

SPECTRUM OF POSTERIOR CRANIAL FOSSA SPACE OCCUPYING LESIONS- OUR EXPERIENCE AT A TERTIARY CARE CENTRE

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ABSTRACT

BACKGROUND

Posterior cranial fossa is located between the Foramen Magnum and Tentorium Cerebelli. It contains cerebellum, pons and medulla oblongata. Tumours of posterior cranial fossa can affect any of the above structures and can lead to pressure symptoms, neurological deficits or sometimes even death. This study has been conducted at our tertiary care centre.

Aims and Objectives- To know the usefulness of intraoperative squash cytology and its diagnostic accuracy in differentiating posterior fossa lesions in correlation with clinical, radiological and histopathological diagnosis. Newer diagnostic modalities like MRI help in detecting these tumours early. Squash cytology and histopathological study diagnose these lesions accurately, which may benefit the patient for further treatment.

MATERIALS AND METHODS

This is a retrospective descriptive study. A total of 28 cases of posterior cranial fossa lesions were analysed with squash cytology and histopathology correlation.

RESULTS

Among the 28 cases analysed, 27 cases (96.4%) showed squash cytology with histopathologic correlation. A case of ependymoma was misdiagnosed as embryonal tumour/ medulloblastoma on squash cytology. Male preponderance was noted. The commonest age group was first decade in this study. Cerebellopontine angle was the most common site of the tumours in our study.

CONCLUSION

Analysing the posterior fossa tumours by squash cytology along with radioimaging in tumour localising and patient's demographics helped in correct histopathological diagnosis for further management and follow-up.

KEY WORDS

Posterior Cranial Fossa SOLs, CP Angle, Squash Cytology, Meningioma, Schwannoma, Prognosis.

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BACKGROUND

Intraoperative diagnosis of posterior fossa Space Occupying Lesion (SOL) by squash cytology is extremely helpful in diagnosing the lesions. It can give a Neurosurgeon proper idea and fairly accurate diagnosis of the tumour. It can be useful to reconsider the lesion and can change the mode of surgery. The spectrum of lesions in this site is varied and differs in a child and adult. Gliomas and embryonal tumours are common in children.⁽¹⁾ (Fig. 1) Posterior fossa metastasis is the most common malignancy in adults. Gliomas and lymphomas can occur and are located in supratentorial compartment. Loss of chromosome-22 is the most frequent genetic alteration in Ependymoma. Epidermoid and dermoid cysts are also common and account for 0.3 - 1% of all intracranial tumours. Haemangioblastoma represent 2% of benign vascular tumours.⁽²⁾

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The study was undertaken to evaluate the various tumours in posterior cranial fossa, their incidence, age and sex distribution, location and evaluate the correlation between clinical, radiological and squash cytological and histopathological diagnosis. Prognosis aspects were also taken into consideration.⁽³⁾

Neurological morbidity is significant with posterior fossa brain tumours. Most common clinical presentations are headache, hearing loss, vomiting, giddiness, seizures and visual disturbances. Headache being the commonest. Sudden unilateral hearing loss is seen in CP angle lesions. Our study also showed similar comparable history. They often present with manifestation of raised intracranial pressure.⁽⁴⁾

Aims and Objectives

To know the usefulness of intraoperative squash cytology and its diagnostic accuracy in differentiating posterior fossa lesions in correlation with clinical, radiological and histopathological diagnosis. Newer diagnostic modalities like MRI help in detecting these tumours early. Squash cytology and histopathological study diagnose accurately these lesions, which could benefit the patient for further treatment.

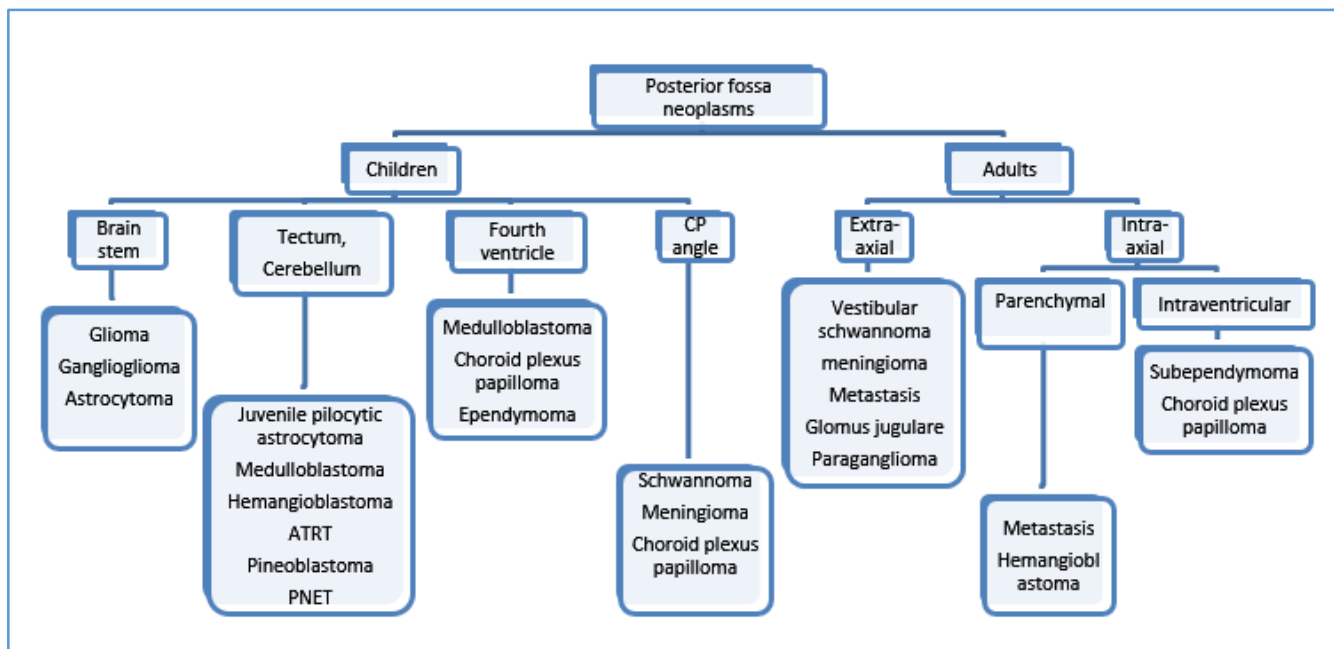


Figure 1. Classification and Distribution of Posterior Fossa SOLs

MATERIALS AND METHODS

A retrospective descriptive study of 28 cases of resected posterior fossa SOL performed in our tertiary care centre for a period of 18 months from January 2017 to June 2018 were included. Squash smears were prepared immediately from the samples, which were sent in isotonic saline and were subsequently stained by Haematoxylin and Eosin and toluidine blue stains. Cytomorphological features were recorded and left-over sample sent for histopathological examination. Routine processing and paraffin embedding was done. Immunohistochemistry was also done in selective cases.

RESULTS

A total of 113 cases of CNS intracranial SOLs squash samples were received in the study period, out of which 28 cases (21.8%) were posterior fossa tumours.

Clinical Features

Headache due to Hydrocephalus is the most common symptom seen in 70% of the cases, radiculopathy with neurological deficit in 20% cases, remaining cases had vertigo and nausea.

Gender Distribution

Males were more commonly involved in a ratio of 4: 3. There were 16 males (57%) and 12 females (43%) (Fig. 2).

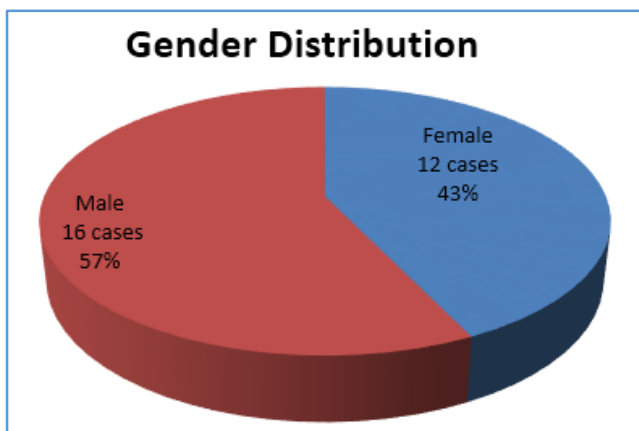


Figure 2. Gender distribution of Posterior Fossa Tumours

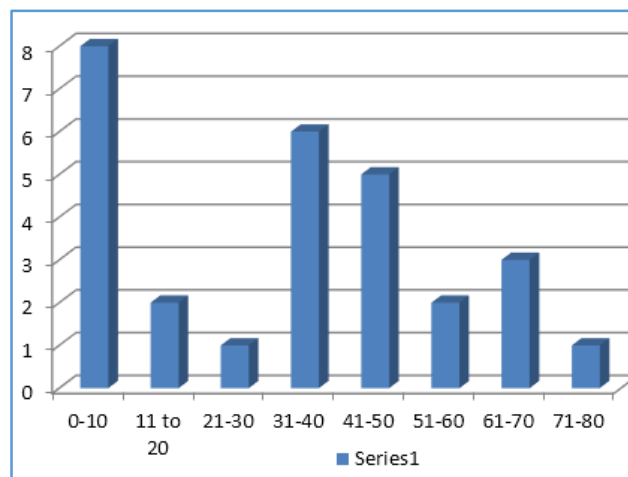


Figure 3. Age Distribution

Age Distribution

Most common age group was the first decade (8 cases, 28%) followed by 4th decade (6 cases, 21%). Rest of the cases were distributed amongst 5th and 6th decade (Fig. 3).

Distribution of Tumours

Most of the lesions occurred in the cerebellopontine angle of posterior fossa, 14 cases, 50%, followed by Cerebellum 13 cases 46% and one case of brain stem 4% (Fig. 4).

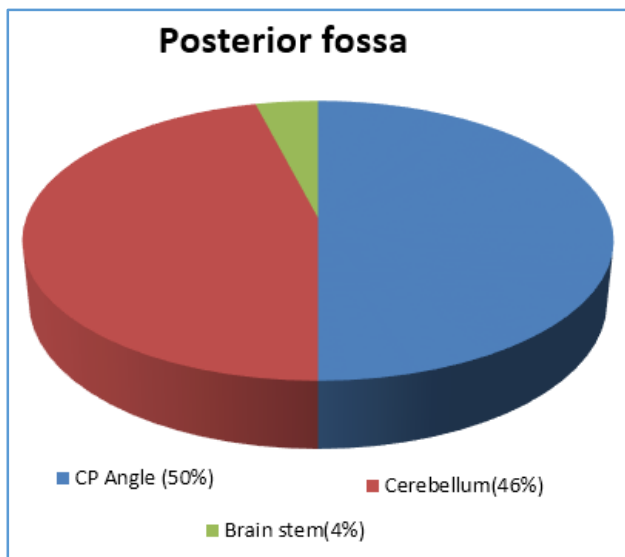


Figure 4. Distribution of Lesions

Most common histologic type was Schwannomas (9 cases 32%) followed by medulloblastomas and meningiomas. We encountered one case of Atypical Teratoid Rhabdoid Tumour in a 6-year-old male child.

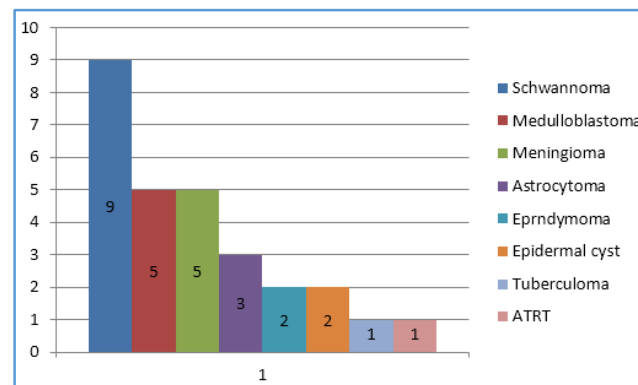


Figure 5. Histologic types of Posterior Fossa SOL

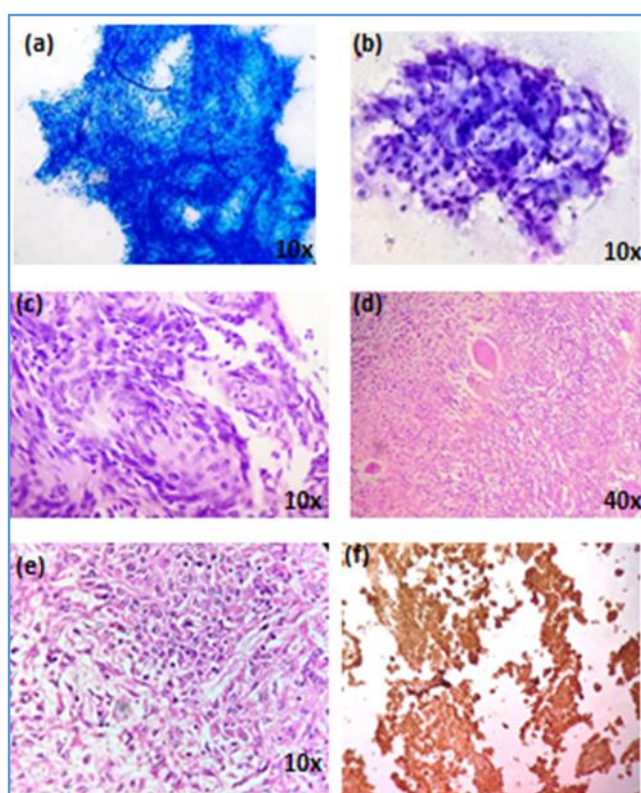


Figure 6(a) 10x, Toluidine Blue Stain, Squash Cytosmear shows cohesive fragments of interlacing bundles of Schwannian cells and Endothelial cells, (b) 10x, Toluidine Blue Stain, Squash Cytosmear shows Meningothelial Cells arranged in Whorls, (c) 40x, Haematoxylin and Eosin Stain, Histopathological Section with Meningothelial Cells in Whorls and Intranuclear Inclusions, (d) 10x, Haematoxylin and Eosin Stain, Histopathological Section of Tuberculoma with Caseating Granulomas, (e) 40x, Haematoxylin and Eosin Stain, Atypical Teratoid Rhabdoid Tumour-Rhabdoid Cells with Abundant Eosinophilic Cytoplasm, (f) Immunohistochemistry- EMA (BioGenex, Clone E-29), Ependymoma: Diffuse Membrane and Cytoplasmic Positivity of Ependymal Cells.

Cyto-Histo Discrepancy

Amongst 28 cases 27 cases (96.4%) showed histopathologic correlation, while (1 case, 3.7%) showed discrepancy. The case diagnosed as embryonal tumour/ medulloblastoma on squash cytology in a 7 years old female child was proved to be ependymoma on immunohistochemical staining.

Prognostic Outcomes

Recurrence was seen in 3 cases (12%). All of them had the primary tumour at CP angle.

DISCUSSION

The posterior cranial fossa of brain is the deepest and most capacious of the 3 cranial fossae. It houses the infratentorial brain- cerebellum, pons, cerebellopontine angle and medulla oblongata. The foramen magnum is located centrally and inferiorly in the posterior fossa and surrounded by deep grooves containing the transverse sinuses and sigmoid sinuses. Bounded by the posterior surface of petrous temporal bone anteriorly, occipital bone posteriorly and squamous and mastoid bone laterally. The brainstem is the portion of the brain connecting the cerebral hemispheres with the spinal cord. It contains the midbrain, pons and medulla oblongata. The brainstem is partially obscured by the cerebral hemispheres and cerebellum.⁽⁵⁾

The most common site for posterior fossa tumours is cerebellum. The lesions are juvenile pilocytic astrocytoma, medulloblastoma, ependymoma, ATRT, haemangioblastoma and metastasis. Brain stem lesions are gliomas and gangliogliomas. CP angle tumours are vestibular schwannoma, meningiomas, epidermoid cyst and arachnoid cyst.⁽⁶⁾

Commonest tumours are Schwannomas, originate from acoustic nerve and constitute 8% of all primary intracranial tumours. Of the 28 cases in our study there were 9 cases of Schwannomas, more than a third of all tumours, in the age group of 3 - 7 decades with a slight female preponderance of 1: 1.25. CP angle is the common site. Brain stem location is very rare. Solid and cystic variants are noted. Spinal intramedullary locations are also more common.⁽⁷⁾ Schwannomas are benign tumours, less than 1% tumours

turn malignant. Histologically, the tumour consists of cellular Antoni A and acellular Antoni B areas and Verocay bodies.

Late recurrences with parotid gland involvement is very rare following irradiation therapy.⁽⁸⁾

Medulloblastomas are the second most common tumours along with Meningiomas in our study 5 cases, 18%. All the cases were seen in children. Medulloblastomas are seen exclusively in posterior fossa. They are non-invasive, fast growing tumours and spread down the spinal canal via CSF drop metastasis.

Signs and symptoms are due to raised intracranial pressure due to block of 4th ventricle. They have repeated attacks of vomiting and headache treated as gastrointestinal disease. Symptoms antedate diagnosis by 1 - 4 months. Medulloblastomas develop from cerebellar stem cells, adjacent to 4th ventricle between brain stem and cerebellum. The tumour is cellular and contains small round cells with scant cytoplasm forming both true and pseudorosettes. They have a better about 50 - 60% survival following treatment in children than in young adults 40%.

We had 5 cases of meningioma, all in adults between 4th and 7th decade. Posterior fossa meningiomas are slow growing, constitute 20% of all intracranial meningiomas. Frequent encroachment of neural and vascular structures is common. CP angle meningiomas and petroclival sites are favoured. Microsurgical excision is the main treatment. Recurrences are frequent and with poor prognosis.⁽⁹⁾

Astrocytomas are next common 4 cases, 2 in children under 10 yrs. with pilocytic variant, 2 in adults aged 35 yrs., female with pilocytic variant, another in a 60 yrs. male diagnosed as low-grade glioma on squash and as astrocytoma on histopathology. GBMs constitute 50% of all primary intracranial tumours in adults with peak incidence around the sixth decade. Infratentorial GBMs are rare about 1%. Common differential diagnosis is metastasis.⁽¹⁰⁾

Ependymomas is the third most common posterior fossa tumour in childhood after medulloblastoma and glioma. In children the location is intracranial, while in adults it is spinal. The common location for intracranial tumours is fourth ventricle. Incomplete resection can lead to recurrence and dismal outcome. Local radiation is recommended. Spinal cord may be the site for recurrences.⁽¹¹⁾ Witt and Colleagues described two distinct subgroups in ependymomas. Group-A of lower age and lateral tumour position with serious prognosis. Group-B older age group and midline tumours.⁽¹²⁾

Epidermal cysts are present at 20 - 40 yrs. with pressure symptoms, mainly headache. We had 2 cases aged 45 and 75 yrs. Intracranial epidermal cysts are congenital inclusion cysts formed during neural tube closure. They make .25 - 1% of intracranial tumours.

Tuberculoma- one case in a four-year male child was seen in our study. The incidence of intracranial tuberculoma in recent times is on rise, because of increased incidence of AIDS. Typical epithelioid cells with Langerhans giant cells were seen on squash.

Atypical teratoid/ rhabdoid tumour was first described in 1987 by Rorke and Colleagues. Histologically resembles rhabdoid tumour of kidney. WHO in 1993 classified it as embryonal grade 4 neoplasms. AT/ RT may be linked to changes in tumour suppressor genes SMARCB1 or SMARCA4. Brain and kidney tumours can occur. Mutations and deletions in the INI gene occur in most. Immunoprofile includes SALL-

4, OCT 3/4, CD 99, FLI-1, Cyclin D1, Beta catenin, p53 and p16. WT-1 genes, PLI-1 and cyclin D1 are potential prognostic markers.⁽¹³⁾

Interesting observation of Cerebellar Mutism Syndrome (CMS) was observed following surgery of posterior fossa tumours in children. Mutism is prominent and strokes, infections and ataxia being less common. Our experience also showed similar findings in few cases with paresis. Dysphagia and other cognitive deficits were observed. Literature quotes dentato-thalamo-cortical pathway damage.⁽¹⁴⁾

The posterior fossa lymphoma are also to be entertained as differential diagnosis in young adults who can present with vertigo and ataxia.⁽¹⁵⁾

Prognosis- The 5-year survival rates exceed 60% for all patients and 80% for certain good-risk individuals with posterior fossa tumours. In cases of pilocytic cerebellar astrocytoma, the 5-year survival rate exceeds 94%.⁽¹⁶⁾

CONCLUSION

Hydrocephalus was the most common symptomatic presentation in our study. Differential diagnosis of posterior fossa tumours should be considered and excluded by using tumour location, patient demographics and tumour imaging. This helps in correct histopathological diagnosis for further management. Squash smears are almost accurate, simple, safe, cost effective and rapid method of diagnosing posterior fossa SOLs. In our study, squash preparations have given extremely valuable intraoperative information in diagnosing the CNS lesions of post. fossa SOLs along with ancillary techniques.

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