Cavernous Angioma

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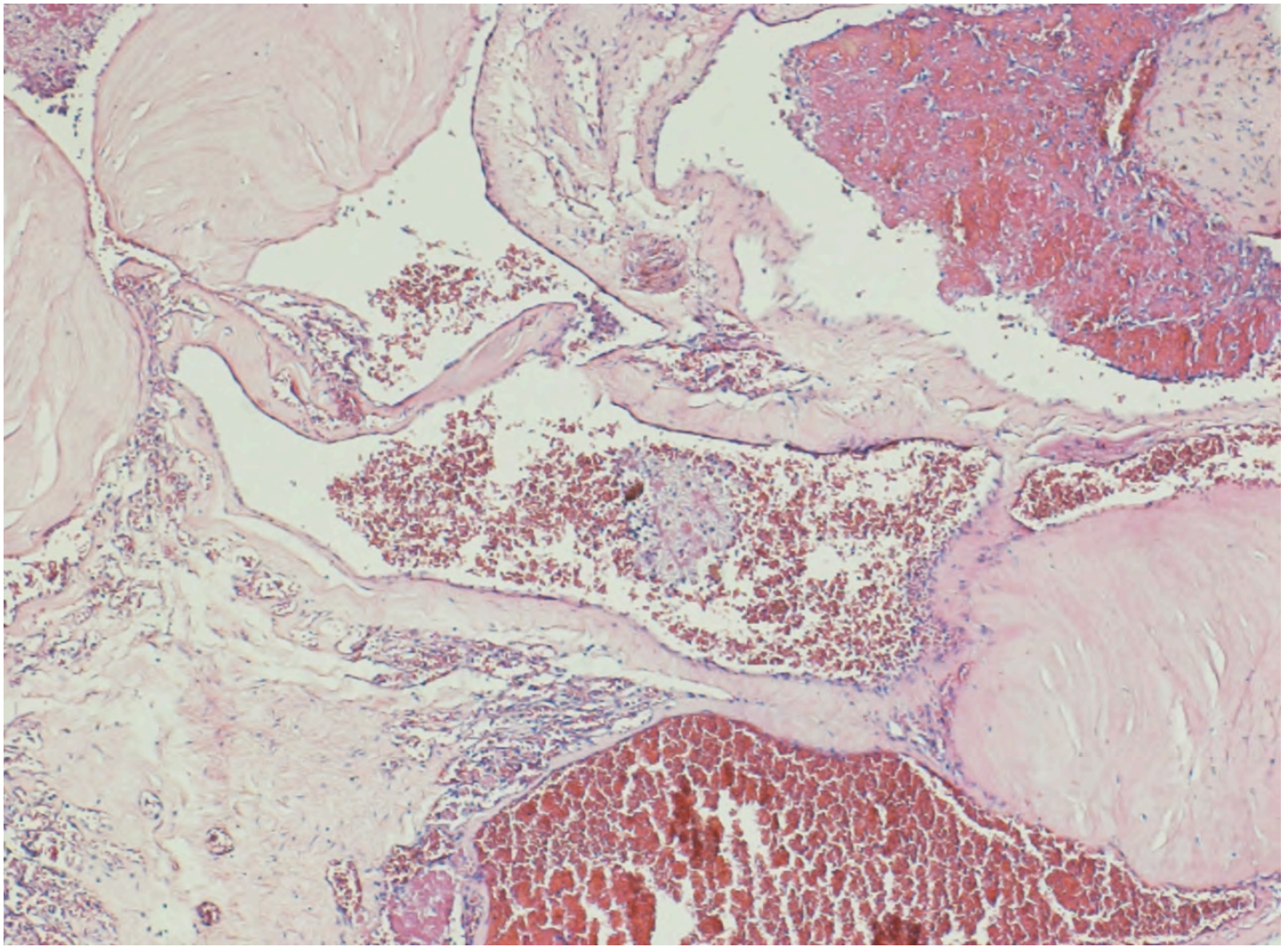
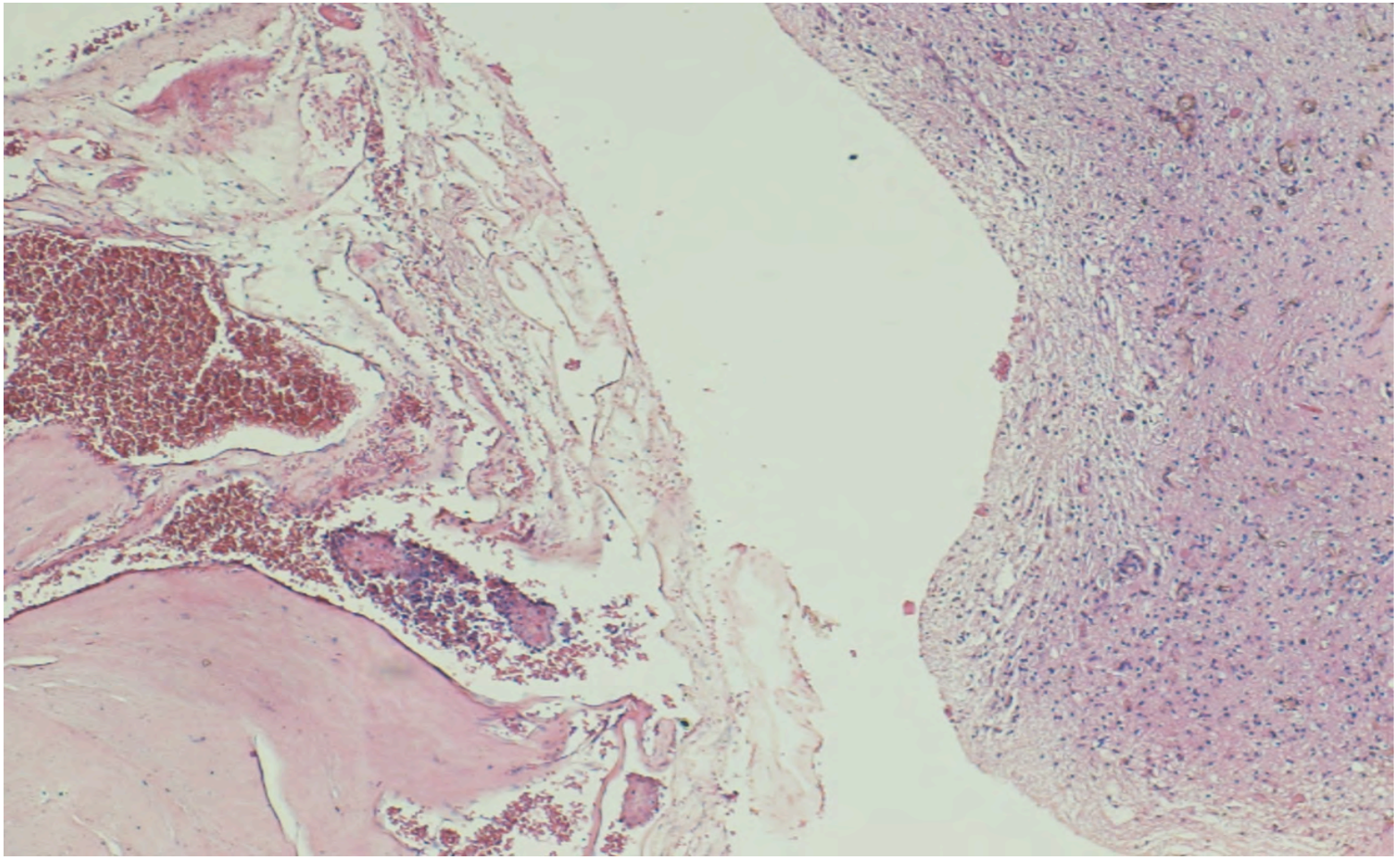
Intracranial cavernous angioma are vascular anomalies consisting of endothelium-lined caverns filled with blood at various stages of thrombosis and organization and separated by a collagenous stroma devoid of mature vessel wall elements

Large, thin-walled blood-filled vascular spaces seperated by a mostly thin fibrocollagenous stroma



Large, irregular blood spaces separated by hyalinized fibrocollagenous tissue. Adjacent glial tissue is also seen in the second picture on the right side.



Cavernous angioma

- The international studies suggest the at the incidence of cavernous angioma is 0.5-0,7% of the population worldwide
- We would like to discuss our limited experience of 47 cases of cavernous angioma, with all of you.
- Period of study between 1990-2010.

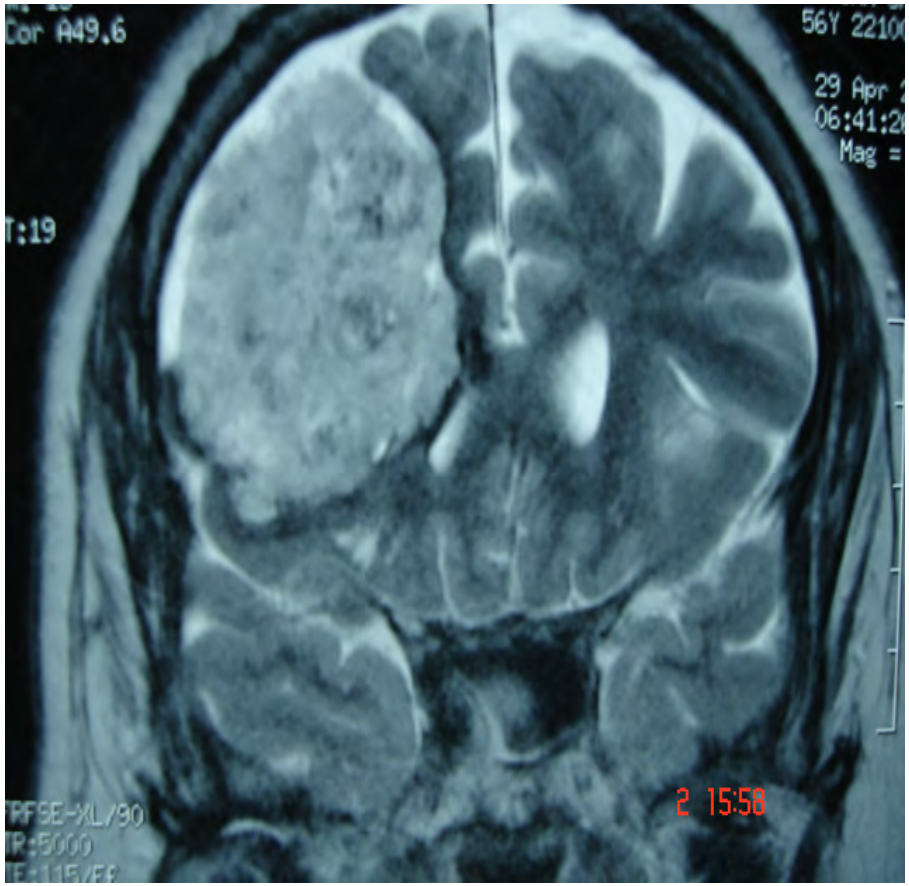
- Age from one day to 64 years old
- 16 males and 14 females
- 36 patients had been operated upon
- 3 of them have been operated more than one time
- 11 patients refused surgery or they have asymptomatic cavernous angioma
- 3 Saudi families were found out that many members of these families harbor cavernous angioma.

Anatomical distributions of the Cavernous Angioma

- Brain stem 4
- Posterior fossa 5
- Supratentorial 40
- Orbit 2
- Multiple 7

The clinical presentation

- Epilepsy 13
- Bleeding 16
- Exophthalmous 2
- Asymptomatic 7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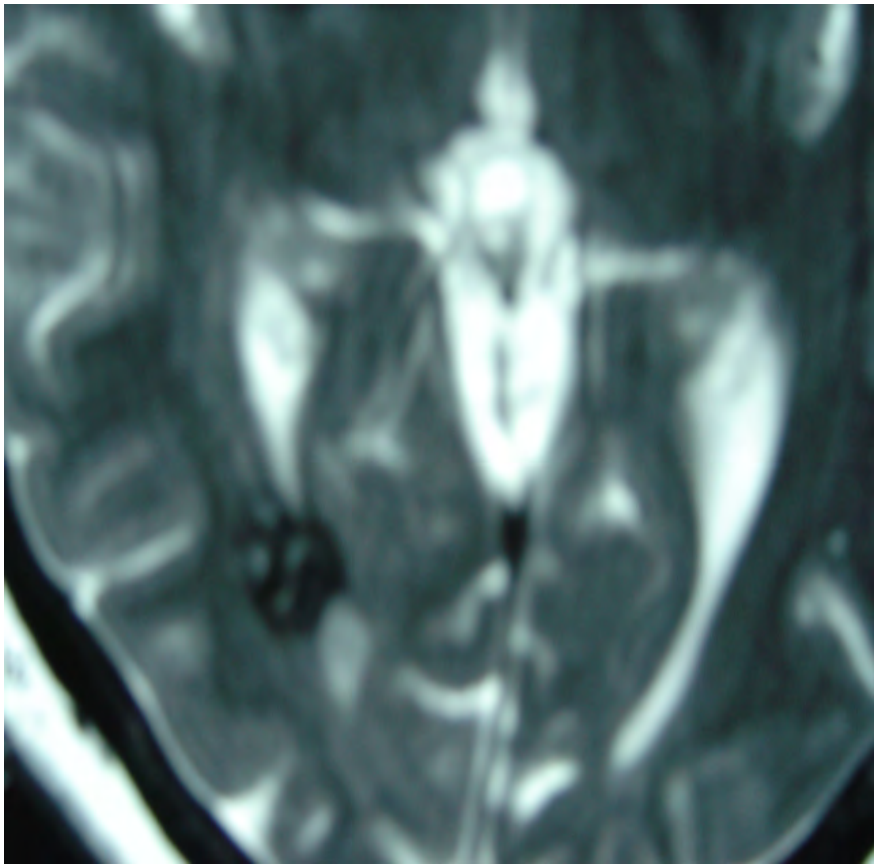
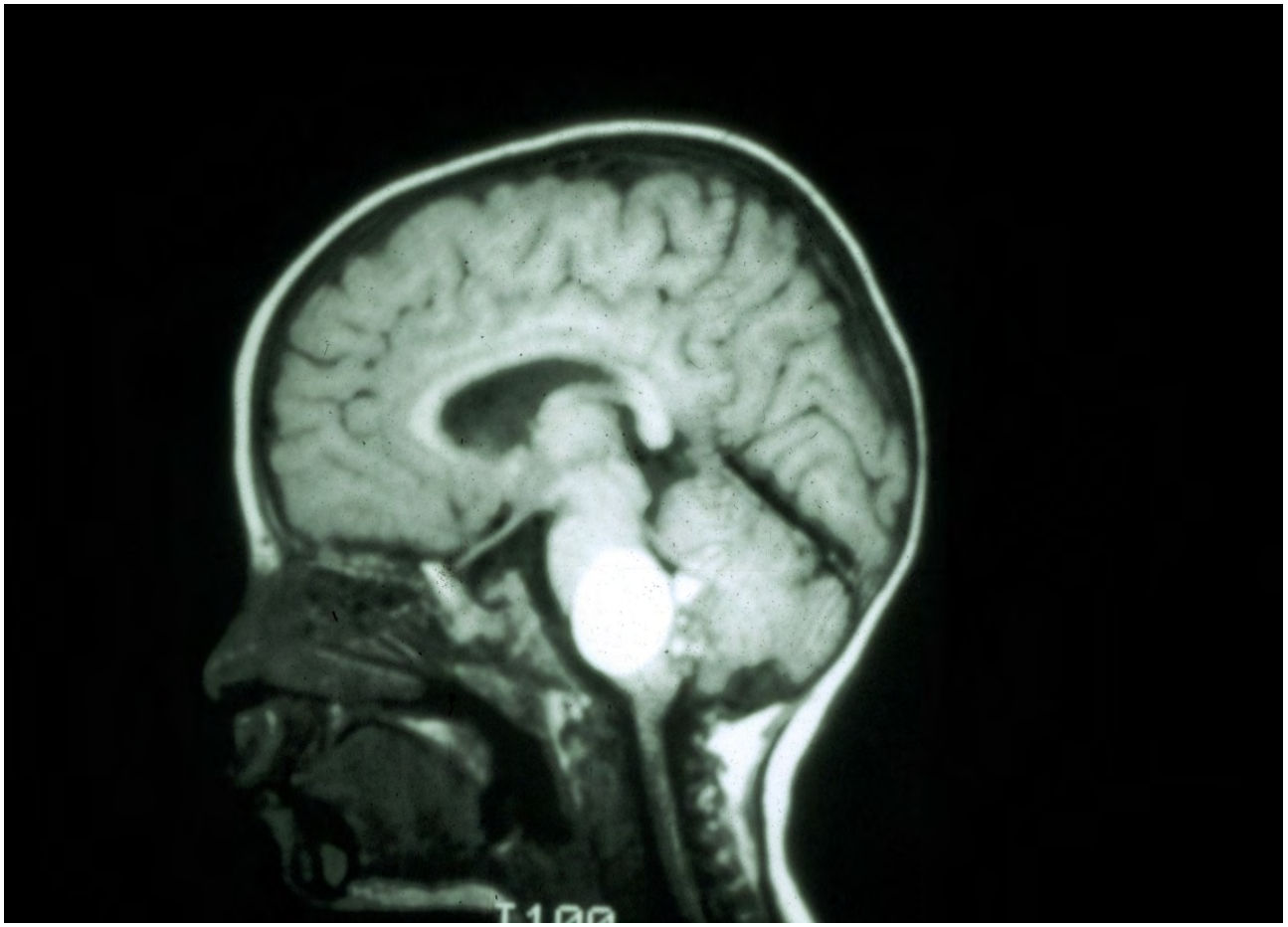


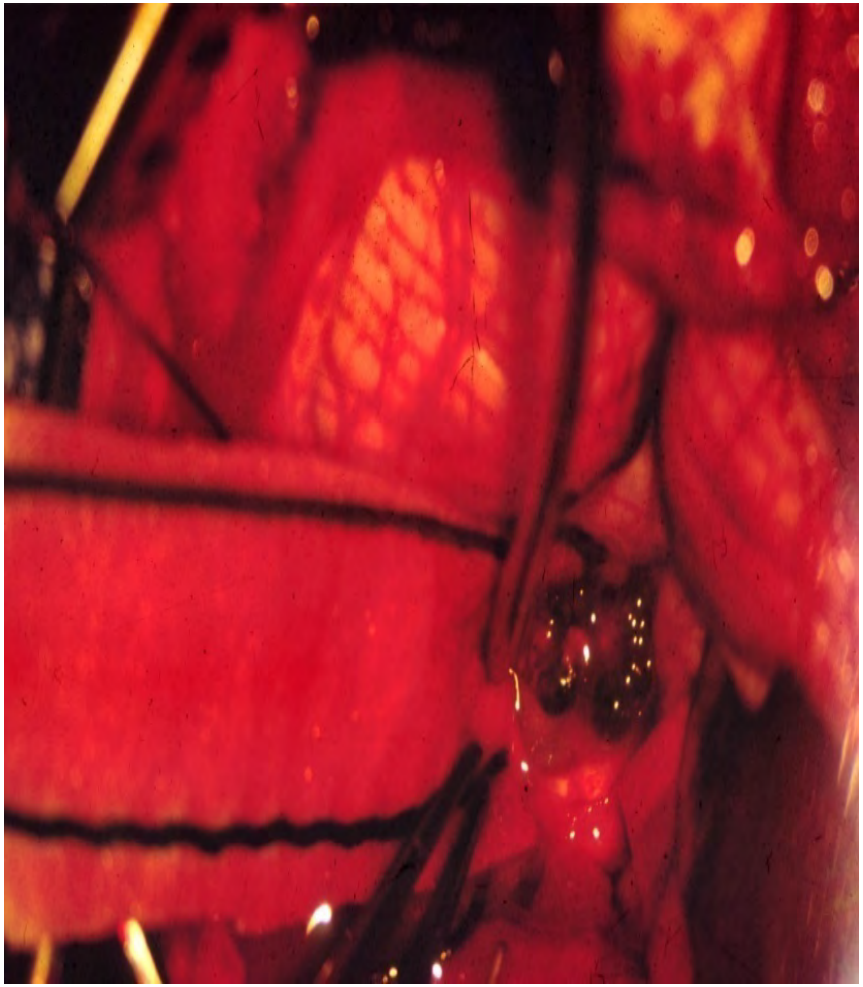
Case 1

- 22 months old baby boy was transferred to our hospital unconscious and ventilated. He suffered a sudden attach of loss of consciousness , III, IV,VI,VII , IX,X cranial nerves palsy. Quadri paresis mainly in the left side.
- CT-Scan and MRI showed brain stem Cavernous angioma

MRI showed large brain stem caverous angioma

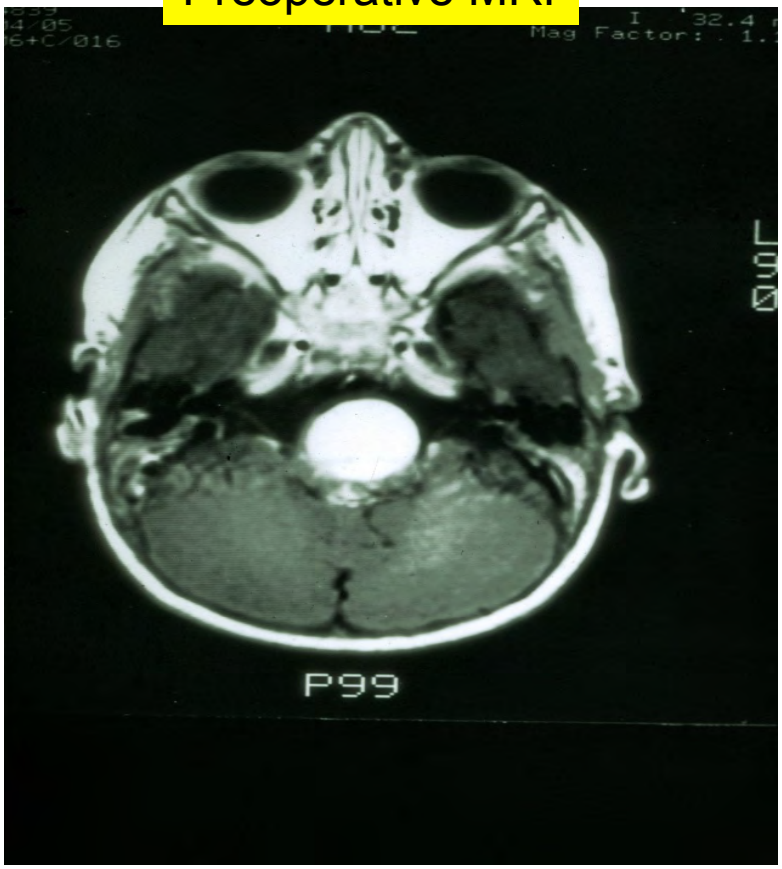


Sub occipital craniotomy and midline approach.



Patient made very good recovery and discharged after 21 days.

Preoperative MRI



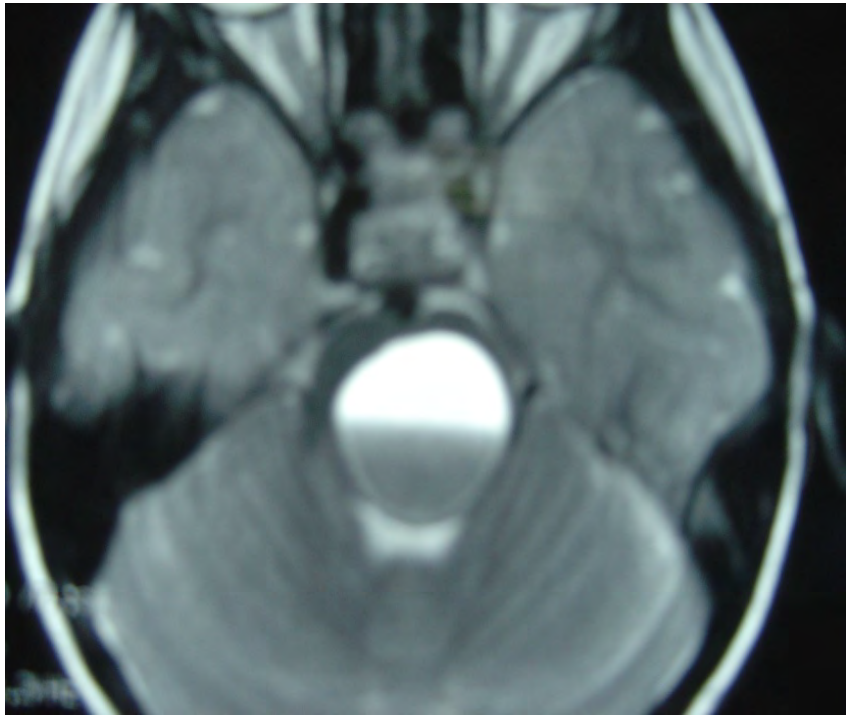
Post operative MRI

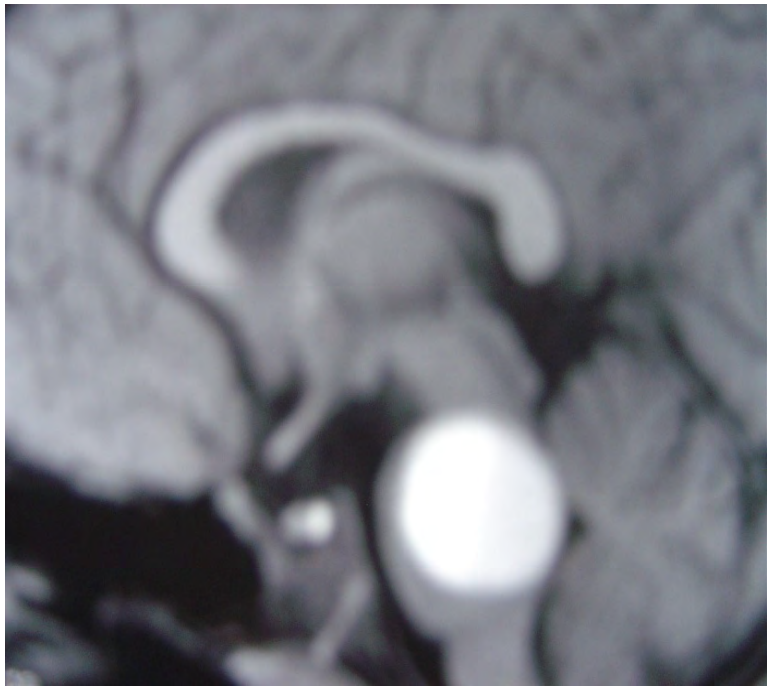
Case 2

- 4 years old girl suffered a sudden attack of sever headache and right side hemi paresis.
- Patient was brought to the ER
- On examination, fully conscious but drowsy girl. She was suffering of headache. Right side 6 and 7 nerves palsy had been detected + Right side hemi paresis

- CT-Scan and MRI showed brain stem bleeding
- The patient was taken to the OR for aspiration of the hematoma and try to find the cause of bleeding.
- The hematoma was aspirated but we failed to find lesion.
- The patient made very good recovery.
- MRA failed to suggest any vascular anomalies.

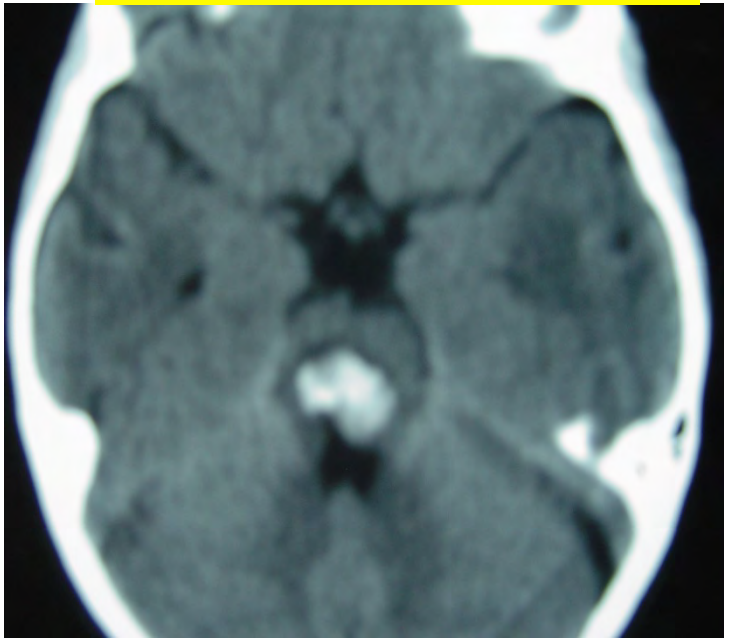
MRI at admission showed brain stem hemorrhage



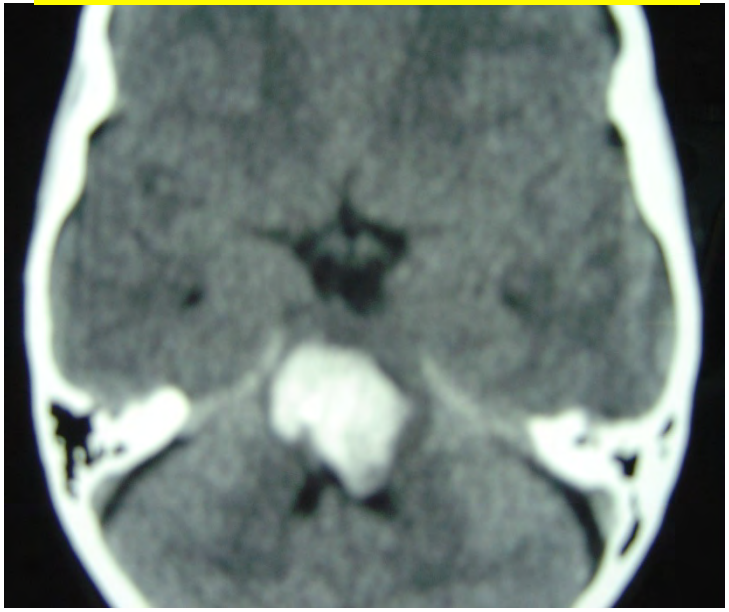


- **Few days later, patient became suddenly unconscious.**
- **MRI showed massive brain stem bleeding**
- **Patient was taken immdeitely to the OR and she was re-operated and the cavernous angioma was found and removed**
- **Patient made excellent recovery.**

MRI after the first surgery

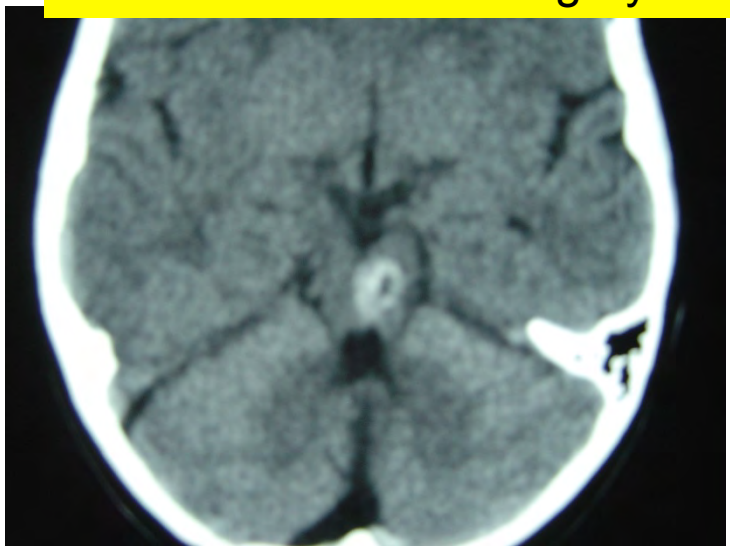


2ND attack of hemorrage

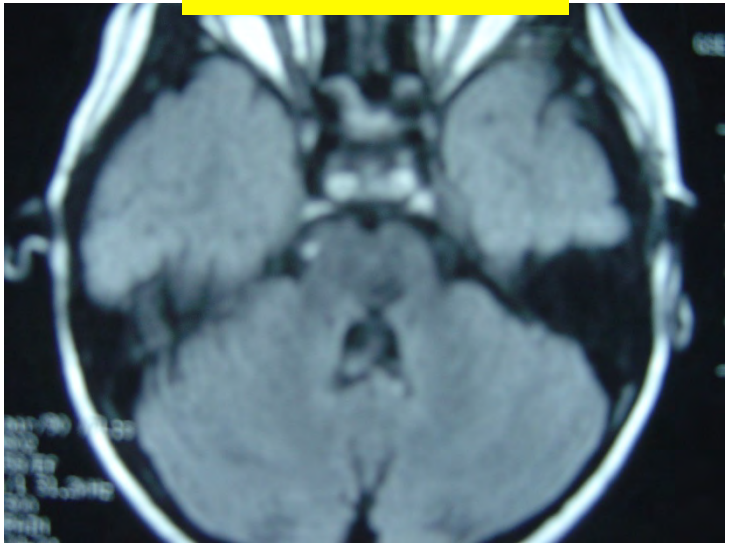


Follow up

One week after 2ND surgery



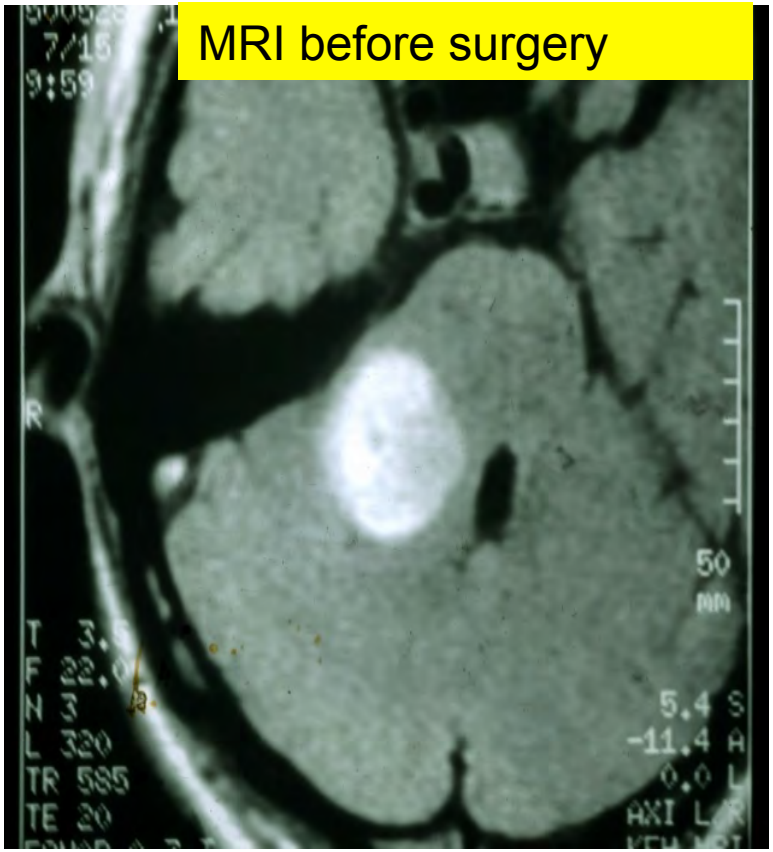
One month later



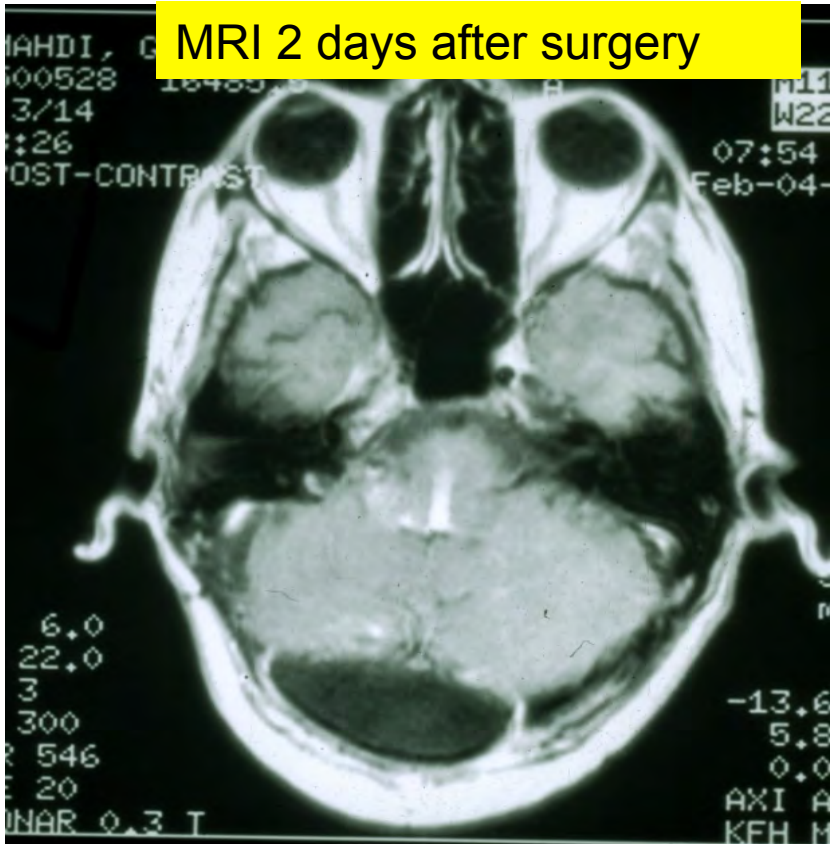
Case 3

- 28 years old man had a sudden attack of headache and loss of hearing as he was making a phone conversation.
- The patient was fully conscious orientated suffers right VII, and VIII cranial nerves palsy. No motor or sensory deficit
- MRI and CT-scan showed Rt CP angle lesion

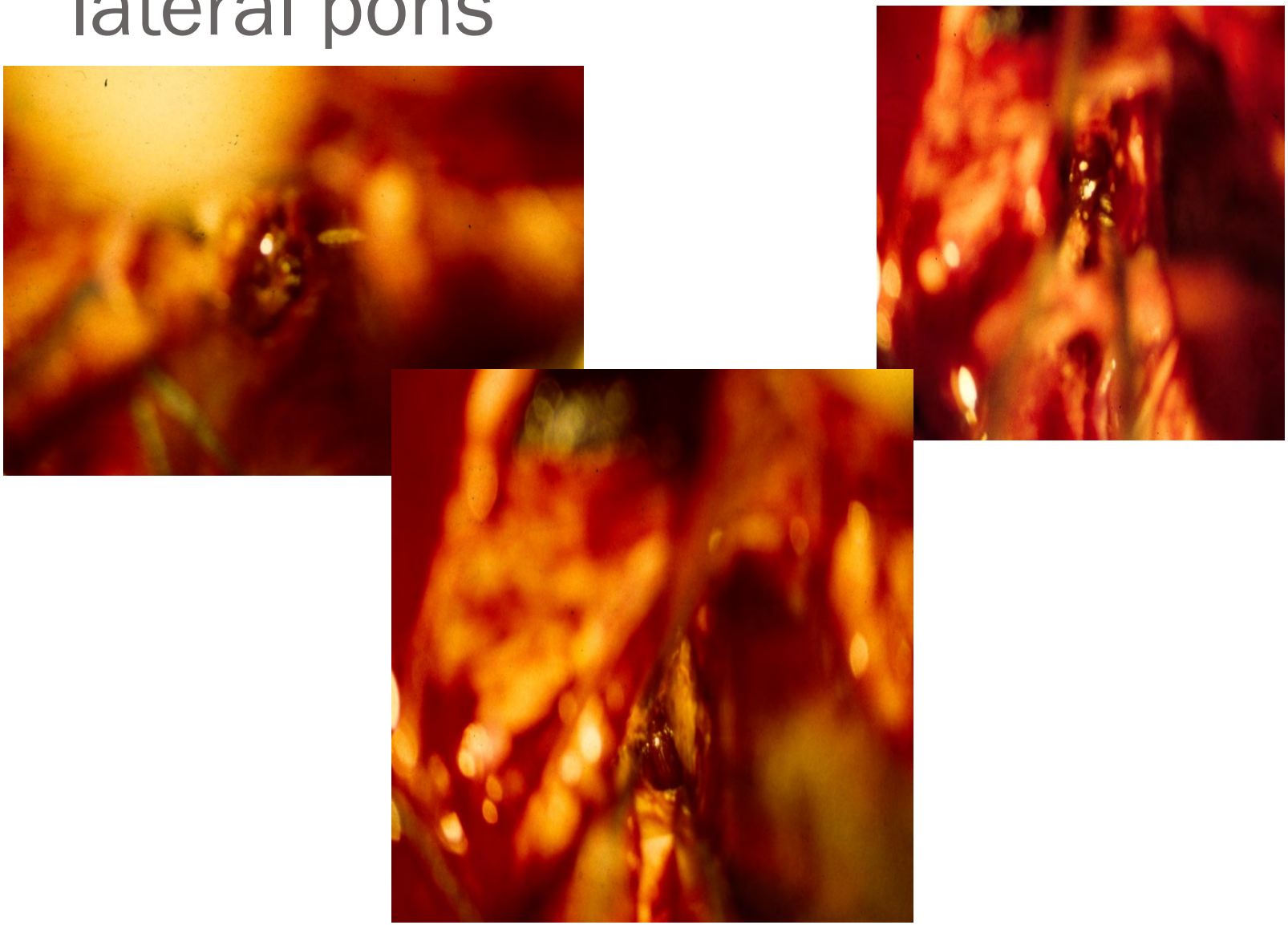
MRI before surgery



MRI 2 days after surgery

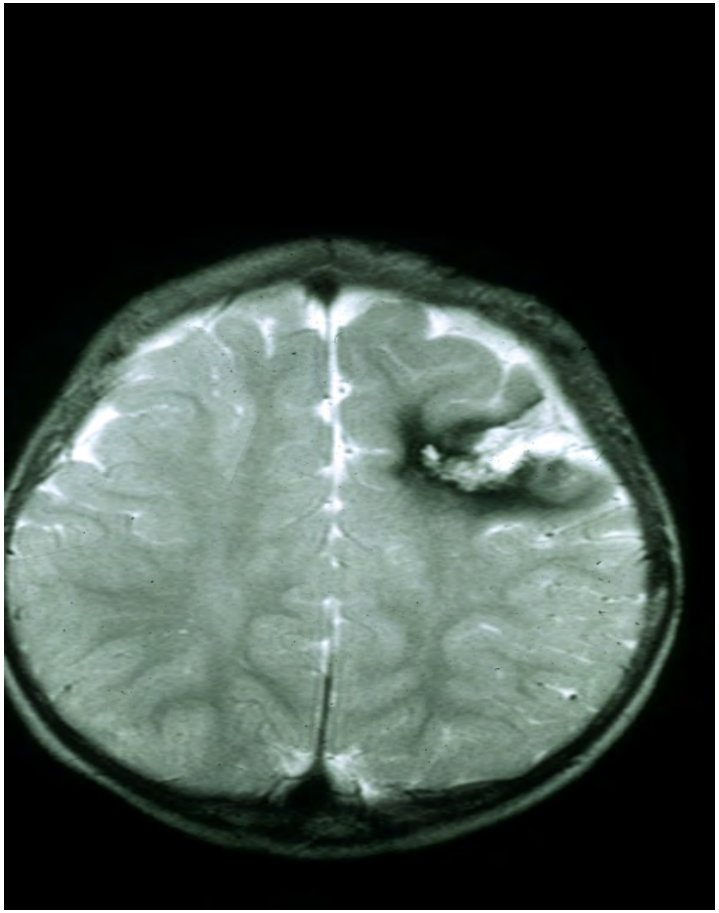


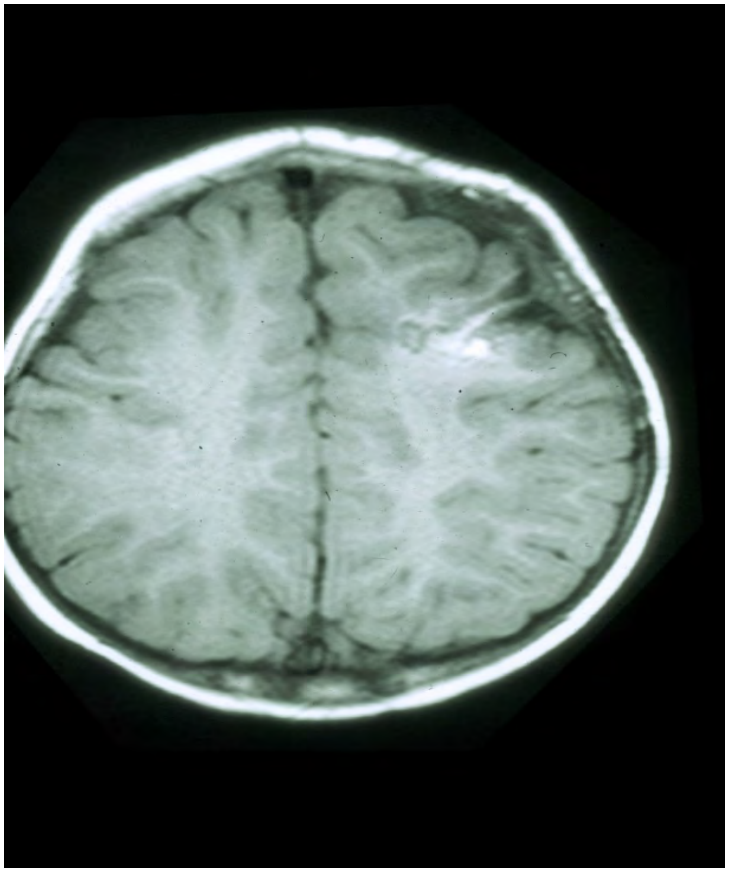
Retromastoid approach to the lateral pons



Case 4

- 64 years old lady suffered an attack of headache and followed 3 days later by an epileptic fit.
- She was seen in our service , neurologically free.
- CT-Scan and MRI done and showed multiple cavernous angioma, One in the left parietal lobe with evidence of bleeding.
- The patient had 2 cavernous angiomas in the right temporal lobe

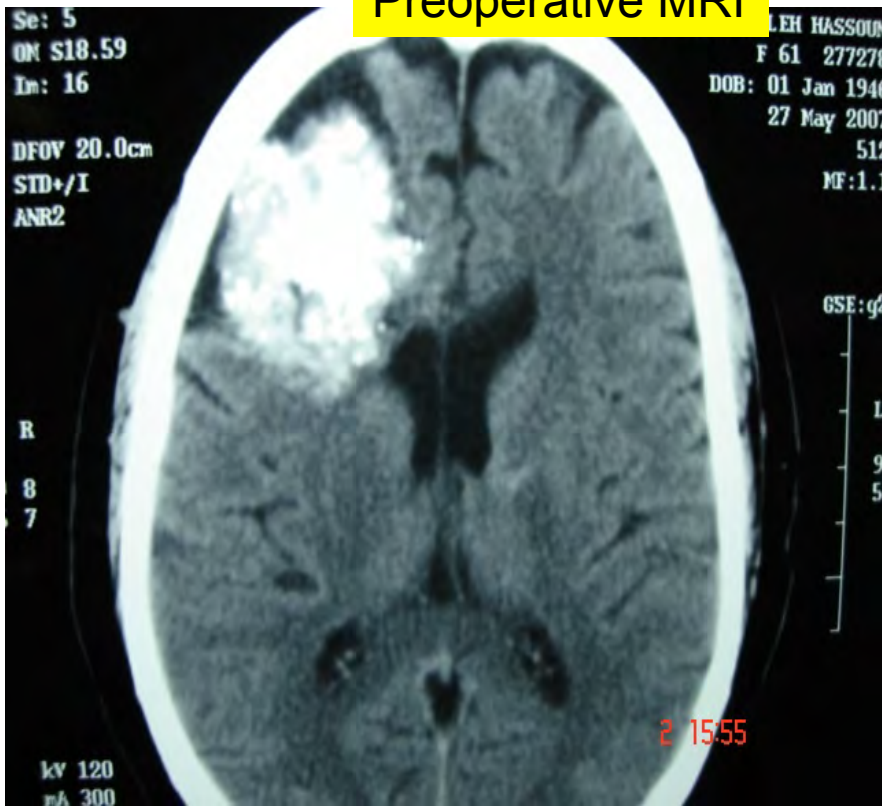




Case 5

- 54 years old lady has been complaining of epilepsy since her childhood. She was also diagnosed as a case of depression and personal behavior changes for last 10 years.
- She was on anti depressant and anti epileptic medication
- A week prior to her admission she suffered status epileptics

Preoperative MRI



P3 days Post operative MRI

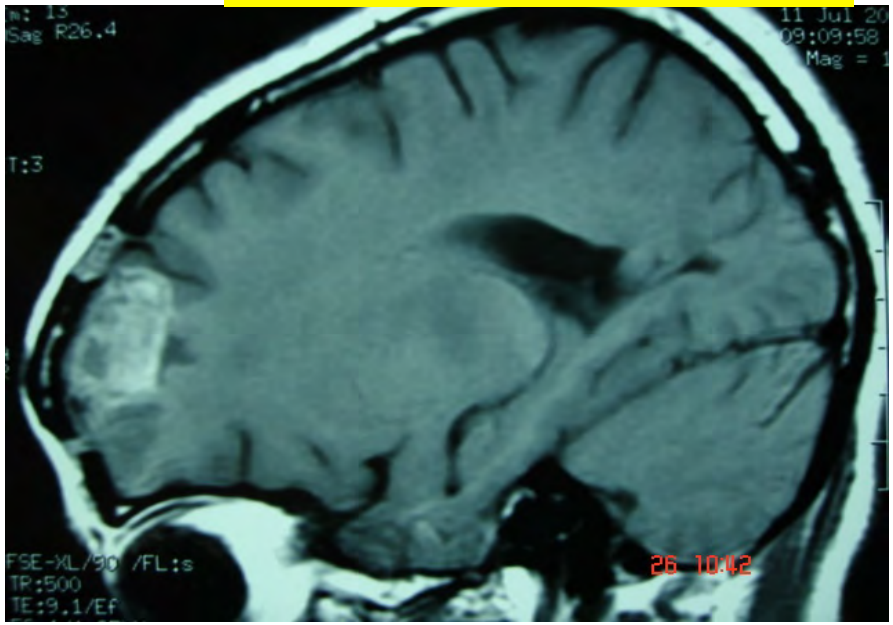
Removal of the frontal cavernous angioma

Case 6

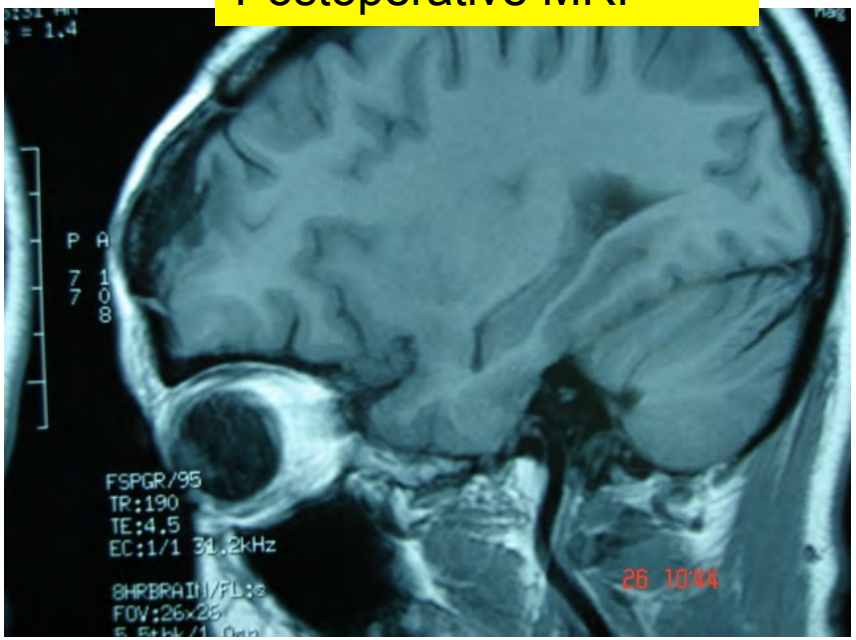
- 15 years old student has been diagnosed as case of epilepsy since age of 2 years
- MRI was done few years ago and was diagnosed as a case of Cavernous angioma.
- At the day of admission, he has a severe attack of headache and loss of conscious for few seconds

Removal of the cavernous angioma through small frontal craniotomy

Prioperative MRI



Postoperative MRI



Cavernous angioma in Neonates
Case 7

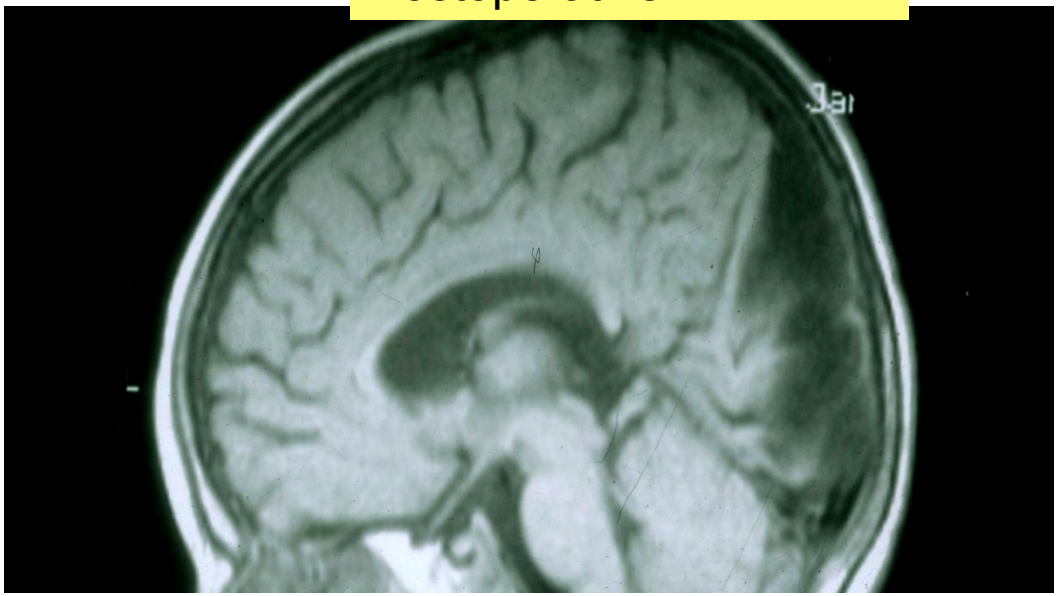
- 31 weeks premature newborn was delivered via CS as the obstetrician detected fetal distress.
- APGAR Score was law (5)
- CT-Scan and MRI was preformed and showed brainstem bleeding and possible cavernous angioma.
- The baby rapidly deteriorated and intubated
- We had been consulted, we decided to take the patient to the operating room for subocciptal approach and removal of the cavernous angioma and the hematoma
- The cavernous angioma was removed, however and unfortunately the patient arrested and died on the table.

Case 8

- Full term baby was delivered APGAR Score was 8. 3 days later the new born suddenly became irritable and subsequently lost consciousness.
- Urgent CT-Scan and MRI showed large right occipital cavernous angioma and bleeding.
- The patient was taken immediately to the OR and the hematoma and cavernous angioma were removed.
- The patient mad excellent recovery.
- He was not put on any anti epileptic drugs!.

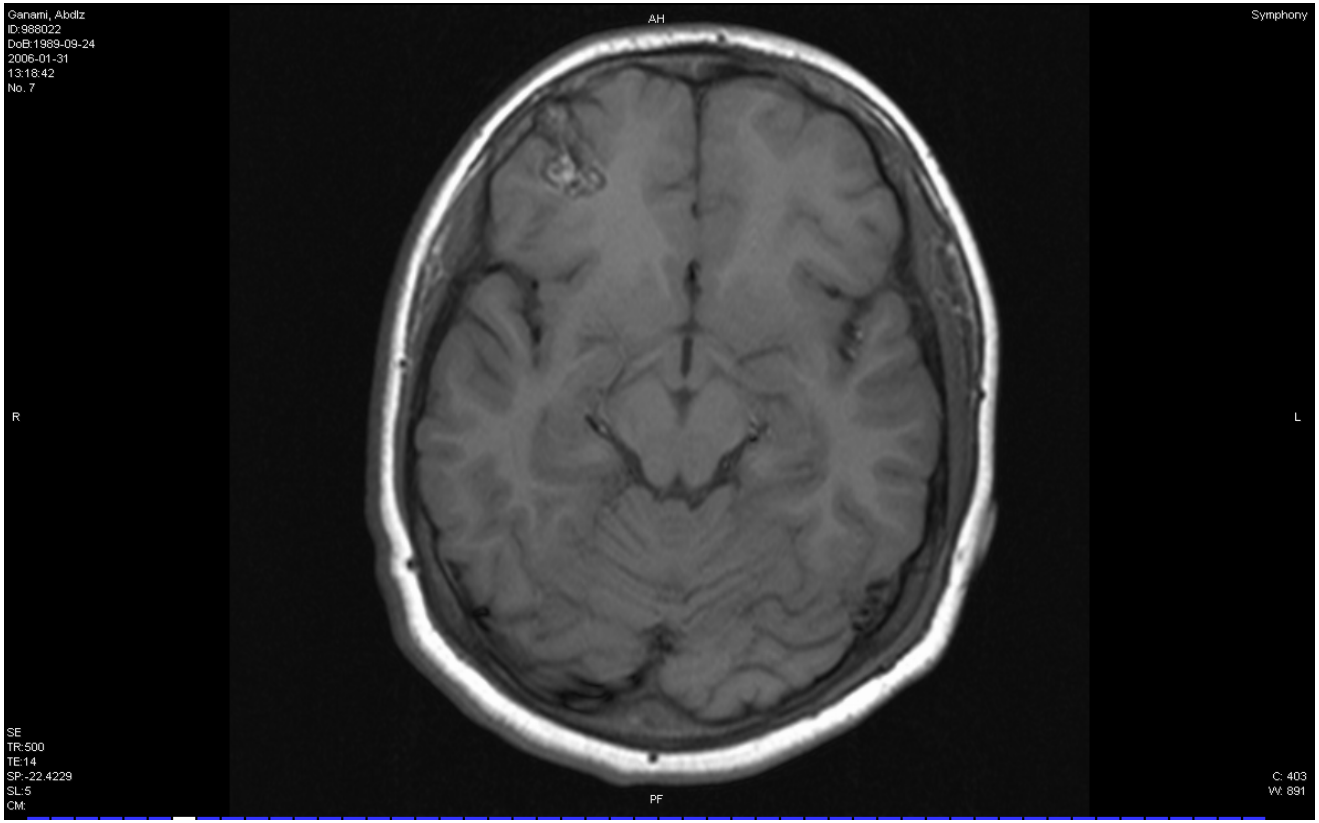
Preoperative MRI

Postoperative MRI



Medically treated patients
Case 9

26 Years old male, Complains of epilepsy which is well controlled on Tegreteol. He refused surgery



Case 10

- 64 years old lady was diagnosed as a case of epilepsy for 22 years.The epilepsy is well controlled on medication.
- She refused surgery.
- She has been followed up for 7 years in my clinic.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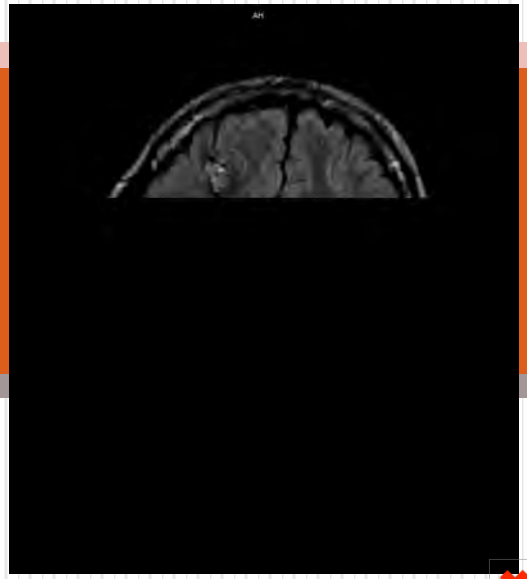
Family Cavernous angioma syndrome



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Case 11

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Case 11

- 19 years old university student complaining of epilepsy since he was 4 years old. He complaints of on and off headache.
- MRI showed multiple cavernous angioma, , left Sylvian fissure, right parietal and right cerebellum.
- He refused surgery as the epilepsy is well controlled on 2 medications
- The father and the brother of this patient have multiple cavernous angiomas too, however, none of them did complaint of epilepsy or clear signs of bleeding. Both complaints of headache from time to time.

Family Cavernous angioma syndrome

We have 3 Saudi families

First family, Grand mother, 2 daughters, 3 grand daughters – No males (five females have been operated , one baby (two years old girl was not operated asymptomatic)

Second family, father and 2 brothers (no females) One brother is operated the others refused surgery.

Third family, mother has multiple small cavernous angioma, and only one of her 3 sons has asymptomatic cavernous angioma.

Characters of family Cavernous angioma syndroem

- 1, Multipilicity
- 2. Various size and location
- 3. The majority complained by epilepsy, 2 asymptomatic one only had bleeding
- 4. Majority supratentorial
- The three families are Saudi families and from the Eastern provience of Saudi Arabia
- They were sent for genetic studies.

Genetic Study

- The results were not conclusive to find special gene. It was suggested that there is autosomal dominant pattern of inheritance.

- This condition is inherited in an autosomal dominant pattern, which means one copy of the altered gene in each cell is sufficient to cause the disorder. In some cases, an affected person inherits the mutation from one affected parent

Genetic causes of familiar cavernous angioma

- CCM may be inherited due to mutations in one of three genes, CCM1, CCM2, or CCM3 – individuals with this inherited form typically have multiple CCM lesions. Alternatively, single lesions are often seen in patients with the sporadic (non - genetic) form of CCM.
- Mutations in these three genes account for 70 percent to 80 percent of all cases of familial cerebral cavernous malformations.
- The remaining 20 percent to 30 percent of cases may be due to unidentified genes or to other unknown causes.

- **It was found as well that mutations in the *CCM2*, *KRIT1*, and *PDCD10* genes cause cerebral cavernous malfor*mation*.**

Question needs to be answered?

How many times patient should be operated for multiple lesions?

Demonstrating case !

- 28 months old girl was refereed to my service as after a severe attack of headache as case of refractory epilepsy
- MRI showed multiple left side parietal cavernous angioma.There is evidence of bleeding from the left side angioma

- **The patient was operated upon for total removal of the left parietal cavernous angioma, she made a remarkable recovery.**
- **The Epilepsy became well controlled . She was put only on one drug (Tegretol)**

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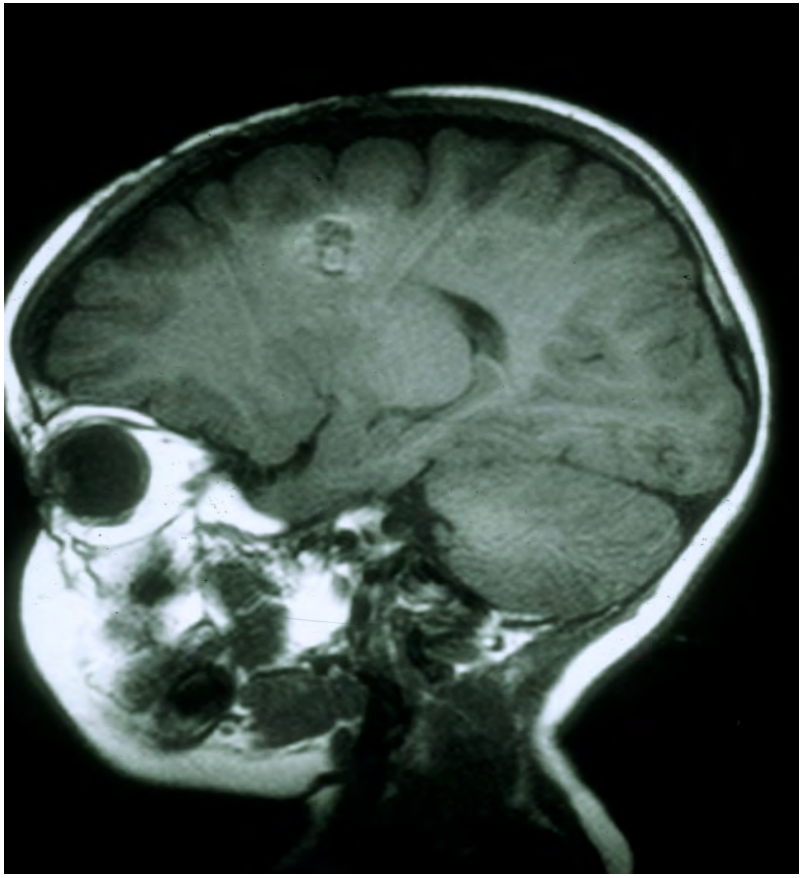
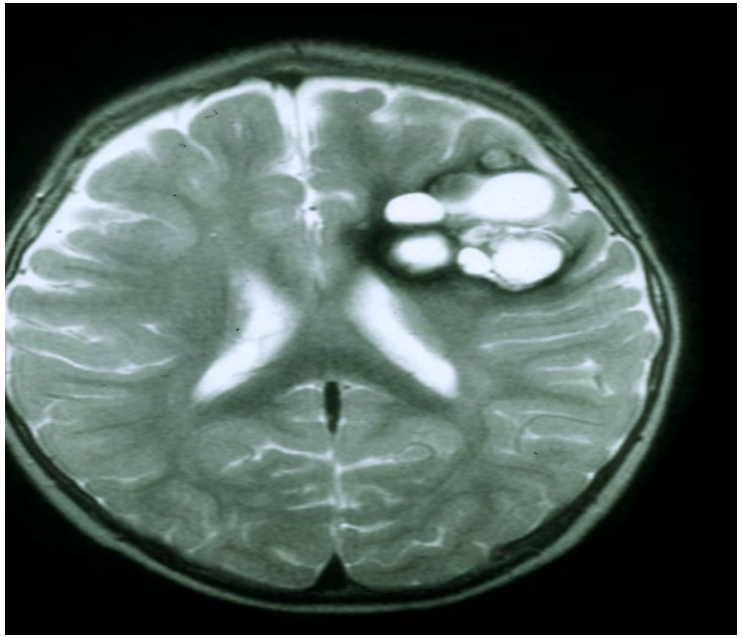
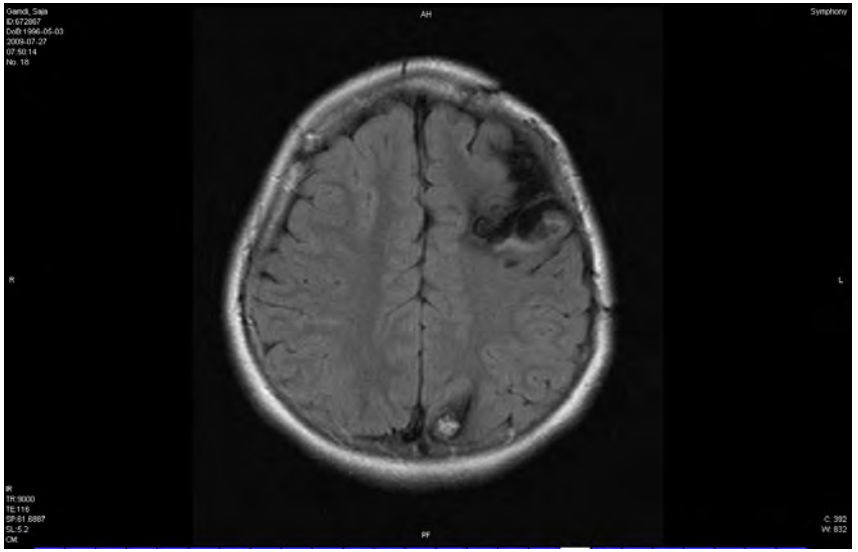


Preoperative MRI shows large cavernous angioma and with areas of bleeding



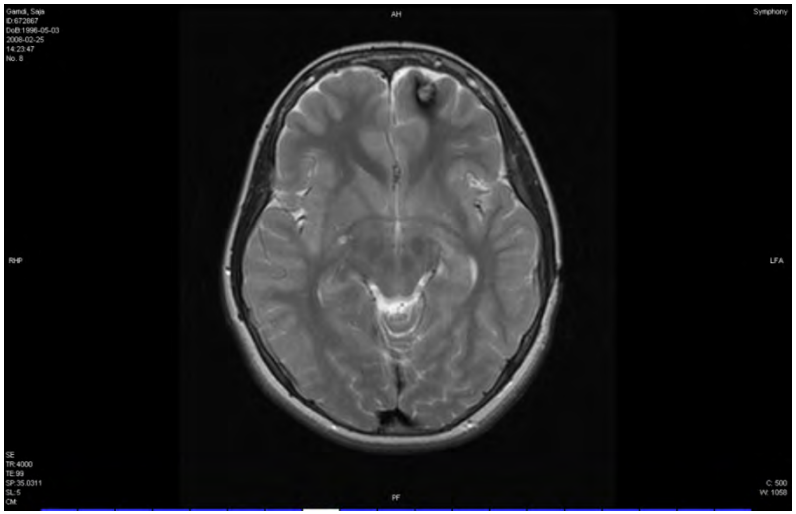
- Post operative MRI, shows total removal

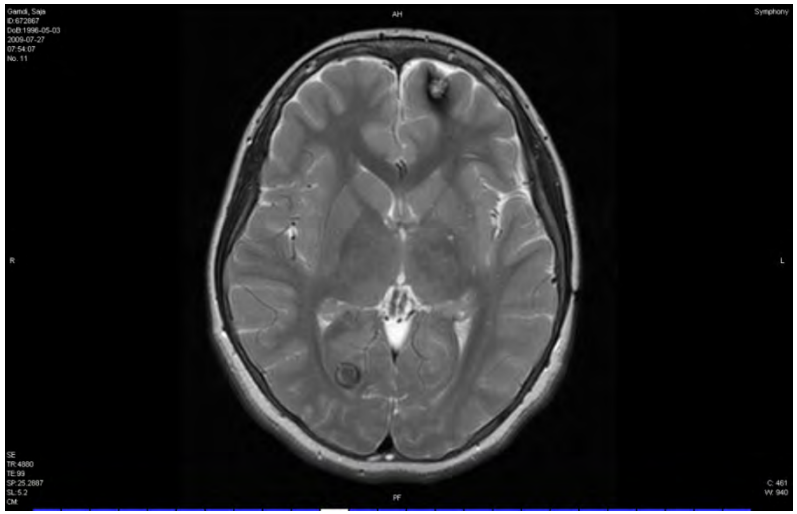


3 days post operative

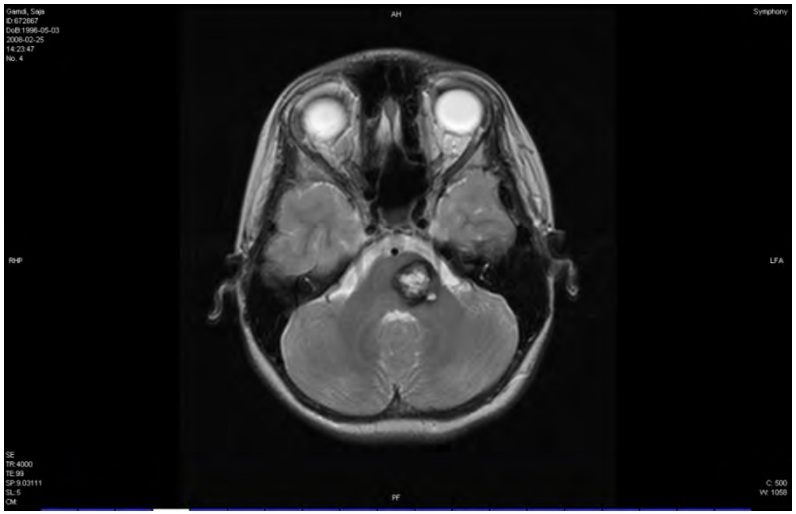
5 years later, she complained of repeated attacks of epilepsy, so MRI was done and shows 2 new cavernous angiomas in the left frontal and in the right occipital area.

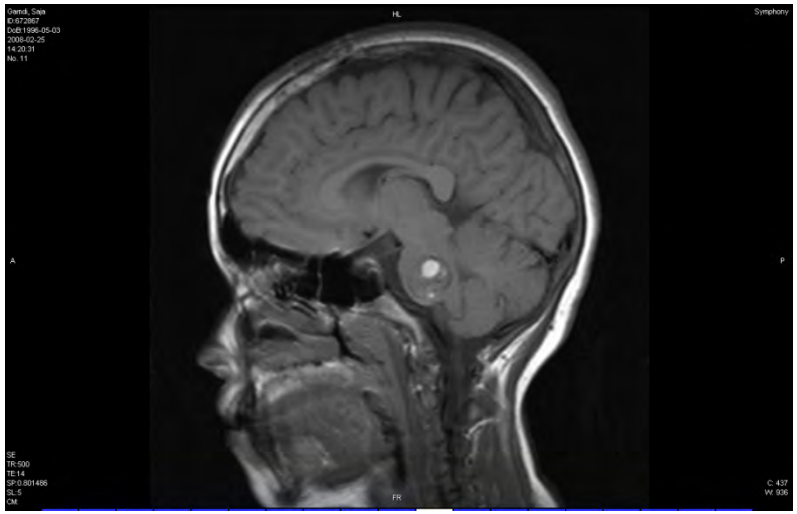
She was operated for second time to remove the frontal cavernous angioma. She made very good recovery. The right occipital cavernous angioma was followed up.



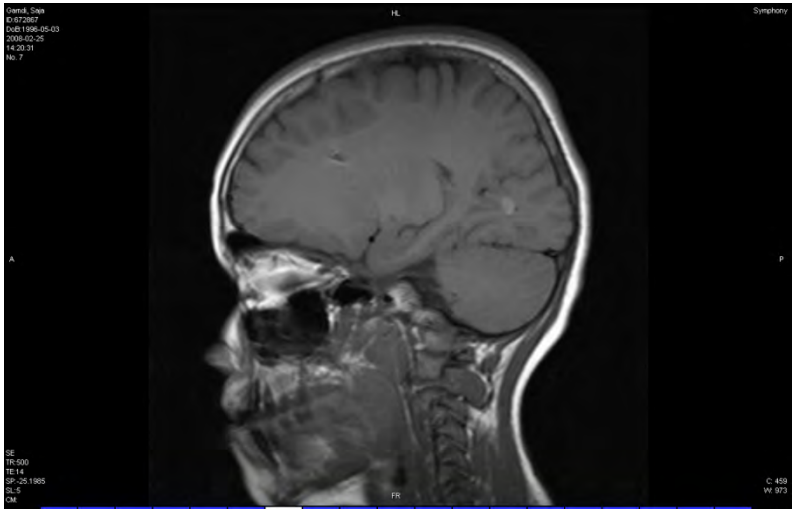


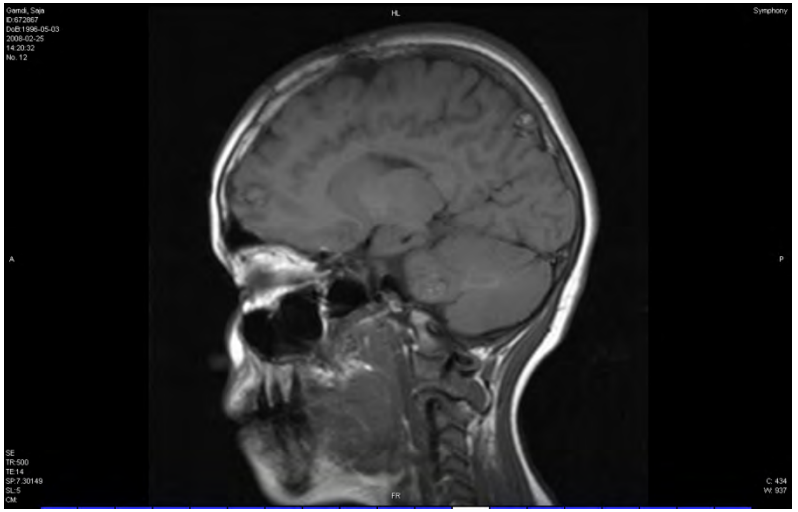
6 years later, the patient started to complaint of headache and can t walk for relatively long distance. No other neurological symptoms. MRI showed Pontine cavernous angioma which was not detected any of her 5 MRI before! She was operated for the third time for total removal of that pontine cavernous angioma. She made good recovery but mild UMN VII palsy, which has improved gradually





Parietal and occipital lesions have been discovered few years later





What to do Next?

- The patient is now 18 years old. In the final year in high school , wants to go to university.
- Epilepsy is well controlled.
- There are other 3 small cavernous angioms, in deep right occipital, in the left pariato- occipital area and deep in the left temporal.
- Patient has absolutely refused to go for surgery again.
- We decided to follow the patient up with annual MRI.

Conclusion

- Surgery is the treatment of choice for cavernous angioma
- The results of cavernous angioma surgery in brainstem is very rewarding
- The role of radiosurgery in cavernous angioma is doubtful
- Medical treatment and careful observation has role in multiple cavernous angioma


Thank you