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Extrarenal manifestation of Autosomal Dominant Policystic Kidney Disease (ADPKD)- A Case reporting

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Abstract: ADPKD is a systemic disease, marked by progressive increase of bilateral renal disease, diverticular disease., liver and pancreatic cystic disease, ICA aneurysm in 6% of ADPKD patients. Other CNS manifestation of ADPKD is arachnoid cyst -8%, chronic SDH. Most of ADPKD with ICA aneurysm incidence is icebergh phenomenon due to asymptomatic nature. In india , in my institute named omanthurar multispeciality hospital Chennai city., there is a case of 45 years aged male, known case of ADPKD., patient coming with c/o head ache. Patient taken for CT Angiogram for which have been revealed multiple aneurysm with varying size were located in Acom , Rt MCA bifurcation., and Pcom region. The Aortogram was revealed abdominal aneurysm too. This patient taken up for surgical procedure all three aneurysm clipped in single sitting surgical procedure.
Key words: ADPKD.,Multiple Aneurysm., autosomal dominant., extra renal .

References: *Text book of pathology by Robinson"s.,*

Text book of neurosurgical technique author by sweet and schimidit. Youman"s text book of neurosurgery.

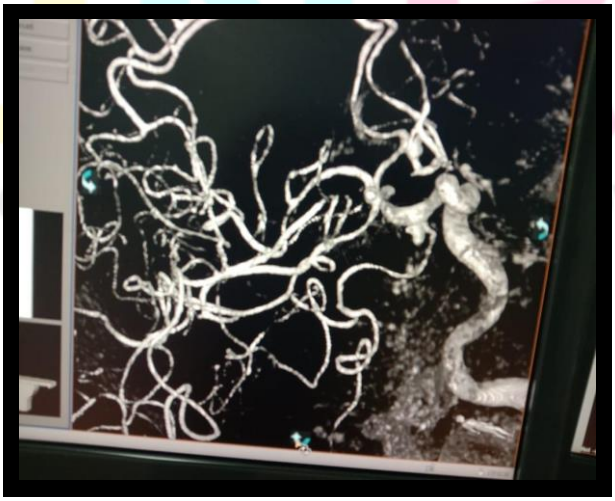
Introduction:

ADPKD compromise 5-10 % of end stage renal disease . ADPKD is caused by mutation in PKD1 or PKD2 genes, with allele frequencies of 1:500 to 1:1000. PKD1 and PKD2 GENE ALLELES located in chromosomes 16. ADPKD is systemic disease with cyst formation over the kidney., liver,., pancrease., aneurysmal formation over the cerebral blood vessels., abdominal aorta. Other manifestation of ADPKD is smallintestinal diverticula., abdominal wall hernia.

CNS manifestations of ADPKD: Arachnoid cyst., spinal meningeal cyst and aneurysm

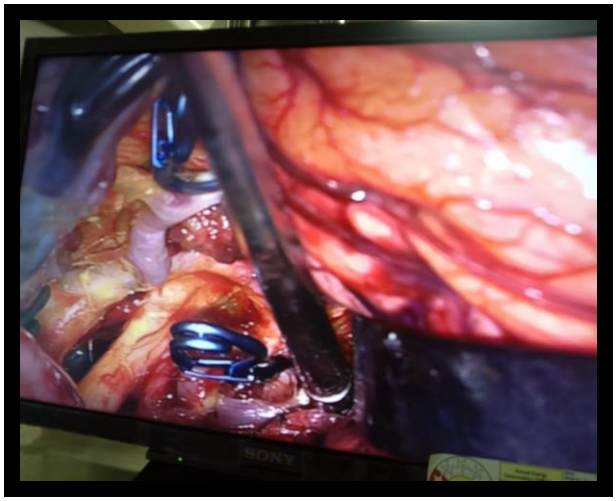
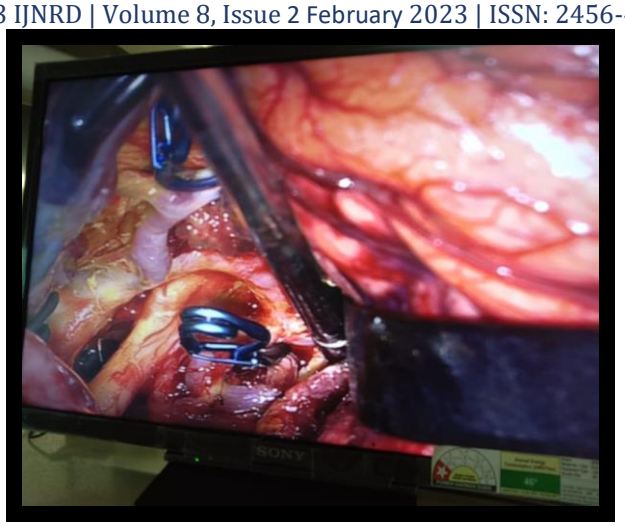
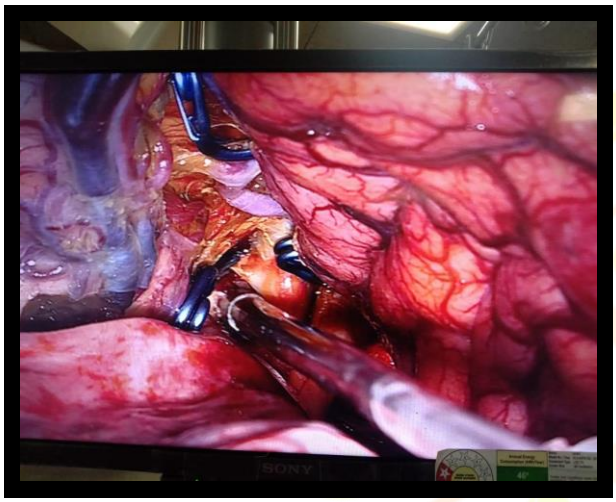
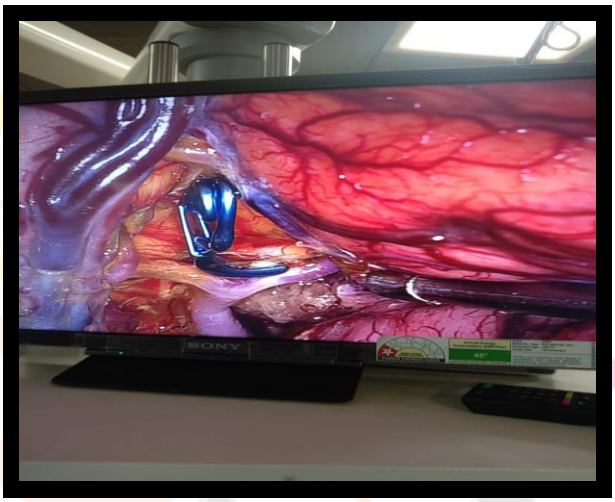
Aneurysm: Asymptomatic intracranial have been detected in - 6%., of ADPKD patients without family history and approximately 20% of patients with family history. In contrast., rate of ICA aneurysm in the general population is 1 to 2%. Rupture of ICA aneurysm in ADPKD is five times higher than general population. Abdominal aortic aneurysm also reported in ADPKD patients.

Case report: A 40 years aged male patient coming with c/o head ache with vomiting., the CT Scan brain shows RT sylvian fissure SAH grading 2. Hence further evaluation patient undergone Digital Substraction Angiography shows RT MCA M1 bifurcation aneurysm., ACOM aneurysm., and PCOM aneurysm. The AORTOGRAM revealed abdominal aortic aneurysm which is more associated frequency of ADPKD. The USG abdomen shows poly cystic kidney disease.



DSA done in omanthur multi super specialty hospital shows Acom, Pcom, RT m1 MCA Aaneurysm

The patient taken for Right pterional craniotomy., all the anterior circulation aneurysm clipped with appropriate clips at single setting.

DISCUSSION: ADPKD is a autosomal dominant PKD1 and PKD2 Genes mutation alleles located in chromosome 16. The ADPKD Is a 5 to 10% of end stage renal disease contribution with extra renal manifestation are GI tract diverticula cystic disease over the liver, pancreas. Cardiac manifestations are early HT, LVH, Mitral valve prolapse. The CNS manifestations of ADPKD is arachnoid cyst of brain and spinal cord, multiple intracranial aneurysm and abdominal aneurysm. In omanthur multi specialty hospital Chennai tamilnadu, india dept of neurosurgery identified a 40 years aged male patient with features of multiple cyst over the liver and kidney, intra abdominal aortic aneurysm, multiple intracranial aneurysm over the Acom., Rt M1 bifurcation aneurysm, PCOM aneurysm which was clipped in single sitting. Autosomal dominant polycystic kidney disease (ADPKD) is the most common form of PKD. Most people with ADPKD have defects in the *PKD1* gene, and 1 out of 6 or 1 out of 7 people with ADPKD have a defective *PKD2* gene. ADPKD affects 1 in every 400 to 1,000 people and is the most common kidney disorder passed down through family members. Abnormal heart valves, brain aneurysms. Abdominal aortic aneurysm, liver cysts; pancreatic cysts; diverticula, are common extra renal manifestations of ADPKD. In this case manifested with adult onset polycystic kidney , multiple liver and renal cyst, abdominal aneurysmal aorta, multiple intra cranial aneurysm over the Acom, Rt Pcom and RtM1 MCA territory.

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Text book of pathology by Robintson"s., 4th edition.
Text book of neurosurgical technique author by sweet and schimidit 6th edition.
Youman"s text book of neurosurgery 7th edition.