Original Article

COMPLICATIONS AND OUTCOME IN GIANT CRANIOPHARYNGIOMA- A CASE SERIES AND META-ANALYSIS

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Abstract:
Craniopharyngioma is a common intracranial tumour with bimodal presentation. It is more commonly seen in pediatric age group and second peak is seen in adult population (above 40). It developes from outpouching of stomadeum during embryogenesis, which involutes with time. Persistence of this outpouching leads to proliferation of cells with various deleterious effects on the nearby brain structures and also through hormonal imbalance on all over body. Many such tumours increase to gigantic proportions before the patient notices symptoms. The large sized tumours pose unique challenges during the surgery and in the post operative periods. Here in this article , we present a series of 6 such cases with intraoperative difficulties and a meta analysis of few other cases.

Keywords: craniopharyngioma, tumor, bimodal

INTRODUCTION:

Craniopharyngioma is a rare intracranial tumour with bimodal presentation, with first peak seen in 10-20 years and second peak after 40 years. It developes as an outpouching of stomodeum during embryogenesis.[1] Normally this out pouching involutes during growth of the fetus, defect in B-catenin pathway leads to proliferation of cell mass and tumour formation. Histologically, two types adamantinomatous and papillary varieties are seen. Both having different proginator cells. [2]Adamantinomatous type is more common in pediatric age group and papillary variant is more common in adult age group. It is a benign tumour but owing to its location in the brain, it presents with myriad of symptoms due to involvement of various structures in its vicinity.[3] Structures nearby are: pituitary gland, optic nerves and chiasm, hypothalamus, various vessels including the distal carotid arteries and its bifurcation. Smaller tumours (<4 cm)usually encroach these structures. And patient presents accordingly with hormonal imbalance, visual symptoms, hydrocephalus,

headache and other features of raised ICT. In cases, where the size exceeds 4 cms, it starts to extend to various other regions of the brain, including retroclival, subtemporal and into the third ventricles. In this series , we came across 6 such cases who had multicompartmental extension of the tumour. [5]Two cases had complete excision of the tumour, despite stalk preservation in one, postoperatively they had triphasic response of DI and SIADH. But has high recurrence rate most.

MATERIAL AND METHODS:

The case series was done at the neurosurgery department of the superspeciality block of SSMC Medical college, rewa. Patients were admitted directly from opd or were referred from other departments of the college. We conducted these surgeries between june 2020 to march 2023. Presenting complains were headache, diminished vision, rapidly increasing weight , stunted growth (Table 1). In these 6 patients, there was multicompartmental extension of the tumour, including clival and subtemooral extension. Complete excision was achieved in two, subtotal in the other two and decompression in the other two. Intraoperatively , these tumours pose unique challenge, different from other tumours of the brain. It obtains blood supply from the feeders which supply the surrounding structures like superior and inferior hypophyseal arteries, and multiple unnamed perforators. Dissection of the tumour not only poses a challenge to the nearby structures like visual apparatus and the hypothalamus, but extensive vascularity that these tumours have, including en passage arteries , vascular injury including injury to the major vessels like carotids is also a risk . Dissection of the tumour is more risky because of its origin from below the infundibulum; vessels get splayed over the tumour with multiple perforators going in to the tumour mass. While debulking the tumour , direct injury may be incurred to the vessels along with vasospasm due to handling, which usually do not respond to papaverine. Retraction of the brain structures is needed to look into the corners this causes additional neurological deficit depending on the part retracted. Traction on the brain structures while teasing off the tumour is another serious assault on the brain which has maximum effect if traction is done on the brainstem , thalamus , hypothalamus.

Serial no	diminished vision	stunted growth	headace	convulsions	gigantism	comatose
pt 1	+	+	+	-	-	-
pt 2	+	-	-	-	-	-
pt 3	+	-	-	-	-	-
pt 4	+	-	+	+	+	-
pt 5	-	-	+	-	-	-
pt 6	+	+	NA	+	-	+

Case one: 13-year-old boy presented to us with mild to moderate headaches, dizziness and gradual loss of vision with stunted growth and also with vomiting on and off. Examination showed diminished vision in both eyes with impaired finger counting at three distance. growth was obtunted and the child was short in height. MRI Brain was done, which showed a suprasellar lesion extending to posterior clival region. We planned to do the surgery right. Right sided pterional craniotomy was done and gross total excision of the tumour was done. Patient went into diabetes insipidus (DI) in the post operative period, followed by syndrome of inappropriate diuretic

hormone release (SIADH) and then resolved after 5days. post-operative hormone replacement was needed after discharge also.

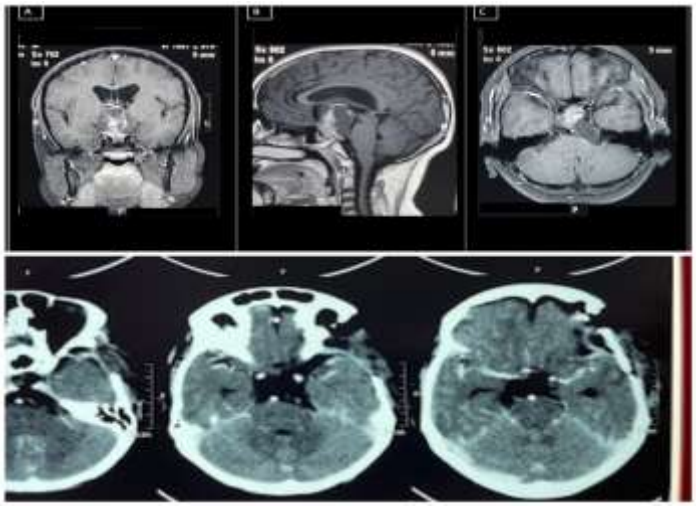


image of pt 1

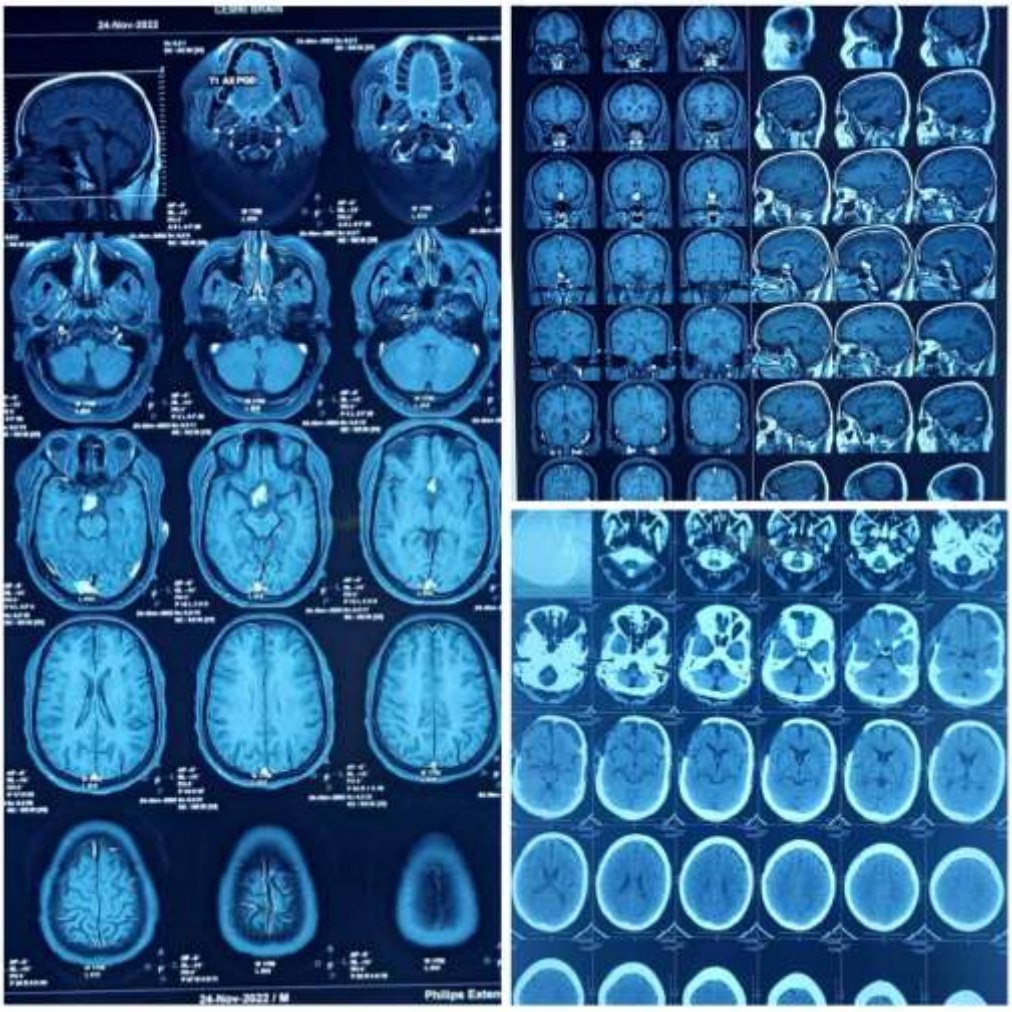


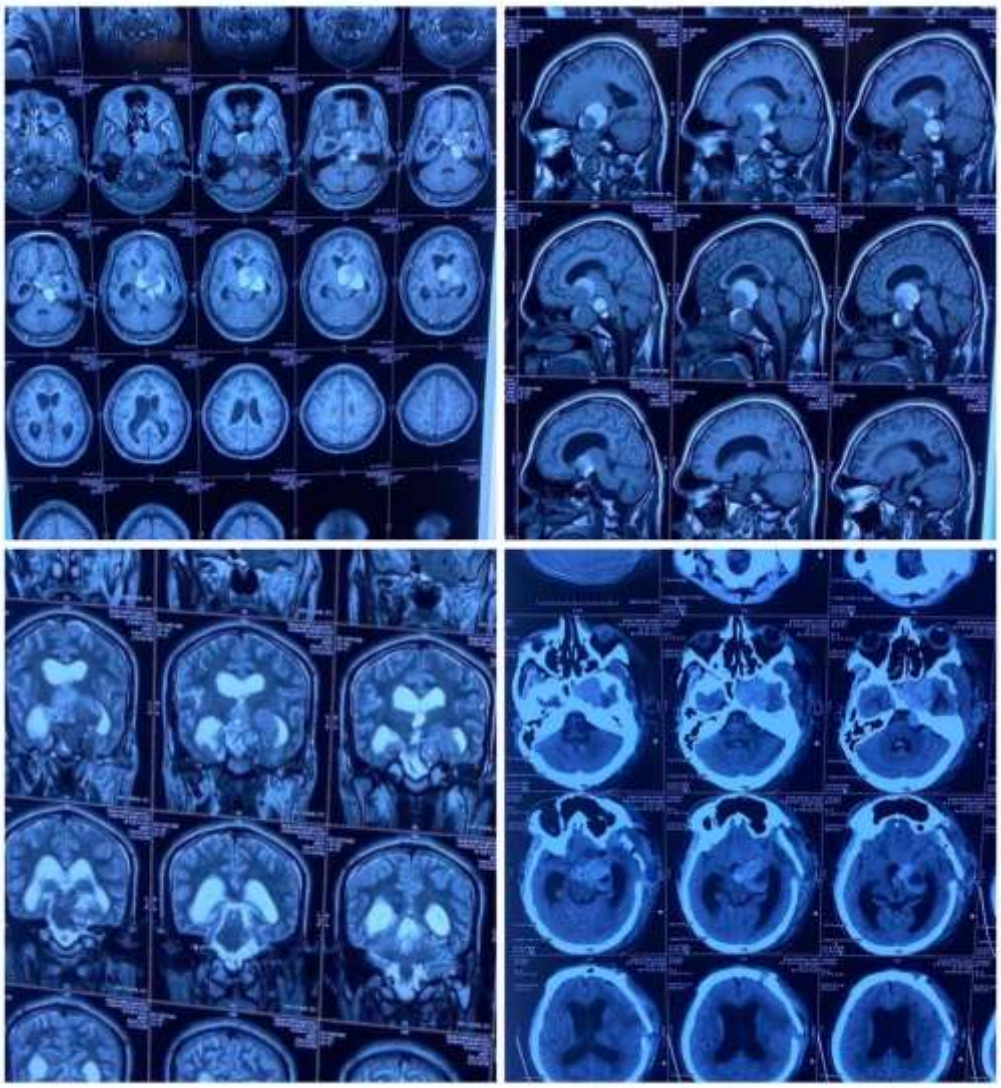
image of pt 2

Case 24 year old man presented with impaired vision, weakness, general, apathy, headaches on, and off . On examination, there was loss of vision in both the eyes to the extent of perception of light

being present in both eyes, hormonal profile was deranged :TSH and IGF were, low cortisol was low too.

MRI brain was done. It showed a giant supra seller reason with compression of the optic apparatus and the hypothalamus on right side. Pterional craniotomy and gross total excision was done. Pituitary stalk was identified and preserved.

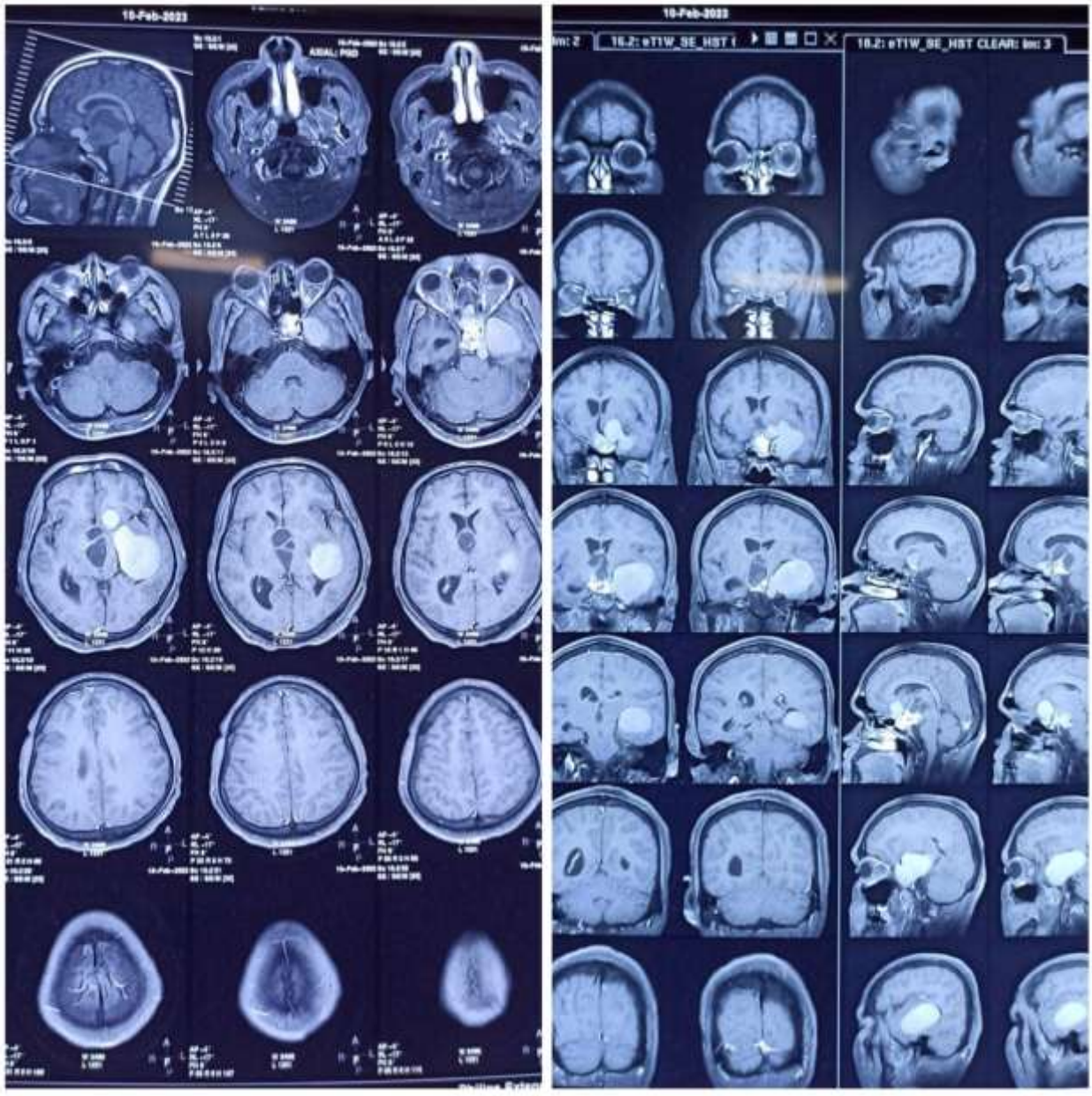
post-operatively, patient went into diabetes, insipidus (DI) for a brief period and recovered after 36 hours. However, the hormonal deficiency persisted and replacement was needed.



pre and post op images of pt 3

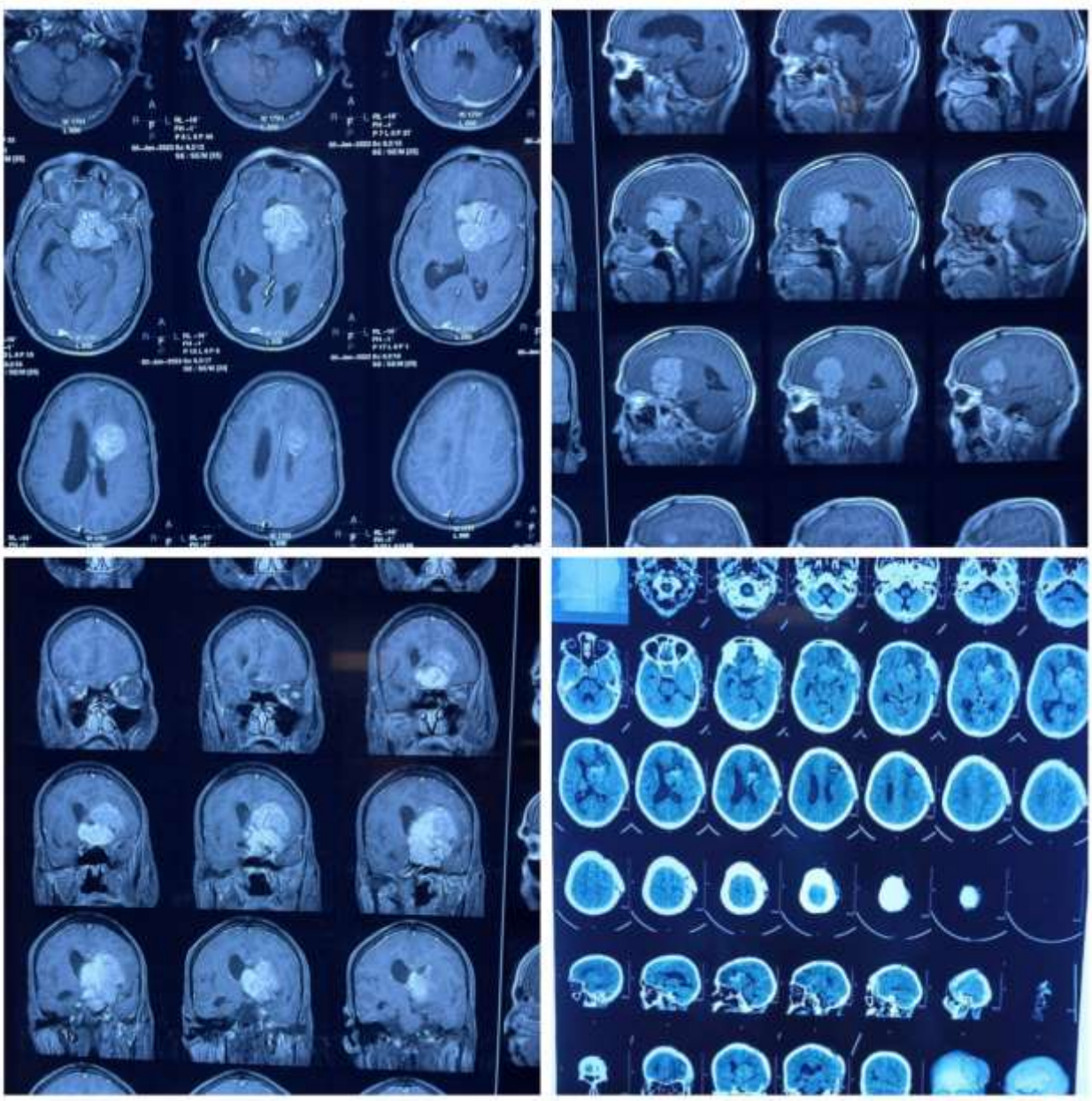
Case three: A 19 year-old boy came with headache, vomiting and loss of consciousness. Patient's relatives gave history of headache, and vomiting on and off since last 4 to 6 months. MRI showed suprasellar reason with compression of the optic apparatus, hypothalamus, third ventricle and also extending into the pre pontinen space. Patient was operated and debulking of the tumour was done. mostly, in the area surrounding the visual apparatus. Other corridors were left aside due to Injury to one carotid artery andmoderate blood loss. Patient also had hydrocephalus,so EVD was placed. Patient showed improvement in the post operative period And did not develop DI. Vision also improved. In the second sitting after general improvement, partial debulking was done from the hypothalamic and clival areas. However, due to adherence to the brainstem and hypothalamus complete resection was not possible. In the post operative period,patient developed right sided

weakness, left sided ptosis, (most likely due to prolonged retraction on the left cerebral peduncle) . Patient remained ventilator dependent and expired after 15 days.



images of patient no 4

Case number 4: A 18-year-old boy presented with repeated bouts of headache with seizure episodes and he had no other complain. Examination showed papilloedema .
MRI was done and it showed a giant craniopharyngioma with multiple compartmental spread involving left subtemporal area, hypothalamus , third ventricle and clivus.
patient under pterional craniotomy. decompression of the cystic portion with subtotal ressection of the tumour was done. patient remained hormone dependent had DI in the post operative period.



images of patient 5

Case number five : A 70-year-old man presented with headache
On examination, he was found to have papilloedema.
MRI brain was done, It showed a supra sellar space occupying lesion (SOL), enhancing with contrast, extending to occupy both the carotid arteries, optic nerves and the optic chasm and also extending into the right parietal lobe.[6] Pterional craniotomy was done and decompression of the optic nerves was done. Tumour appeared highly vascular and was plastered over the carotids. In view of the above findings and a lack of a plane in between the tumour and the major blood vessels, a trans cortical route was also taken and some decompression was achieved in the same sitting. patient had some relief in the symptoms.[7] A redo surgery was being planned, but due to the patient's age, patient's elatives were not keen for second surgery and decided to take him for radiotherapy.[8]

Complications

Sr no	optic pathway injury	vascular injury	hypothalamic injury	post op hydrocephalus	hormonal deficiency	death
pt 1	-	-	+	-	+	-
pt 2	-	-	-	-	+	-
pt 3	-	+	-	-	-	+
pt 4	-	-	-	+	+	-
pt 5	-	-	-	-	-	-
pt 6	-	-	-	-	+	+

Conclusion :

If the tumour is multi compartmental, it is better to do a staged surgery. Even if possible to do in a single sitting, outcome may be unfavourable due to vasospasm , traction edema, prolonged retraction of the vital areas or brainstem and also due to subsequent hydrocephalus, the outcome may be poor.[9]

Various approaches need to be taken like transsphenoidal endoscopic, trans Sylvania transpetrosal,retrosigmoid, depending on the tumour location.[10]

Giant craniopharyngioma are a rare and unique category of tumours which can have varied presentations like visual deterioration, raise ICT, hydrocephalus, hormonal disturbances, diencephalic syndromes, seizures and many other symptoms. It may have already spread to multiple pockets in the same cranial fossa or to multiple cranial fossas including anterior, middle and posterior and surgery being the primary modality of treatment needs to be tailored and staged as per individual cases. Radiotherapy and chemotherapy are useful Post-operative adjuncts.

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