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Fatal outcomes in Third Ventricular Colloid Cyst: Analysis of Cases, Review of literature with emphasis on prompt intervention in Emergency Department to reduce case fatality rates.

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ABSTRACT:

Colloid cyst of the third ventricle is a rare benign lesion accounting for 0.5% of all brain tumors, a small subset of these patients can present with acute neurological deterioration which can be fatal. Estimate the risk of acute deterioration in patients and to assess the outcomes in patients with symptomatic colloid cysts. Retrospective correlative study of all patients diagnosed to have colloid cyst based on neuroimaging studies[CT and or MRI] at JSS Medical College and Hospital during the period January 1, 2014 to January 31, 2016. Of the 24 patients included in the study, 8 patients presented with acute neurological deterioration of whom, four died suddenly following diagnosis. Estimated risk of deterioration was 36%, but mortality in the subset of patients with acute neurological deterioration was as high as 50%. Risk factors for acute deterioration based on patient age, cyst size and density on plain CT, and presence of hydrocephalus were evaluated. Positive correlation was obtained for cyst size and hydrocephalous. While most patients have an insidious clinical course, a small unpredictable cohort of patients present with a cascade of neurological which can culminate in sudden death, necessitating early surgical intervention. Early intervention with ventriculostomy as an interval procedure in the setting of Emergency Department can often prove to be life saving.

Keywords: Colloid cyst, Sudden death, Ventriculostomy.
Abbreviations used in this paper: CT = computerized tomography; MRI = magnetic resonanceimaging; Colloid Cyst [CC]

INTRODUCTION

Third ventricular colloid cysts are rare benign epithelial cysts which account for 0.5-2% of all the intracranial tumours with a reported incidence is 3.2 per 100,000 populations. Clinical presentation with symptoms is during mid-adulthood.[1,2,7] Despite their benign histology, they carry a poor prognosis by virtue of its location with a documented mortality greater than 10% in symptomatic cases. The risk of fatality is reported to be as high as 1/3rdof all symptomatic patients.

Embryologically, these cysts are derived from the primitive neuroepithelium of the tela choroidea or the ectopic endodermal migration in the velum interpositum. The cysts are located in the rostral aspect of third ventricular roof. Subsequently they project inferiorly to occupy the anterosuperior quadrant adjacent to the foramina of Monro.[2,3]The cyst is attached by a short stem to the wall of the third ventricle; it is this attachment that provides partial stability within the ventricular lumen.[4,5 6]

The natural history of colloid cysts is varied but is mostly an insidious course. Majority are a symptomatic and are often discovered as an incidental finding. Symptomatology at presentation is varied; the commonest initial symptom being headache in about 2/3rd's of the patients. Episodes of paroxysms of headache (with change of head position), vomiting and amblyopia, or profound mental changes are classical of colloid cysts. Other symptoms include disturbed mentation, vomiting, vertigo, drop attacks and sudden attacks of leg weakness, rarely can it manifest with seizures. [7] Recent studies also highlight the prevalence of cognitive symptoms, with symptoms ranging fromanterograde amnesia to gustatory hallucination. These symptoms can occur with or without the presence of

hydrocephalus, and are thought to be secondary to compression of connecting pathways between the mesocortices and subcortical limbic regions.[8]

The ominous facet of this benign cyst is the occurrence of acute neurological deterioration precipitated by sudden death. Proposed mechanism for this lethal outcome is a matter of debate. This has been attributed to acute obstruction of cerebrospinal fluid (CSF) resulting in herniation and brainstem compression or alteration of the hypothalamus-mediated cardiovascular reflex control.[10] Atleast one-third of symptomatic patients are at risk for precipitous decline or death.[11,12]

The main stay of therapy for symptomatic colloid cyst, with or without neurological deterioration issurgery. Varied approaches have been advocated for the cyst excision. Trans-callosal, trans-cortical, trans-ventricular or a sub-frontal lamina terminalis approach have been described. In areas where-in neurosurgical expertise is limited, prompt unilateral or bilateral ventriculostomy to reduced intracranial pressure has proven to be lifesaving in the setting of acute neurological deterioration if intervened early. Cyst excision can be done as an interval procedure following the shunt. [12]Despite our increasing awareness of the natural history of colloid cysts, the cohort of patients who are at risk for acute neurological deterioration cannot be accurately defined. Guidelines pertaining to surgery or follow-up in patients with asymptomatic colloid cyst are still unclear. [14,15]

MATERIALS AND METHODS

Sole inclusion criterion was neuroimaging documentation of a Colloid cyst during the period January1, 2014, and January 31, 2016. All imaging studies were performed on Philips Ingenuity-128slice CT and Philips Ingenia-3Tesla MRI at JSS Medical College and Hospital, Mysore.

24 patients with an imaging diagnosis of colloid cyst were retrieved from the archives and evaluated for outcomes. Two categories were distinguished, one cohort with acute neurological deterioration characterized by a cascade of neurological symptoms accompanied with vomiting, altered consciousness suggestive of raised intracranial pressures and the other in whom the diagnosis was incidental, the patients who underwent neuroimaging for other systemic illnesses.

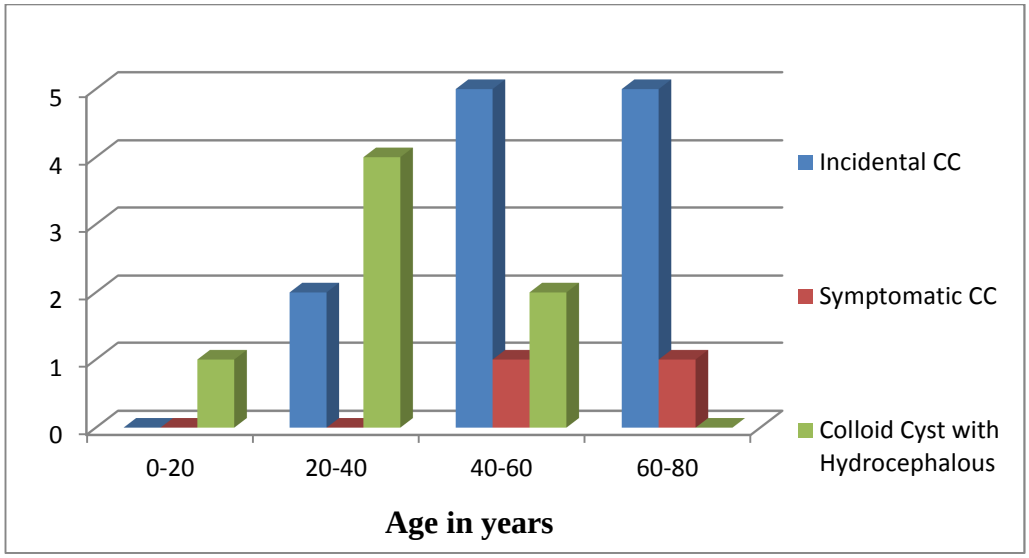
Neuroimaging was performed using CT in 18 patients (75%) and MRI in 6 patients (25%). Data on patient age at presentation, size and density of the cyst, presence of hydrocephalous were retrieved for statistical analysis. Each factor was assessed as an independent variable for the cause of deterioration. Risk factors for acute deterioration were explored using univariate analysis with the Fisher exact testand crude relative risk assessment. Computer statistical software (Software Package for the Social Sciences, version 10.0.5 for Windows; SPSS Inc.) was used to perform statistical analysis.

RESULTS

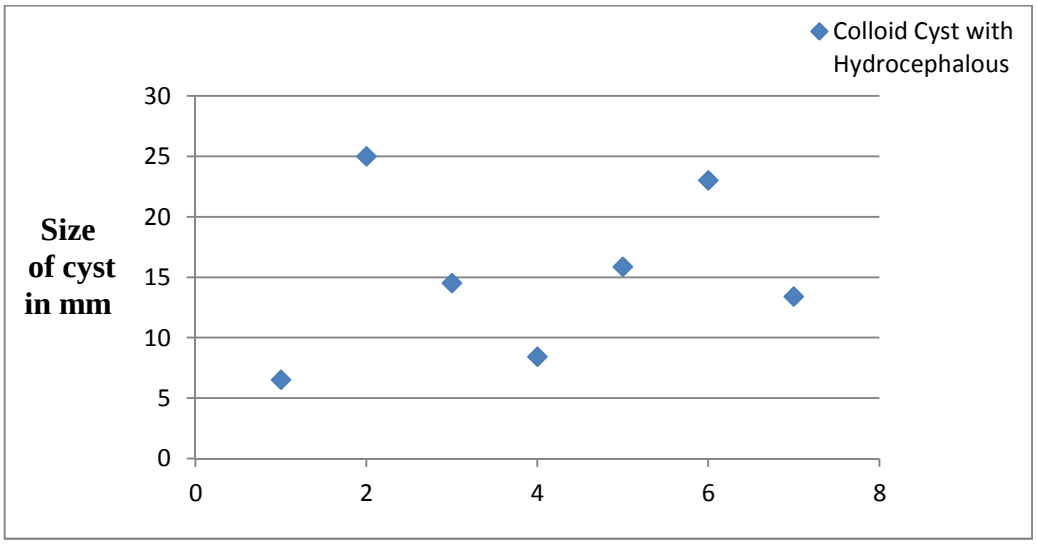
Of the 25 patients, 8 presented with acute neurological deterioration, 4 died and 4 were operated upon with good outcomes. The mean age of patients at the time of diagnosis was 46.9 years (range 21-77 years) with a definite male preponderance of 24%. Estimated risk of deterioration was 36%, but mortality in the subset of patients with acute neurological deterioration was 50%. The natural history of colloid cysts and the pathophysiology of acute deterioration are insufficiently known to calculate the risk for an individual patient based on these figures.

The age group at presentation was also varied the youngest in our series was 13year old, the oldest was 77years. Sizes of the cyst were varied, the smallest cyst was 3mm, and the largest was 25mm.[Graph 1, 2] Of the 8 symptomatic patients, the youngest was 13years and the oldest was 44yrs, both of whom died during MRI. The time lapse between CT and MRI was about 6.2hours. One patient died immediately after CT scan in emergency department (0.4hrs), one died in transit to operation theatre, both of these patients could not be resuscitated. 4 other patients were operated and discharged with no neurological complications. Multivariate statistical analysis of risk factors revealed positive correlation for the occurrence for hydrocephalus and size of cyst.

Graph 1: Age at presentation of colloid cysts



Graph 2: Scatter graph depicting the varied sizes of colloid cysts complicated by hydrocephalous.



The second cohort of patient in whom the diagnosis of colloid cyst was incidental, were educated about the cyst and its complications. Patients were advised to report to the emergency department in event of occurrence symptoms, follow up imaging was advised on a yearly basis for a period of three years, if clinically quiescent, they were advised clinical evaluation and cessation of sequential imaging.

DISCUSSION

Despite articles documenting the benignity of this lesion, review of literature documents the occurrence of acute deterioration and sudden deaths, a 1996 review of the literature by Hernesniemiand Leivo recorded a 21% of the 939 patients presented with compromised consciousness. [1] Even in our own case series, 8 patients (33.3%) presented with acute deterioration, of whom four died, of the four two patients succumbed during the MRI.

The true prevalence of colloid cysts is unknown, but based on a series of consecutive CT scans it has been estimated to be lower than one in several thousand, however the autopsy series document a prevalence of 1 in 8500 persons. Although the prevalence of colloid cysts is quite low, the incidence is even lower. [17]

In our series, we noted that the estimated risk of acute deterioration for incidentally discovered asymptomatic colloid cysts is only a fraction of the estimated risk for symptomatic patients. These results are in agreement with recent data on untreated asymptomatic patients given the natural history of these cysts.[14,15,22]A life time risk of acute

deterioration in patients harbouring symptomatic colloid cyst is varied, highest reported till date is about 34%; though this cohort of symptomatic patients may constitute a composite of subgroups with different risk variables, pertaining to age at presentation, presence of hydrocephalous and the cyst size & cyst density.

For instance, it has been suggested that colloid cysts smaller than 10 mm in diameter are associated with a minimal risk of acute deterioration.[16,17,18,19] In our study, incidental colloid cysts were mostly in the range of 3mm to 12mm, mostly less than9mm, only two patients were 10mm and other was 12mm, both of whom had no clinical symptoms pertaining to the colloid cysts. In the cohort of patients with presented with acute neurological deterioration, the size ranged from 6.5mm to 25mm [Figure 1, 2].

Figure 1: Axial, Coronal and Sagittal Non contrast CT of a 21year old lady who died soon after imaging, showing a hyperdense third ventricular Colloid Cyst complicated by hydrocephalous & brain herniation

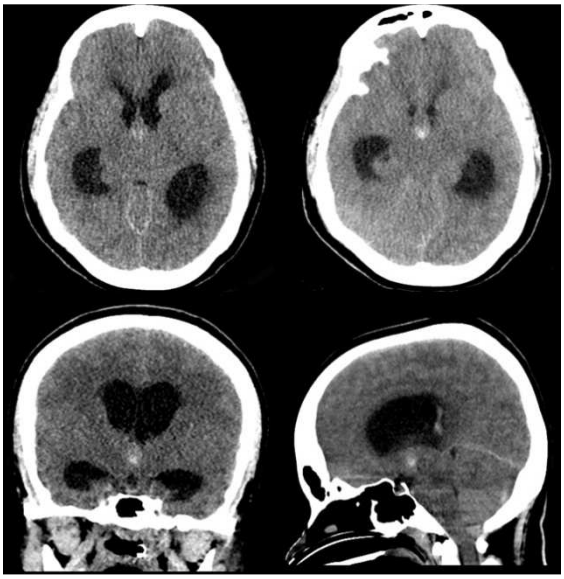
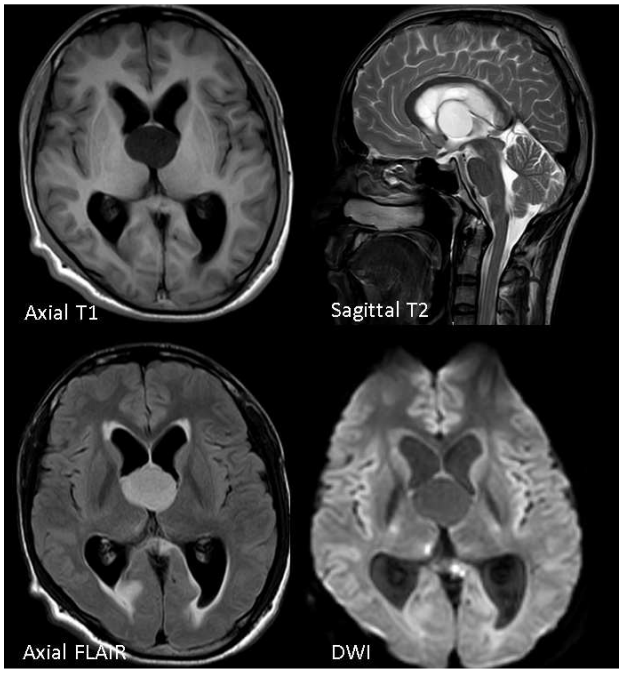


Figure 2: Multiplanar and Multisequence MRI of a 26year old male showing T1 hypointense and T2 hyperintense lesion with no restricted diffusion, third ventricular Colloid Cyst complicated by hydrocephalous & brain herniation. No enhancement was seen in the post contrast images, not depicted here



Pollock and colleagues proposed a theory of the natural history of colloid cysts. The occurrence of symptoms is thought to depend on the interaction of three factors: rate of cyst growth, development of cerebrospinal fluid

obstruction, and arrested cyst growth in the older patient. [14]The theory posed by Pollock and colleagues does not identify subgroups of patients with higher or lower risks of acute deterioration.

With increased availability of neuroimaging, asymptomatic colloid cysts will be encountered more frequently. The dilemma for therapeutic approach is in assessment of headache as the presenting symptom concerns whether the headache is caused by the colloid cyst. Pollock et al, in their review opined that if the headache was thought to be unrelated to the colloid cyst, the patient is categorized as Class I or II; neurosurgical intervention is not warranted in these patients. If however headache is associated with other symptoms, then such patients would be deemed as symptomatic (Class III) and prompt neurosurgical intervention is resorted to in this subset of patients.

Macaulay and colleagues postulated that in patients with colloid cysts, as the patient ages, the ectopic goblet cell rests secrete an amorphous, proteinaceous material resulting in cyst formation and expansion. If the cyst enlarges rapidly, obstruction of CSF flow occurs and symptoms of increased intracranial pressure develop, and if there is abrupt blockage of CSF sudden death occurs due to increased intracranial pressure and secondary brain herniation. If, however, the cyst enlarges more gradually, the same is accommodated at the foramen of Monro without disruption of CSF flow and the patient remains asymptomatic as a steady state of CSF flow and absorption is achieved. Owing to this it would be difficult to ascertain as which subgroup of patients are at risk for neurologically deterioration.[18,19,20,21]

The imaging appearances of the cysts in our study were varied in terms of HU on plain CT and intensity on MRI. On MR imaging the three cysts were hyperintense on T1 and T2weighted sequences due to high mucin content. In particular, when cysts on CT were isodense or hypodense, MRI delineated the cysts better than CT. Two patients who underwent MRI had isodense cysts on CT; MRI was advised following CT for optimal delineation. However, regression analysis pertaining to cyst density and risk of deterioration was not significant, though the numbers encountered in our study are small, our findings concurs with what has been documented in literature.[24,25]

Given the high estimated risk of acute deterioration in symptomatic patients, lack of definite attributable risk factor, occurrence of refractory herniation & documented overall mortality as high as10%, prompt intervention should be resorted to particularly in symptomatic patients. The pathophysiological mechanisms responsible for acute deterioration and sudden death are the same despite the risk factors and condition at presentation, owing to irreversible cerebral herniation, which can be refractory to even emergent ventricular drainage, let alone surgical excision.
Early intervention in terms of unilateral or bilateral ventriculostomy can be lifesaving if instituted early. CT is adequate for diagnosis, subjecting the patient to MRI for confirmation or to corroborate findings can be done subsequent to ventriculostomy if need be.[12]

One of the limitations of this study, is that is a possibility that using the present recruitment method, the prevalence maybe underreported, as there would be a subset of patients who were seen as outpatients in whom neuroimaging was not done and presumably there is a considerable number of people harbouring colloid cysts who experience only minor symptoms or express no complaint which could result in underestimation of presentation. Lastly, not all patients who died suddenly undergo autopsy to ascertain the cause of death.

CONCLUSION

Desai et al in their series advocated emergency shunt procedure in patients with acute neurological deterioration as an interim procedure to cyst excision. None of the patients in whom a shunt was performed died. In our series, estimated risk of deterioration was 36%. The pathophysiological mechanisms responsible for acute deterioration and sudden death are the same despite the risk factors and condition at presentation, owing to irreversible cerebral herniation, which often can be refractory.

The purpose of this article is to highlight the necessity of early intervention in terms of unilateral or bilateral ventriculostomy which can be lifesaving if and when instituted early. In our own experience, given the circumstances in which mortality occurred, we advocate ventriculostomy as an interim procedure to corroborative imaging with MRI or definitive surgery.

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REFERENCES

[1] Hernesniemi J, Leivo S. Management outcome in third ventricular colloid cysts in a definedpopulation: A series of 40 patients treated mainly by transcallosal microsurgery. Surg Neurol 45:2-14, 1996.
[2] M.S. Greenberg, Tumor, in: M.S. Greenberg (Ed.), Handbook of Neurosurgery, Georg Thieme,New York, pp. 437-439, 2001.
[3] K. Kuchelmeister, M. Bergmann, Colloid cysts of the third ventricle: an immunochistochemicalstudy, Histopathology 21 (1992) 35-42.
[4] M. Skerbinjek Kavalar, R. Kavalar, T. Strojnik, A colloid cyst of the third ventricle—the cause ofepisodic headache and sudden unexpected death in an adolescent girl, Wien Klin. Wochenschr. 117(2005) 837-840.
[5] D.M. Hadley, Colloid cyst of the IIIrd ventricle, J. Neurol. Neurosurg. Psychiatry 72 (2002) 15.
[6] T. Beems, T. Menovsky, M. Lammens, Hemorrhagic colloid cyst: case report and review of theliterature, Surg. Neurol. 65 84-86, 2006.
[7] H.H. Schmidek, Operative Neurosurgical Techniques, WB Saunders, Philadelphia, 2006.
[8] Lawrence JE, Nadarajah R, Treger TD, Agius. Neuropsychiatric Manifestations of Colloid Cysts: areview of the literature. Psychiatr Danub. Sep; 27 Suppl 1:S315-208, 2015.
[9] A.P. Amar, S. Ghosh, M.L.J. Apuzzo, Ventricular tumors, in: H.R. Winn (Ed.), Youmans Neurological Surgery, Saunders, Philadelphia, 2004, pp. 1245-1246.
[10] M.W. Partington, A.J. Bookalil, Familial colloid cyst of the third ventricle, Clin.Genet. 66 (2004)473-475.
[11] P.C. de Witt Hamer, M.J. Verstegen, R.J. De Haan, W.P. Vandertop, R.T. Thomeer, J.J. Mooij,W.R. van Furth, High risk of acute deterioration in patients harbouring symptomatic colloid cysts ofthe third ventricle, J. Neurosurg. 96 (2002) 1041-1045.
[12] Camacho A, Abernathey CD, Kelly PJ, Laws ER Jr. Colloid cysts: Experience with the management of 84 cases since the introduction of computed tomography. Neurosurgery 1989;24:693-700.
[13] Desai KI, Nadkarni TD, Muzumdar DP, Goel AH. Surgical management of colloid cyst of thethird ventricle- a study of 105 cases. Surg Neurol. 2002 May: 57(5):295-302.
[14] Pollock BE, Huston J III: Natural history of asymptomatic colloid cysts of the third ventricle. JNeurosurg 91:364-369, 1999.
[15] Pollock BE, Schreiner SA, Huston J III: A theory on the natural history of colloid cysts of thethird ventricle. Neurosurgery 46:1077-1083, 2000.
[16] Buttner A, Winkler PA, Eisenmenger W, et al: Colloid cysts of the third ventricle with fataloutcome: a report of two cases and review of the literature. Int J Legal Med 110:260-266, 1997.
[17] Chen TC, Krieger M, Hinton DR, et al: The colloid cyst, in Apuzzo MLJ (ed): Surgery of theThird Ventricle, ed 2. Baltimore: Williams & Wilkins, 1998, pp 1071-1132.
[18] Lewis AI, Crone KR, Taha J, et al: Surgical resection of third ventricle colloid cysts. Preliminaryresults comparing transcallosal microsurgery with endoscopy. J Neurosurg 81:174-178, 1994.
[19] Little JR, MacCarty CS: Colloid cysts of the third ventricle. J Neurosurg 40:230-235, 1974.
[20] Luke JL, Helpern M: Sudden unexpected death from natural causes in young adults. A review of275 consecutive autopsied cases. Arch Pathol 85:10-17, 1968.
[21] Macaulay RJ, Felix I, Jay V, et al: Histological and ultrastructural analysis of six colloid cysts inchildren. Acta Neuropathol 93:271-276, 1997.
[22] Mathiesen T, Grane P, Lindgren L, et al: Third ventricle colloid cysts: a consecutive 12-yearseries. J Neurosurg 86:5-12, 1997.
[23] Antunes JL, Louis KM, Ganti SR: Colloid cysts of the third ventricle. Neurosurgery 7:450-455,1980.
[24] Guner M, Shaw MDM, Turner JW, et al: Computed tomography in the diagnosis of colloid cyst.Surg Neurol 6:345-348, 1976.
[25] Powell MP, Torrens MJ, Thomson JLG, et al: Isodense colloid cysts of the third ventricle: adiagnostic and therapeutic problem resolved by ventriculoscopy. Neurosurgery 13:234-242, 1983.