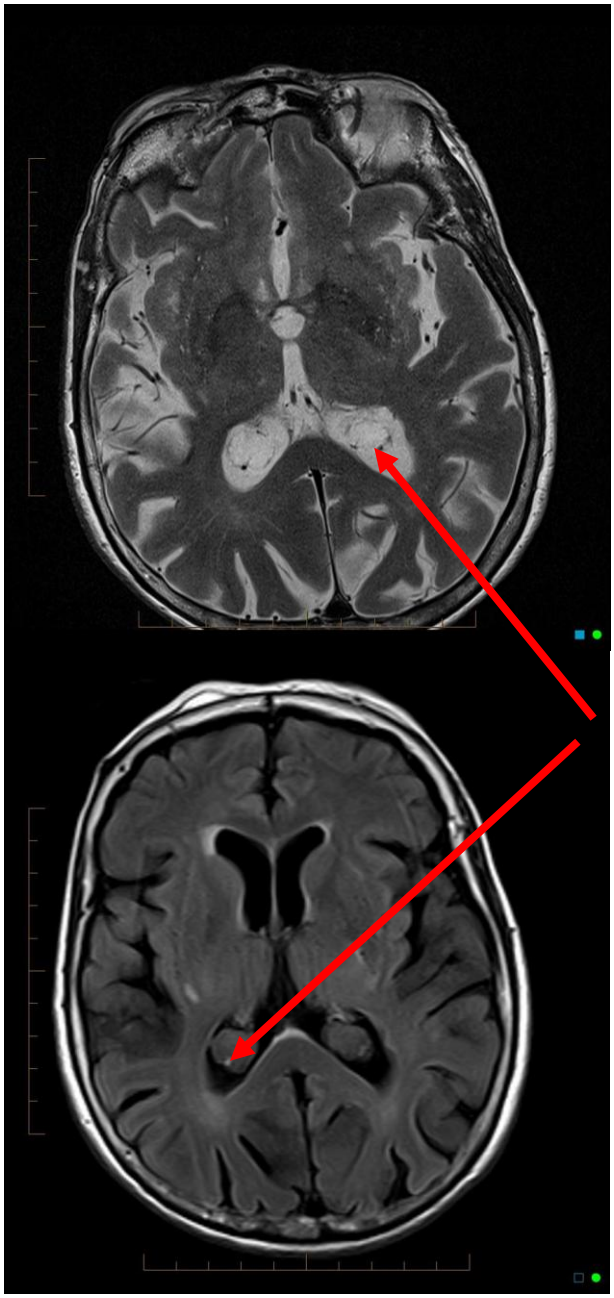

Figure 1: MRI Brain: Abnormal high signal intensity lesion noted involving bilateral choroid plexus of occipital horn of lateral ventricle on T2WI which appear hypotintense on T1W1, FLAIR and shows diffusion restriction on DW1 sequence. Size measuring approximately 1.6×1.1 cm on Right side and 1.2×1.2 cm on left side, possibility of xanthogranuloma of occipital horn of bilateral choroid plexus (Arrow).

DISCUSSION

Xanthogranuloma is common in choroid plexus of ventricles and has been reported in third ventricle, lateral ventricle as well as fourth ventricle.[8,9] Choroid plexus xanthogranuloma is also known as "plexi choroideorum xanthomas". Symptoms may arise if it cause obstruction of the ventricular system[6,9] or is complicated with haemorrhages. Patients may have headache due to hydrocephalus. They may present with confusion, short term amensia or gait disturbance. Symptoms may depend upon the site where tumor is located. Xanthogranulomas of the glomus of the lateral ventricles could be different from that of third ventricle cystic lesions which are like colloid cysts.[3]

In 1900, Blumer first reported case of xanthogranuloma of the choroid plexus; however in1973, work of Kepes et al gave evidence for presence of non-ventricular fibrous xanthoma.[10] Intracranial Xanthogranulomas is described in adults and children (juvenile Xanthogranuloma JXG) which could be solitary or multiple with or without skin involvement.[11,12] Cutaneous form can present like solitary or multiple yellowish, orange-red or tan-hued papules.[11] Multiple extensive intracranial lesions are described in cerebellum, brain stem, thalami, and bilateral cerebral hemispheres.[12]

Our patient presented with headache, mild cognitive impairment and hemiplegia; which partially recovered without any specific treatment. Headache may be due to obstruction of cerebral circulation. Hemiplegia is difficult to be explained on the basis of bilateral xanthogranuloma of choroid plexus situated in the occipital horns of lateral ventricles. Could this MRI finding of bilateral choroid plexus be incidental? This middle aged, post menupasual woman may have had vascular stroke leading to hemiplegia. However, imaging findings did not favour it. Her lipid profile was abnormal.

Hemiplegia in XG is described however it is described more in Juvenile xanthogranuloma (JXG) that also in extraventricular XG. Hemiparesis, hyperreflexia and hypoalgesia in neck, trunk and limbs due to intradural xanthogranuloma in the upper cervical spine was reported in an 18 year female by Oyama H et al.[13] Hemiperesis because of marked displacement and stenosis of the middle cerebral artery, having intracranial xanthogranuloma in the middle cranial fossa was reported by Koyama *et al.*[14] Review based on 35 symptomatic intraventricular xanthogranulomas of brain, Shuangshoti S., showed hemiplegia as one of the reported symptoms.[14] As per this review, patients may present with headache, nausea,vomiting. optic atrophy, papillodema, diplopia, nystagmus, ataxia, seizure, fifth and seventh cranial nerve involvement, sphinctor disturbance, distortion of sense of smell and taste, altered consciousness, personality changes psychomotor retardation, confusion and aphasia.[15]

Foamy cells fat-laden macrophages which are seen in XG. They are seen in atherosclerosis also in form of fatty deposit on the blood vessel walls. It may lead to plaque formation in atherosclerosis, which is risk factor for coronary artery disease and stroke.[16,17] Foamy macrophages are also found in infections due to Chlamydia, Toxoplasma or Mycobacterium tuberculosis. The cholesterol forms a rich food source for the bacteria. Caseation found in the center of tubercular granuloma is basically a mass of cholesterol.[18] Thus foamy cells are common in athrosclerosis, chronic infections and XG.

Origin of the foamy cells in XG is unclear. Investigators have found that there is dyslipidemia with xanthogranuloma of the choroid plexus. They have tried to find link between lipid metabolism and XG.[1] Wolf and co-workers (cited by Shuangshoti S, Netsky MG) reported 16 of 20 patients having mild to severe generalized arteriosclerosis. They postulated that hyperlipemia was one of the causative factors in the formation of xanthogranuloma.[1] Our middle aged patients did have dyslipedemia. We do suggest that lipid metabolism alteration may be playing role in formation of xanthogranulomatosis. Albeit, further research is required.

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