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Pineal Region Tumours in Pediatric Age Group#

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Abstract

16 cases of pineal region tumours in the pediatric age group constitute the subject of this study. After thorough investigations both clinical, radiological and laboratory, they were all operated upon by a ventriculo-peritoneal shunt to divert the cerebrospinal fluid. This was followed in 15 cases by the favoured occipital transtentorial approach and in one by the supracerebellar infratentorial approach. The microsurgical bipolar suction irrigation technique was used in all cases. The aim was to decompress as much as possible of the lesion mounting to complete excision and to attain ample biopsy for tissue diagnosis. There were no surgical mortalities. The outcome was excellent in 14 cases, one surgical morbidity in a highly invasive tumour, one incident of transient visual field defect and only one mortality due to haematemesis two weeks postoperative.

Introduction

ALTHOUGH pineal region tumours are responsible for only a small percentage (0.4 to 1%) of neoplasms of the brain, yet they have raised an interest out of proportion to their frequency [1]. In the pediatric age group the incidence increases and acounts for 3 to 8%. More than 50% of all pineal tumours are found in patients under 20 years of age [2].

The pineal region is limited dorsally by the splenium of the corpus callosum and the tela choroidea, ventrally by the quadrigeminal plate, rostrally by the posterior point of the third ventricle and caudally by the vermis of the cerebellum [3].

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However, on the basis of radiological diagnosis, many authors include the midbrain, posterior third ventricular and falcotentional masses in the pineal region [4, 5, 6, 7, & 8].

The variant types of neoplasms arising in the pineal region follow the Russell and Rubinstein classification of 1977:

1. Tumours of germ cell origin 2-tumours of pineal parenchymal cells 3-tumours of glial and other cell origin 4-non neoplastic cysts and masses [9].

It is also worth mentioning that the clinical presentation of tumours in the pineal region vary in 4 categories depending upon the size of the lesion, whether it invades the surrounding neural structures, it grows into and expands the ventricular cavity, it has a secretory function or is associated with a suprasellar component [10, 2].

Recently several modalities of investigations have been used in the diagnosis of pineal region tumours, facilitating the path to an optimum management. These investigations usually fall into three main groups, namely:

1. Radiological investigations mainly C. T. scan and MRI.
2. Cerebrospinal fluid study both cytology and tumour markers.
3. Others like neurophthalmological investigations and endocrinal assessment [11].

Inspite of the good surgical results, it is still debatable whether it is in the patients best interest to explore these lesions or whether to treat the hydrocephalus by a shunt procedure followed by blind tumour irradiation [12].

However, in the last 15 to 20 years, the science of radiotherapy, chemotherapy and lastly immunotherapy has become very much dependent upon the nature of the tumours to be treated and the role of blind irradiation progressively regressed [13 & 14].

The surgeon should try to fulfill the objective of the direct attack in the following order : 1) Obtain an adequate tissue biopsy for planning an optimal treatment; 2) Decompress the posterior third ventricle and the aqueduct thus treating hydrocephalus without the need for a permanent shunt device if possible; 3) Excision of the whole tumour achieving complete surgical cure [15].

Several approaches were used for surgery of the pineal region tumours. Only three approaches survived after the use of the neurosurgical microscope. These approaches are : 1) The posterior transcallosal approach; 2) The supracerebellar infratentional approach; 3) The occipital transtentorial approach. Now only these three approaches stand for exploring the pineal region tumours in modern neurosurgery [16].

Inspite of adequate tissue biopsy a controversy exists regarding the optimal dose, the volume to be irradiated, and indications of prophylactic spinal irradiation. This is due to the heterogenicity of pineal region tumours and their different biological behaviour [17].

The accepted radiotherapy for pineal region tumours in most centers consists of 2000 rads to the ventricular system with a focal or a boost dose of 3000 rads to the tumours bed, to bring the total midplane tumour dose to 5000 rads [18].

The overall incidence of spinal seeding from pineal tumours is approximately 10% being much more common in germinomas and pineoblastomas [17].

Yet the most accepted indications for cranioaxial irradiation are summarised as follows : 1) Intracranial seeding; 2) Positive cerebrospinal fluid cytological examination for tumour cells 3) Positive myelography and/or neuroimaging techniques for spinal seeding [18 & 19].

Material and Methods

16 cases are included in this study with a male to female ratio of 1 : 1. The age ranged from 2.5 years to 12 years with a mean of 5.7 years.

All the patients were subjected to thorough history taking, general and neurological examination.

The radiological investigations entailed plain X-rays to the skull, C.T. scan and MRI to the brain with control C.T. scans to all the patients and even follow-up MRI for 2 cases (Figs. 1 & 2).

Cerebrospinal fluid study both cytological and tumour markers (alpha-fetoproteins and beta human chorionic gonadotrophins) were done to all the patients. The sample was obtained during the cerebrospinal fluid diversion procedure. All the cytological studies were negative, while none showed elevation in alpha-fetoprotein alone. Two patients (one pineoblastoma and the other germinoma) showed elevation of beta human chorionic gonadotrophins alone. Three cases of mixed germ cell tumours had elevation in both tumour markers. Further investigations for the detection of seeding were done, myelography in one case, abdominal sonography in two cases with abdominal symptoms after ventriculo-peritoneal shunts. A postoperative MRI study was done with contrast in one case to show spinal seeding.

All the 16 cases had hydrocephalic changes and needed the application of a ventriculo-peritoneal shunt before the direct approach. Since a right occipital transtentorial approach is the favoured procedure, the shunt was applied on the left side so as not to hinder surgery. The right occipital transtentorial approach was the chosen approach in 15 cases (Figs. 3, 4, & 5). Only one patient was operated upon through a supracerebellar infratentorial approach.

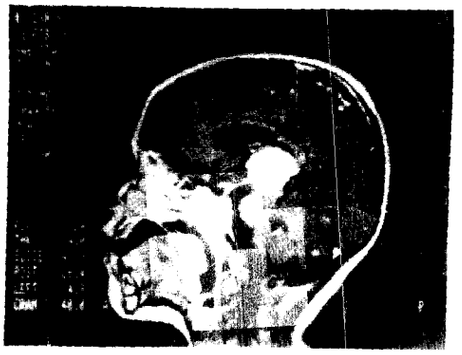

Fig. (1): MRI, T1 weighted image with Gd-DTPA showing an enhancing S. O. L. in the pineal region which proved to be a pineoblastoma.

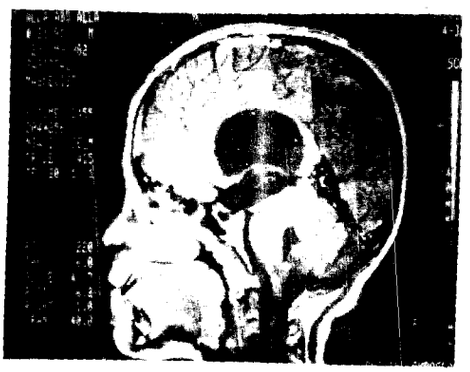

Fig. (2): MRI, T1 weighted image with Gd-DTPA showing an enhancing S. O. L. in the pineal region which proved to be an astrocytoma Gr II.

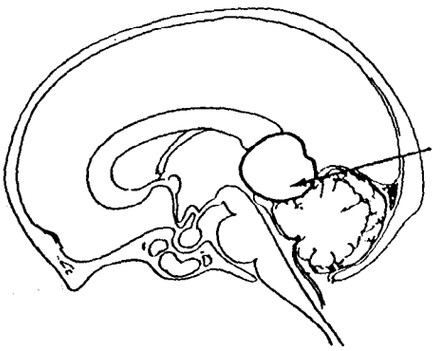

Fig. (3): A diagram of the occipital transtentorial approach to a pineal region tumour.

As regards the adjuvant therapy, all the patients received cranial irradiation by two opposing lateral fields in a dose ranging from 5000 to 5500 rads. The local field irradiation ranged from 3000 to 3500 rads to the tumour site, and whole brain irradiation in a dose ranging from 2000 to 2500 rads. Three cases of variant pathologies meeting the criteria for spinal irradiation received an additional radiation of 2500 to 3000 rads. Those criteria were : 1) Positive cerebrospinal fluid cytology, 2) Positive spinal MRI with Godolinium or myelography for spinal seeding, 3) Positive radiological or operative findings of other tumour foci in the brain indicating cerebrospinal fluid spread.

Fig. (4): Splitting of the tentorium after bipolar coagulation at the site of the tentorial opening. The arachnoid over the tumor mass is apparent.

Fig. (5): Appearance of the ependymal lining of the posterior part of the third ventricle after decompression. There is still a remainder of the mass beneath and vein of Galen.

The cases were all followed up clinically every 2 weeks. A radiological follow-up and an estimation of the level of tumour markers in the cerebrospinal fluid were done every 8 weeks or earlier in case of need. This was done for a period ranging from 4 months at the minimum and 2 years at the maximum. Chemotherapy was given in two cases (one germinoma and the other pineoblastoma) due to elevation of the tumour markers and evidence of recurrence following surgery and radiotherapy.

Results

The cases of this series were followed for a mean period of 9 months with the overall results being excellent in 14 cases without any postoperative deficits. One case had visual field defect postoperatively which disappeared completely after 6 weeks.

Another case already had cerebellar manifestations preoperatively which did not improve or resolve but actually increased after surgery. This was mostly due to minimal oedema following excision of the part of the tumour invading the cerebellum. The ataxia regressed after one week of surgery but did not disappear completely.

There was no operative mortalities in our series. Only one poor outcome was due to severe haematemesis ending fatally 2 weeks after surgery.

The histopathological verification of the nature of the tumours encountered in this series are shown in table (1).

Table (I): Results of Histopatological Examination of the Variant Tumours Encountered in This Series.

Histopathological Nature	No. of Pts
Germinoma	4
Germinoma with elements of endodermal sinus tumour	1
Mixed germ cell tumour	1
Pineoblastoma	6
Pineocytoma	3
Astrocytoma	1

Discussion

Although pineal region tumours are not among the most common brain tumours, they are certainly one of the most controversial. This is in part due to the unique site within the brain mandating perfect knowledge of the microsurgical anatomy of this region, and awareness of the variability of the deep venous system.

The current neuroradiological techniques are sensitive in picking up small lesions but are not able to specify the pathology. The diversity of pathological lesions in this area makes ample biopsy the only method for proper tissue diagnosis. This has been

repeatedly emphasized and is now the state of the art for the management of pineal region tumours, especially with the refinements of the surgical procedures and adjuvant therapy [20 & 21].

The belief that empiric blind irradiation of pineal region tumours has been recently questioned and with the advancement in adjuvant methods of therapy the mode of management has been standardized. The biopsy results is considered the hallmark for the further management of pineal region tumours. The ratios of parenchymal tumours to germ cell tumours was 1.5 : 1 in this study and the pineoblastoma to germinoma was also 1.5 : 1, which coincided with Herrick's series in 1984 [1].

This fact emphasized the importance of obtaining a proper tissue diagnosis before proceeding to radiotherapy and that the actual incidence of germinoma has decreased markedly. It is also of importance the fact that pineal parenchymal tumours especially pineoblastoma may simulate any pathological lesion on C. T. scan and MRI, which led to the conclusion that ample tissue biopsy is still the only proof of the correct pathological nature of the pineal region tumours. Another surgical goal would be to achieve third ventricular decompression for restablishment of the cerebrospinal fluid pathways. The limits of excision would be total in benign lesions, aiming at total removal in well localized and curable lesions or biopsy only in malignant and infiltrative or highly vascular lesions.

If the decompression is not achieved a drainage or diversion of the trapped cerebrospinal fluid should be planned to relief the increased intracranial tension.

The choice of the surgical approach to the pineal region tumours remains up to the surgeon. This series has shown the ease of the occipital transtentorial approach, its wide exposure to the area and its low morbidity.

The surgical outcome of this study was comparable to the series of Neuwelt, Stein and Lapras, which had respectively 6%, 6.1% and 7.3% operative mortality and morbidity [23, 24, & 15]. This was attributable mainly to the policy of safe surgery without rushing to undue triumphs by aiming at potentially dangerous total excision.

All the three cases showing recurrence after surgery and radiotherapy were managed by chemotherapy. They are still being followed-up and doing well.

It appears however that there is a great future for chemotherapy as one of the main and first lines of management of pineal region tumours [10 & 22].

The policy of starting with chemotherapy as an adjuvant before radiotherapy may be promising especially that the pineal gland is devoid of a blood brain barrier permitting the choice of chemotherapy on

the basis of the tumours inherent sensitivities to the drug after proper tissue diagnosis.

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