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POSTERIOR CRANIAL FOSSA TUMOURS IN CHILDREN AT KENYATTA NATIONAL HOSPITAL, NAIROBI
P. K. Wanyoike, MBChB, MMed (Surg) Consultant General Surgeon and Senior Registrar Neurosurgery, Kenyatta National Hospital, P.O. Box 20723, Nairobi, Kenya

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P. K. WANYOIKE

ABSTRACT

Background: **The management of children with posterior fossa tumours is a challenge to health care professionals worldwide. Difficulties in diagnosis especially in children less than three years is well documented. Limited diagnostic modalities and lack of awareness of the symptoms and signs as well as societal perception of children's complaints contributes to late presentation. Kenyatta National Hospital Neurosurgical unit is the only specialized unit among the public hospitals in Kenya where such patients are referred.**

Objective: **To review the management of posterior fossa tumours in children at Kenyatta National Hospital.**

Design: **A retrospective analysis of children treated for posterior fossa tumours at the neurosurgical unit of Kenyatta National Hospital between 1996-2003.**

Setting: **Neurosurgery unit, Kenyatta National Teaching and Referral hospital.**

Results: **Thirty seven children were treated for posterior fossa tumours between 1998 and 2003. Twenty four were females while thirteen were males giving a male: female ratio of 1:1.8. The age varied between 2-16 years with a mean of 6.7 years. Cerebellar symptoms were the most common mode of presentation (30%) followed by headaches and vomiting. Twenty percent of our patients were blind at presentation probably due to chronic effects of raised intracranial pressure. Out of 11 patients with histological diagnosis of meduloblastomas, over 99%, were females and only one was a male. Astrocytomas were evenly distributed at five males and six females. The mean duration of symptoms was 3.7 months while it took eight weeks between time of diagnosis and treatment.**

Conclusion: **Posterior fossa tumours in our set-up are more common in females than in males, M:F ratio of 1:1.8. Over 90% of medulloblastomas are found in female children making it a predominantly female tumour as opposed to available literature. The delay in diagnosis is probably due to lack of information both to the parents and health care providers and expensive diagnostic tools. A high index of suspicion, and a good history and clinical examination is required in the diagnosis of posterior fossa tumours in children especially those below three years.**

INTRODUCTION

Tumours of the central nervous system are the second commonest childhood tumours (20%) after leukaemia (37%) and are the most common solid paediatric tumours comprising 40-50% of all tumours(1,2). Posterior fossa (infrateritorial) tumours comprise between 54-70% of childhood brain tumours compared to 15-20% in the adult population(3). Worldwide the commonest paediatric posterior fossa tumours are medulloblastoma (20%) astrocytoma (15%), brainstem gliomas (15%) and epedymomas (8%) in that order(4).

The estimated incidence of brain tumours in children is 2-3.5 per 1 00,000(5). The mean age at diagnosis has decreased from about 13 years in 1930(6) to nine years in 1970(7) to the current figure of 6.5 years in 2000(3). In our setup the mean age at diagnosis was 6.7 years.

The clinical presentation of a child with posterior fossa mass is dependent on the anatomical location, histologic type and the presence or absence of hydrocephalus(8). Because posterior fossa is only one tenth of the intracranial volume and contains structures that are responsible for vital functions, tumours in this region are quite challenging to the health care team. Symptoms result from either compression of vital structures or from raised intracranial pressure. Common presenting features include cerebellar symptoms, multiple cranial nerve palsies, headaches, vomiting and blindness due to raised intracranial pressures.

Most cerebella astrocytomas are low grade pilocytic astrocytomas. These are mostly cystic with a mural module. Solid astrocytomas usually found in the vermis are more aggressive with a poor prognosis depending on the histological grade. However of all childhood tumours, cerebella astrocytoma have the best prognosis(9).

Medulloblastoma is the commonest primary neuroectodermal tumour and also the most common solid childhood tumour. It comprises approximately 20% of posterior fossa tumours in childhood(10). They are extremely malignant tumours and are characterised by the ability to seed along cerebral spinal fluid pathways. The tumours are diffusely blue in colour on staining and are often called blue tumours. They are treated surgically and in most cases, complete macroscopic removal is possible. Controversy exists on whether to perform pre or post operative shunting procedure. Adjuvant therapy involves chemotherapy or radiotherapy. Radiotherapy however is not given to children less than five years, due its side effects.

Epedymomas arise from the epedymal lining of the ventricles (4th ventricle in posterior fossa). They are mostly benign but fast growing tumours(7). They recur frequently and tend to seed along cerebral-spinal fluid pathways. They are reddish grey, lobulated tumours and microscopically have a diagnostic feature of rosette formation.

Brainstem gliomas account for 25% of tumours arising in the posterior fossa in children. They present with multiple cranial nerve palsies, signs of long tract involvement and inco-ordination of movement. Classically, they present with diplopia due to involvement of 4th and 6th cranial nerves. Blockage of the aqueduct leads to signs of raised intracranial pressure. Brain stem gliomas are classified into; (i) Diffuse tumours, which balloons the brainstem and involves all layers. These are malignant with a very poor prognosis. It involves several cranial nerves particularly the 5th, 6th, 7th and 10th, (ii) Focal tumours that lie within the brainstem and (iii) Exophytic tumours that arise from the brainstem but extend into the cavity of the 4th ventricle.

Both focal and exophytic brainstem tumours are usually pilocytic and surgery is the treatment of choice, unlike the diffuse type which is malignant and only chemotherapy and radiotherapy can palliate.

MATERIALS AND METHODS

A six year retrospective review of records of patients presenting with posterior fossa tumours at the neurosurgical unit of Kenyatta National Hospital was carried out from 1998 to 2003.

Data recorded included age and sex distribution, duration and nature of symptoms, whether pre or postoperative ventriculo-peritoneal (V-P) shunt was done and the tumour type on histological examination. The time interval between onset of symptoms and diagnosis (calculated from time of onset of symptoms as recalled by parents and CT scan diagnosis) and the time between diagnosis and definitive treatment were analysed.

RESULTS

In our series 50% of paediatric posterior fossa tumours were in children less than five years with 30% in the 0-3 year age group, while 15% was in older children (11-15 years) (Table 1).

Table 1

Age distribution

Age (years)	No.
0-5	18
6-10	12
11-15	7

Table 2

Sex distribution

Sex	No.
Male	13
Female	24
Total	37

Out of 37 patients 24 were females and 13 were males giving a M:F of 1:1.8. (Table 2).

Table 3

Common signs and symptoms

Symptom	Mean frequency score	Mean duration in months
Headaches	17	6.5
Vomiting	14	5.0
Neck stiffness	3	2.6
Cerebellar signs	20	4.6
Blindness	7	1.0
Speech disturbance	6	2.4
Total mean duration		3.68

Cerebellar symptoms were the commonest mode of presentation followed by headaches and vomiting. Blindness was found in seven or 20% of our patients probably due to late presentation. Neck stiffness was described in only three of our patients (Table 3).

Table 4

Shunting procedure

Procedure	No.
Pre-operative V-P shunt	23
Post-operative V-P shunt	5
No shunt at all	5
Total	33

Most of our patients (70%) got a pre-operative V-P shunt inserted before definitive craniotomy and excision was done.

Five patients had a shunt inserted post-operatively while five patients did well without a shunt procedure (Table 4).

Table 5

Tumour type and distribution

Histological tumour type	Freq.	Male	Female
Astrocytoma	11	5	6
Medulloblastoma	11	1	10
Ependymoma	3	1	2
Tuberculoma	3	3	-
Meningioma	1	-	1
Total	29	10	19

Medulloblastoma showed a marked female predominance with 10 patients compared to one male while astrocytoma had an equal sex distribution. The three cases of tuberculoma were all in the males. Most studies quote both medulloblastoma and astrocytoma as being common in the males (Table 5).

Out of 37 patients reviewed over the six year period 12(40%) died at various times after surgery. Eleven are on active follow up while seven (24%) have been lost to follow up. Of those patients who died, the mean duration between surgery and death was 20 days with a range of 0-66 days.

The mean duration from onset of symptoms to diagnosis was 3.7 months. This was calculated from onset of symptoms as recalled by parent to time of CT scan diagnosis. It took an average of eight weeks from time of diagnosis to definitive management.

DISCUSSION

The results of this study show a female predominance of posterior fossa tumours compared to the male (M:F 1:18). It also shows a marked female predominance of medulloblastomas (99%) compared to the male. This trend contrasts with reported world literature of a male predominance.

There was an equal histological tumour type distribution between astrocytoma and medulloblastomas and this compares well with the world series.

A significant proportion of our patients (20%) presented blind. This is most likely due to delay in presentation and diagnosis and the long-term effects of raised intracranial pressure. In our set-up this can be attributed to lack of knowledge and awareness by the parents and lack of diagnostic tools especially in the rural areas. However, there is now a tendency to early diagnosis with the presence of computerized tomography scans in five major towns in Kenya. This facility however remains out of reach of most patients due to the cost implications.

Unfortunately most of the patients have to be referred to the Neurosurgical unit at Kenyatta National Hospital causing a delay in management, which at present stands at 7.5 weeks.

The overall mean duration of symptoms of 3.7 months is better than the Irish series of 4.7 months. This improvement has been seen in recent years from a mean of 5.7 months prior to 1998 to a minimum of one month in our series. This is attributed to early diagnosis and referral with a better postoperative outcome, which encourages parents to seek help assured of positive outcomes.

The difficulty of diagnosis of posterior fossa tumours in children under three years is well documented. Thirty percent of our patients were under three years. A thorough history and clinical examination with high index of suspicion is therefore paramount in diagnosis

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