

Tumours of Third Ventricle: Diagnosis and Treatment Techniques and Strategies

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SUMMARY

Tumours in and around IIIrd Ventricular region have always presented a challenge because of difficulty in diagnosis and treatment, the anatomy and pathology of this region is complex. Clinically symptoms and signs are related to either increased intracranial pressure or infiltration or compression of nuclei and pathways. MRI is the investigation of choice. The management includes surgery, irradiation and chemotherapy. Operative approaches from every conceivable angle have been developed but choice of the right route and good microsurgical technique has made surgery safe.

INTRODUCTION

Tumours in and around IIIrd ventricular region have raised an interest out of proportion to their frequency because they are difficult to diagnose and treat. The challenge presented by these tumours is due to their central position in the cranial cavity and difficulty in approach¹. The first report of successful removal of tumour from IIIrd ventricular region was published by Openheim and Krause in 1913².

Dandy perfected the transcallosal approach to tumours of IIIrd ventricular region³. More recent advances in development of operative microscope, microsurgical technique and anesthesia have considerably reduced the mortality.

Improved tumour definition has not been limited since modern neurodiagnostic tools have provided sophisticated techniques for identifying and localizing the tumours and their boundaries while reducing the risk and invasiveness of these procedures⁴.

Anatomical considerations

The anatomy of the IIIrd ventricle is very complex owing to the surrounding vital vascular and neural structures with important functions, which makes the tumours in this region difficult to expose and remove⁵.

The third ventricle is located in the centre of head below corpus callosum and body of lateral ventricle; above the sella turcica, pituitary gland and midbrain; and between the cerebral hemispheres, thalami and the walls of hypothalamus. It is intimately related to the circle of Willis and its branches and the vein of Galen and its tributaries.

Pathology

The region of the third ventricle has a large variety of tissues and structures. The pathological processes in the area encompass a wide spectrum of pathological entities. Lesions are primarily intraventricular like colloids cysts and craniopharyngiomas, in the walls like astrocytomas, pineal tumours and germ cell tumours in the pineal region^{6,7} and those that are primarily extra-axial in basal locations may compress or invade the walls of the third ventricle. Ependymal cysts and Ependymomas are also found in adults and children⁸.

Clinical behavior

The clinical presentation of tumours in this region to a large extent are dependent upon the size of the lesion and whether it invades the surrounding neural structures (gliomas) or grows and expands the ventricular cavity (colloid cyst).

1. Related to Increased Intracranial pressure (ICP)

Symptoms of increased ICP are seen with lesion at the Foramen of Monro or lesion that grow into third ventricle ultimately obstructing it and include headache, vomiting and altered mental state⁹.

2. Related to infiltration or compression

Infiltration or compression of nuclei and pathways lead to specific symptoms. Thalamic invasion leads to hemianaesthesia. Invasion of internal capsule may produce hemiparesis or hemiplegia. Basal ganglia involvement result in extrapyramidal syndrome. Hypothalamic invasion may cause disturbances of water regulation, body temperature, somnolence and weight gain. Tumours compressing quadrigeminal plate lead to Parinaud's syndrome. Short term memory problems may develop by invasion of mamillothalamic pathways.

Diagnostic work up

Skull radiography and air or positive contrast ventriculography were once the back bone of diagnosis of third ventricle which has been relegated to only a historical position. The diagnosis and management of these lesions have now been revolutionized initially by computed tomography and now Magnetic resonance imaging (MRI)¹⁰⁻¹². MRI to its incomparable ability to image soft tissue by means of its unique feature of increased sensitivity and tissue discrimination compared to CT, direct multiplanar axial, sagittal and coronal imaging capability and lack of the beam hardening artifact encountered with posterior fossa CT^{13,14} is the investigation of choice.

Angiography is helpful in demonstrating the nature or lack of vascularity of a mass. It is being replaced by MR angiography and is also performed to differentiate vein of Galen aneurysm, angioma and meningioma¹⁵.

Tumour markers alpha-fetoproteins and beta subunit of Human chorionic gonadotropin are useful in evaluating sub groups of pineal tumours. In choriocarcinoma, elevation of serum and CSF beta-HCG levels always occurs. Embryonal cell carcinoma, Endodermal sinus tumours and occasionally immature teratomas secrete AFP. Elevated levels of plasma or CSF beta-HCG and AFP

have also been found in patients with pineoblastomas¹⁶. Tumour markers are usually sensitive indicators in diagnosis during follow-up to indicate total removal or later recurrence.

Plasma melatonin level as marker is useful for neoplasms and other lesions of pineal gland. It is secreted in a circadian rhythm with high serum levels during the daytime. After pineal tumour resection serum melatonin levels may serve to demonstrate complete pinealectomy. Experimental studies for other markers include S-antigen for pineal tumours, placental alkaline phosphatase for primary intracranial germinomas¹⁷.

MANAGEMENT

A) Surgery

Primary objectives in management of masses affecting the IIIrd ventricular chamber include:

1. The wide variety of pathology and lack of specificity of tumour markers makes histological evaluation mandatory. Biopsy may be achieved by a stereotactic or endoscopic approach.
2. Achieving maximum lesion excision for the benign lesions to normalize CSF dynamics and decompressing focal pressure effects for the malignant lesions.

As third ventricle is located in geometric centre of intracranial cavity, so operative approaches from every conceivable angle have been developed.

A number of surgical approaches are used, the most appropriate for a given lesion depends on exact location as determined by neuro imaging studies, size of the lesion and the clinical status of the patient.

Approaches for anterior third ventricular tumours

In 1930 Dandy first used the transcortical approach. It requires incision of cerebral cortex to reach the lateral ventricle. Transforaminal approach described by Van Wagenen in 1931 and Sano described subchoroidal transvelem interpositum approach^{18,19}.

The most frequent complications of these approaches are post operative seizures, the frequency of which is as high as 27%²⁰. Fornix section results in memory loss and confusion with mutism were also reported²¹.

Transcallosal approach permits the surgeon to enter IIIrd ventricle without cortical incision and ventriculomegaly is not a necessary prerequisite for this approach²².

Memory disturbances have been a frequent topic of concern²³. Fornix can be sectioned inferiorly to enlarge the foramen and many authors feel that section of a single fornix should not be associated with memory loss²⁴⁻²⁶.

Approaches for posterior third ventricular tumours

Occipital transtentorial approach was first accomplished by Foerster in 1928 and described in detail by Poppen in 1966^{27,28}. This opened the avenue for most widely used approach to lesions in posterior third ventricle. Damage to veins of deep venous system constitutes the major cause of morbidity and mortality.

Ausman in 1984 reported successful use of three quarter prone position with operative side down which decreases the risk of air embolism and minimizes occipital lobe retraction²⁹.

Infratentorial supracerebellar approach

Infratentorial supracerebellar approach was first described by Krause in 1913 and revived by Stein in 1971³⁰. The wide suboccipital craniotomy with opening of the torcula and transverse sinuses is made with patient in a sitting position. Complications include damage to major veins and CSF leaks.

Minimal invasive techniques

Stereotactic approach utilizes computed tomography or magnetic resonance imaging. Lesion can be biopsied, drained or removed depending on their consistency. On occasion radioactive substances can be stereotactically directed into cysts³¹. Complications include intracranial haematoma, seizure and abscess formation in 8%^{32,33}.

Endoscopic procedure was first performed by Power in 1983 using rigid endoscope. With endoscopic surgery operating time on average appears to be half that required for open microsurgical approaches. Stay is reduced to half and patients return to work in half the time as compared with open surgical techniques³⁴.

B) Radiation

Initial irradiation has several advantages:-

1. CT has made it possible to identify

radiosensitive tumours following small dose of radiation and thus save the morbidity and mortality of an operative procedure.

2. Most patients operated require post operative irradiation.

The recommendation for 5000 to 5500 rads is based on retrospective analysis of several series. Other therapeutic modalities are stereotactic radiosurgery and interstitial irradiation³⁵.

C) Chemotherapy

In 1977 De Tribolet with Barrelet reported the successful use of chemotherapy in a patient with a large pineal tumour³⁶. Combination of cisplatin, bleomycin vincristine and ACNU, vincristine with radiotherapy and high dose cyclophosphamide were used^{37,38}. Chemotherapy is used first in cases of recurrences after irradiation alone, secondly in association with radiotherapy as a primary treatment for embryonal carcinomas, germinomas and other tumours with poor prognosis. Chemotherapy has proven to be effective in 85 per cent of germinomas. In many centres as soon as germ cell tumour diagnosis is made, chemotherapy is advocated as first line treatment. This has the advantage of sparing the hypothalamus which is very sensitive to radiotherapy. It is taking on an increasingly important role in the treatment of pineal region tumours for non-germinomatous germ cell tumours and it should be considered as an adjuvant to operation plus irradiation^{39,40}

CONCLUSION

The management of tumours in and around third ventricle depends mostly on their histologic type and includes surgery, irradiation and chemotherapy and this multimodality approach has markedly decreased the morbidity and mortality.

Surgery of these tumours requires a solid knowledge of anatomical boundaries and landmarks, versatility in the choice of the right route, good microsurgical techniques, meticulousity and lot of patience since the field is relatively confined.

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