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Title of the presentation: A rare case report on giant craniopharyngioma in adult female

Authors and Institute: DR. PRAFULLA RAMANATH KUTE,3RD YR Junior Resident
Co-author-DR.NIDHI AJAY MANDHANA,2ND YR Junior Resident
Department of Radio-diagnosis,DUPMCH,JALGAON,MAHARASHTRA

Introduction:

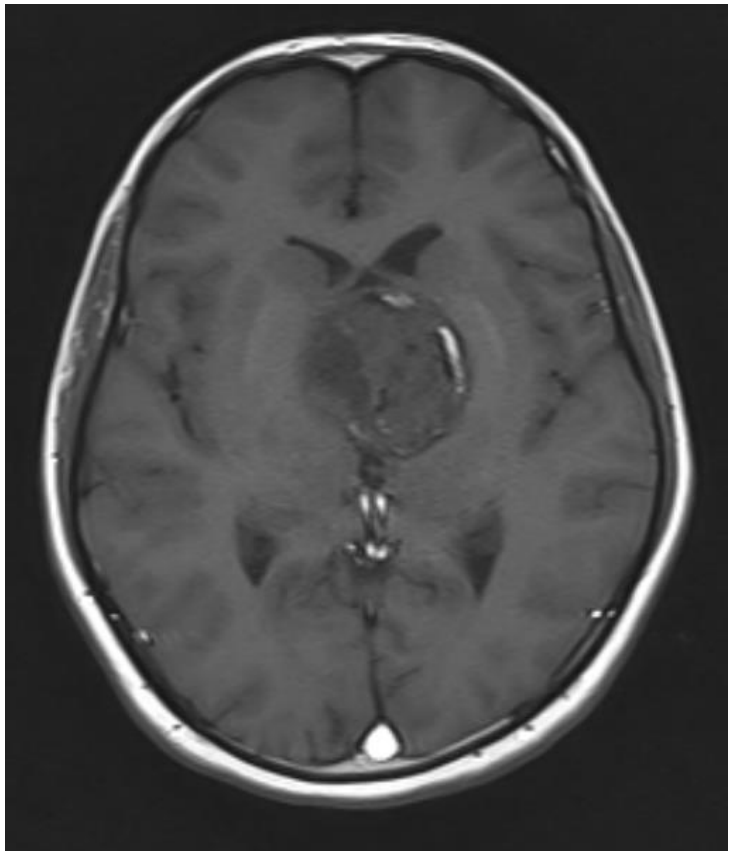
Craniopharyngiomas are a rare group of intracranial tumors that typically develop in the sellar or suprasellar region of the brain. Among these, adamantinomatous craniopharyngiomas are notably more prevalent than papillary craniopharyngiomas. The former primarily occurs in children, while the latter is predominantly found in adults.

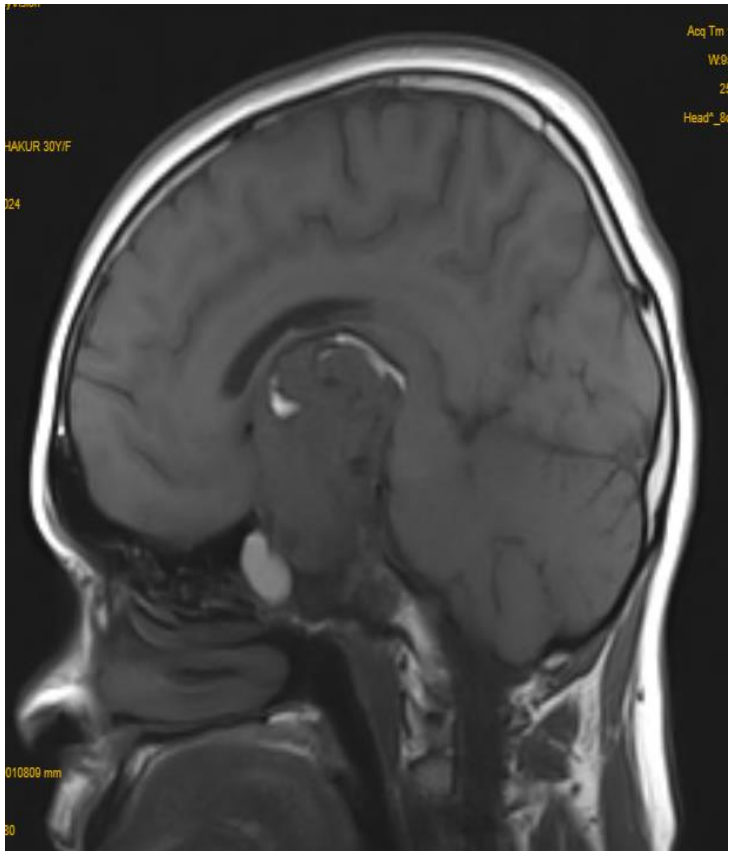
Methodology:

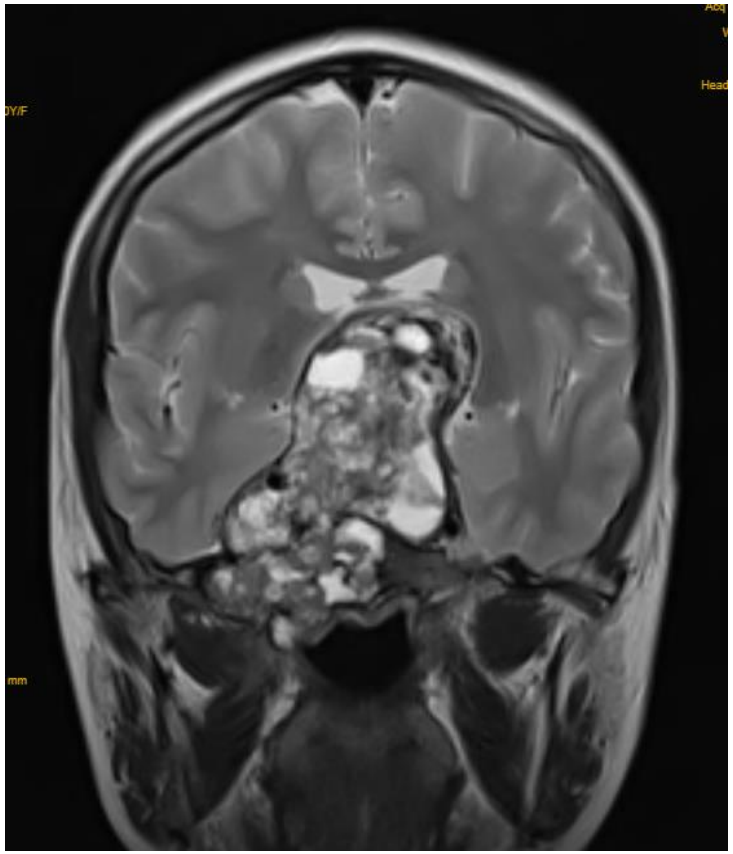
Case presentation-

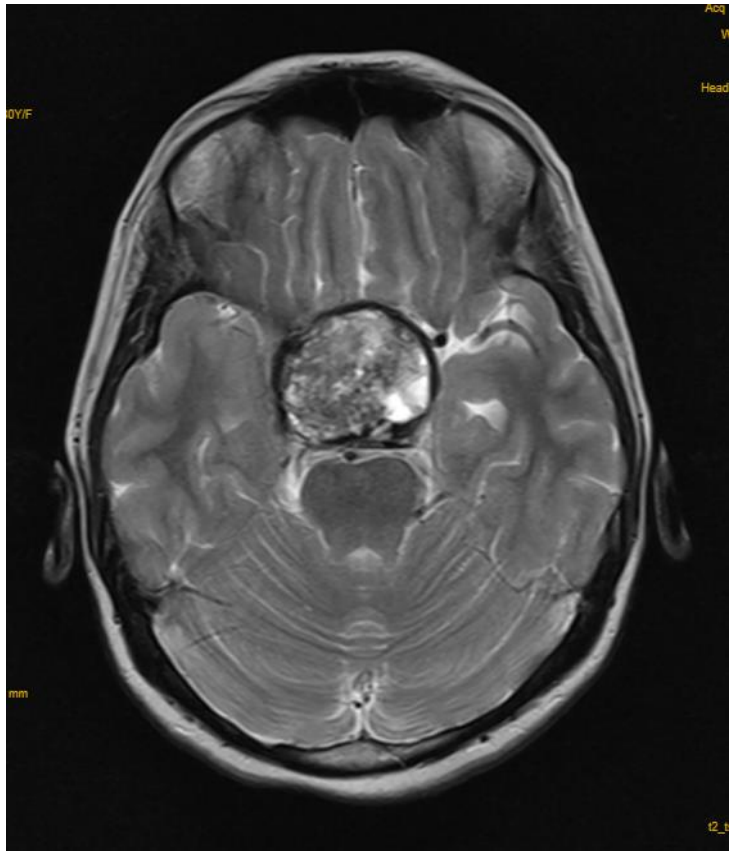
We present the case of a 30-year-old female with visual disturbances and other vague complaints. MRI scan of the brain was performed using T1 & T2 weighted sequences in multiple planes. Found to have a large lobulated well defined heterogenous extra-axial solid mass approximately measuring 3.5cmx4cmx7.5cm in the sellar and Suprasellar region with widening and complete destruction of the sella. Mineralization and enhancement was seen in the mass. Mass is extending superiorly upto the interventricular region at the level of atria of lateral ventricles and extending into the adjacent right medial temporal lobe and middle cranial fossa posteriorly. No extension seen into ventricles , there is no hydrocephalus.Anteriorly the mass was causing severe compression on orbital apices and optic nerve on both sides. There was significant bony erosion and destruction of petrous part of temporal bone, anterior part of clivus and inferolateral wall of sphenoid sinus. Right cavernous sinus was involved with complete encasement of right ICA. Left cavernous sinus was also involved with left ICA pushed laterally.On post contrast study the lesion was showing vivid enhancement of solid component.Features are suggestive of Craniopharyngioma.

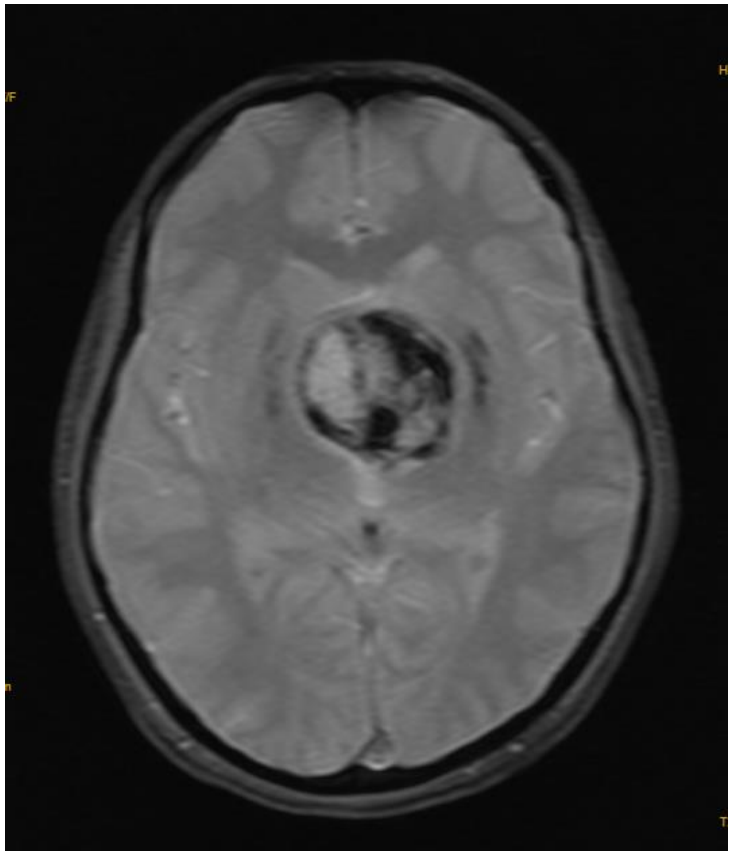
Representative images:

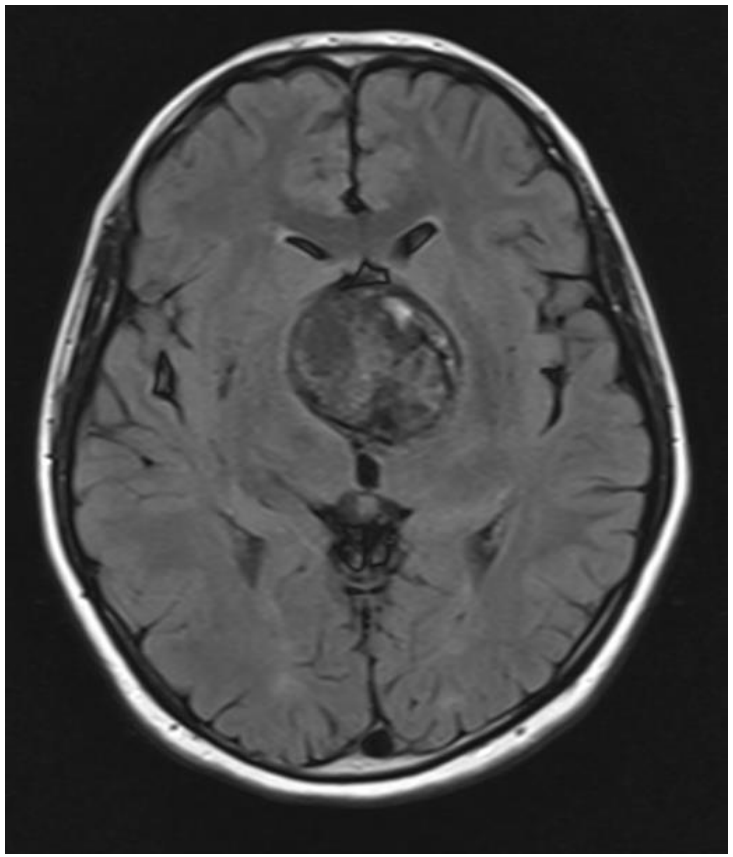


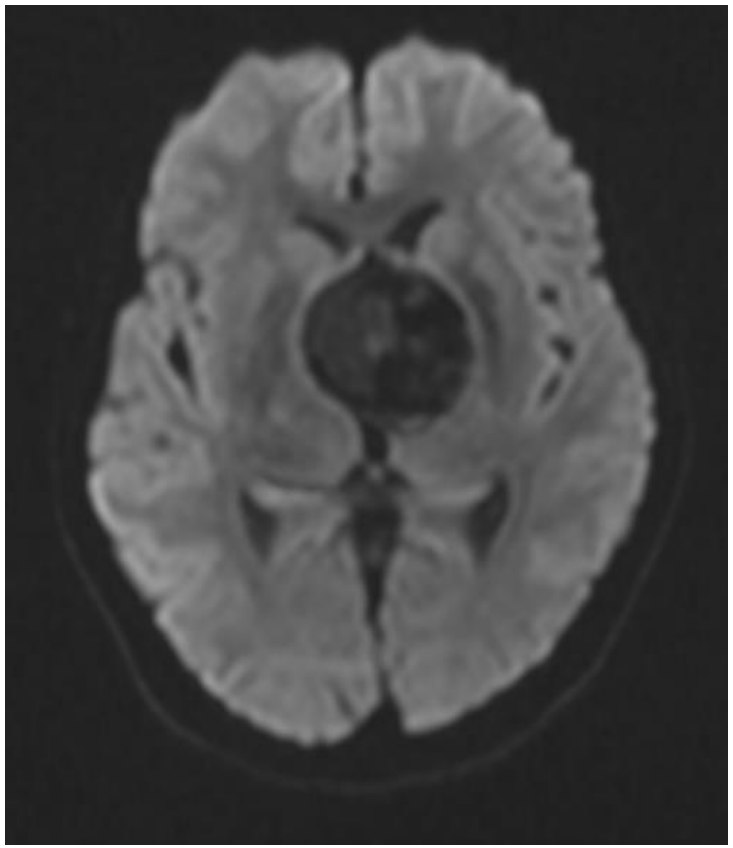


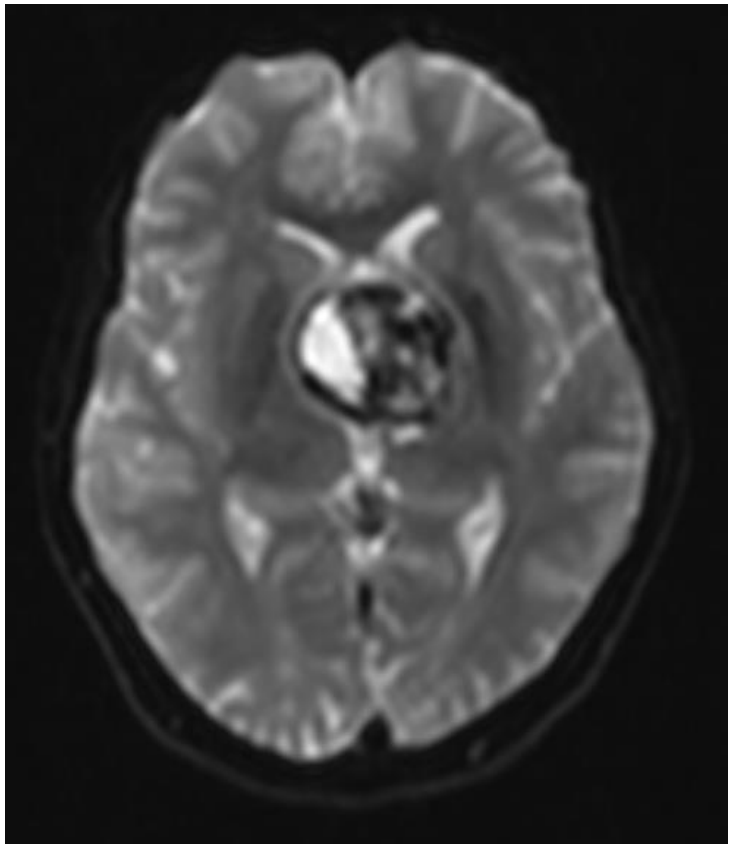












Discussion:

Craniopharyngioma is a slow-growing benign sellar and suprasellar region mass likely arising from squamous epithelial remnants of the Rathke's pouch. They represent 2–5% of all primary intracranial tumors with an incidence rate of 1–2 cases/million a year. About 30–40% of cases present during adolescence while around 25% are diagnosed in patients over age 25.[1] Craniopharyngioma is the most common non-glial neoplasm in children.

Two types of craniopharyngiomas are recognized – adamantinomatous type (90%) and papillary type (10%). Adamantinomatous craniopharyngiomas are infiltrative, multilobulated solid-cystic suprasellar masses containing viscous "machinery oil" fluid rich in cholesterol crystals. Papillary craniopharyngiomas are solid encapsulated cauliflower-like masses that do not adhere to adjacent structures.[2] The former is seen in children while the latter occurs in adults. Clinical symptoms include visual defects, pituitary and hypothalamic disorders, and symptoms of intracranial hypertension. The larger the mass is, worse is the prognosis. Seizures are commonly seen with larger masses and are thought to be due to mass effects exerted over the suprasellar and/or temporal areas, eliciting an epileptic discharge. Regardless of size, these tumors rarely undergo malignant degeneration.

Giant craniopharyngiomas, described as a tumor larger than 5–6 cm on their largest axis, are more common in children and extremely rare in adults. They are usually adamantinomatous craniopharyngiomas and can range from 5 cm to 11 cm.[3] Giant craniopharyngiomas may extend into both anterior and middle cranial fossae. Posteroinferior extension between clivus and pons down to foramen magnum is seen in exceptionally large lesions.

Microscopically, adamantinomatous craniopharyngiomas are characterized by cystic portions and stratified squamous epithelium surrounding calcified wet keratin with prominent peripheral palisading. Papillary craniopharyngioma has solid sheets of mature squamous epithelium.

CT and MRI are the imaging modalities for diagnosis of craniopharyngiomas. In CT, solid cystic portions, calcifications can be well differentiated and enhancement can be observed. MRI shows the relationship with surrounding neurovascular anatomy and whole extent of tumor into surrounding structures. Craniopharyngiomas have variable signals on T1W1 and are usually hyperintense on T2/FLAIR. Nodular or rim enhancement is seen in 90% of cases. MR spectroscopy shows a large lipid lactate peak.

Gross total resection is the best treatment option;[2] however, recurrence is a problem, especially in larger and incompletely excised lesions. Hypothalamic injury is the major risk of resection surgery. Long-time survivors are more likely to experience reduced quality of life, mostly due to morbid hypothalamic obesity. Most cases of craniopharyngioma malignant degeneration occur in patients with multiple recurrences and prior radiotherapy, it is thus recommended that patients undergo regular MRI follow-up after surgical resection to ensure that no relapse or malignant degeneration occurs.

References:

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