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Posterior fossa haemangioblastomas in Northern Ireland: a clinico-epidemiological study

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SUMMARY

A retrospective study of 25 patients who presented with posterior fossa haemangioblastomas to the Northern Ireland Regional Neurosurgical Centre over the past 25 years has been carried out. The epidemiological and clinical features and the results after operative treatment are presented and compared with other series. Posterior fossa haemangioblastomas were more common in female than in male patients and solid tumours accounted for 40% of all cases. While only two patients had associated polycythaemia, five patients (20%) were found to have persistent leucocytosis pre-operatively, while 24% had von Hippel-Lindau complex including two patients who were operated upon for an associated spinal haemangioblastoma. Of particular interest was a patient who had neurofibromatosis. There was no perioperative deaths. Patients with solid tumours fared badly in the long term compared with those who had a cystic type.

INTRODUCTION

Haemangioblastomas account for 7.3% to 12% of all tumours arising in the posterior cranial fossa.[1, 2, 3, 4, 5] Considerable interest has been expressed in their epidemiology, the concurrence of polycythaemia[6, 7, 8] and von Hippel-Lindau complex.[9, 10] Many studies have been published from centres in Europe and the USA but no cases have previously been reported from Northern Ireland. We have therefore reviewed all the posterior fossa haemangioblastomas treated in this centre between 1960 and 1985 with special emphasis on the epidemiology, their association with von Hippel-Lindau complex and various haematological changes. An attempt was made to compare the results of operation with nine other series.[1, 2, 4, 11, 12, 13, 14, 15, 16]

Polycythaemia indicates a haemoglobin level greater than 18g% or a peripheral red blood cell count of more than 6.5×10^6 per ul.[7, 8] The von Hippel-Lindau complex is defined by Jeffreys as a 'clinico-pathological syndrome in which at least one haemangioblastoma of the neuraxis occurs with at least one intra-abdominal example of the following — cysts of the kidney, pancreas or liver, renal carcinoma or phaeochromocytoma. The term may also be applied to cases with haemangioblastoma of the retina and another haemangioblastoma within the neuraxis. The complex may be sporadic or familial'.[6]

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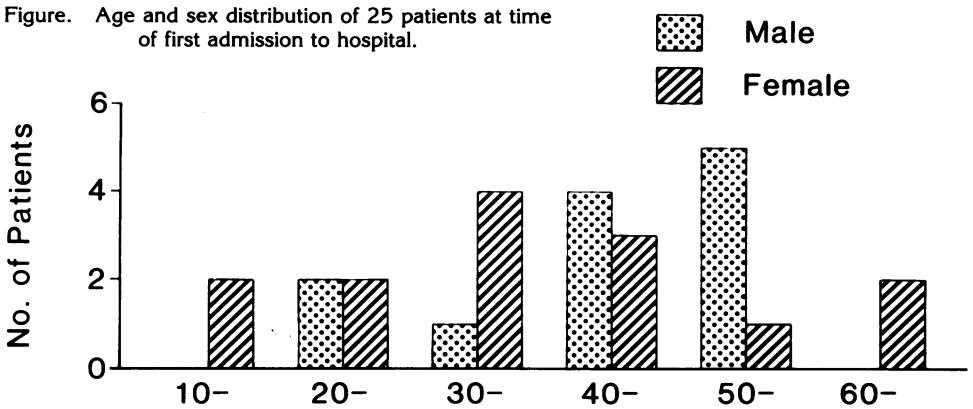
PATIENTS

All the 25 patients admitted to the Regional Neurosurgical Centre, Belfast, with posterior fossa haemangioblastoma were evaluated. There were 11 male and 14 female patients — a sex ration of 1 : 1.3. The age group of highest frequency was the sixth decade for male but the fourth for female patients (Figure). A study of both the ABO and Rhesus blood group distribution in 20 of these patients did not reveal any significant difference when compared with that of the general population.[17, 18]

Figure. Age and sex distribution of 25 patients at time of first admission to hospital.



The presenting symptoms and signs during the first admission are presented in Tables I and II. The average length of time that elapsed between initial symptoms and referral to hospital ranged from one month to five years with an average of 13 months. Sixteen patients (64%) presented with features of raised intracranial pressure. Papilloedema was detected in 36% of the patients which is lower than the reported incidence of 56% to 90%.[1, 2, 4, 13, 14, 15]

TABLE I ·

Symptoms of 25 patients with posterior fossa haemangioblastomas

Symptoms	*Number of patients*	*%*
Headache	20	80%
— occipital	6	24%
— frontal	6	24%
— hemispheric/global	8	32%
Vomiting	12	48%
Blurring of vision	4	16%
Diplopia	3	12%
Vertigo	3	12%
Mental changes	3	12%
Unsteady gait	15	60%
Tinnitus	2	8%
Neck stiffness	5	20%
Epilepsy	1	4%

TABLE II

Neurological signs of 25 patients with posterior fossa haemangioblastomas

Symptoms	*Number of patients*	*%*
Papilloedema	9	36%
Cerebellar deficit	12	48%
— nystagmus	6	24%
— dysmetria	4	16%
Ataxia	13	52%
Cranial nerves palsy	5	20%
Corticospinal tract involvement	3	12%
Occipital tenderness	2	8%

Six patients had haemangioblastomas elsewhere (four retinal and two spinal) leading to a diagnosis of von Hippel-Lindau complex in 24% of the whole group. The four patients with retinal angiomas had a strong family history of this condition. One other patient had an associated neurofibromatosis (von Recklinghausen disease).

Two patients (8%) had polycythaemia and both were found to have solid posterior fossa haemangioblastomas at operation. In five instances (20%), there was a persistent leucocytosis of more than 11,000 per ul without any focus of infection being discovered. One of these, a 52-year-old woman with a cystic cerebellar tumour had a leucocytosis of more than 25,000 per ul for more than six months before the cerebellar lesion was found. Her bone marrow examination did not reveal any abnormality.

Skull X-rays carried out on 10 patients did not reveal any abnormality. Electroencephalography (EEG) was normal in six patients and abnormal in seven, but correct localisation of the lesion was only found in two instances. Radioisotope scans were normal in five patients and abnormal in three, two giving a correct tumour location in the posterior fossa. Although vertebral angiography was performed in only nine cases it always gave the exact situation of the tumour. Prior to 1980, ventriculography was the only definitive diagnostic procedure achieved in nine patients. Since computed tomographic (CT) scanning became available in 1979, 13 cases of posterior fossa haemangioblastomas were diagnosed with its help. Four cases during the last two years were diagnosed by CT scan alone while in others angiography and ventriculography were used as adjunctive investigations.

Surgery was carried out on all the patients. As one later developed a second tumour, a total of 26 procedures was completed. Cystic haemangioblastomas were found in 15 cases situated in the cerebellar hemispheres. The size of these cysts ranged from 10 to 30 ml and that of mural tumour nodules between 0.5 and 2cm diameter. All of these tumours were removed totally. Solid tumours were found in 11 cases (42%), most of which were in the floor of the fourth ventricle or upper cervical cord. Two of the solid cerebellar masses were extirpated, two others partially removed, and the remainder biopsied and given radiotherapy. (Table III).

TABLE III

*Site and consistency of posterior fossa haemangioblastomas**

Site	Number		
	Cystic	*Solid*	*Total*
Right cerebellar hemisphere	11	2	13
Left cerebellar hemisphere	4	1	5
Vermis	0	1	1
Floor of fourth ventricle	0	5	5
Upper cervical cord	0	2	2
Total	15	11	26

*One patient presented with a solid upper cervical cord tumour six years after total removal of a right cerebellar cystic tumour.

All 25 patients have been followed up for periods varying from nine months to 14 years, with a mean of four years. Ten patients have died. The cause of death (Table IV) shows that five died suddenly, suggesting that the tumour had invaded the cardiac or respiratory centre, but post-mortem examination was not carried out. In two others, a medullary infarct was confirmed at autopsy.

TABLE IV

Ten cases of posterior fossa haemangioblastomas who died during follow-up

Case	*Year of death*	*Age at first admission*	*Sex*	*Site and consistency*	*Circumstances of death*
1	1963	65	F	(R) cerebellar, cystic	Sudden death, 3 years
2	1965	15	F	Upper cervical cord, solid	Sudden death, 2 years
3	1971	50	M	(L) cerebellar, solid	Features of increased ICP, 3 years after partial removal
4	1971	38	F	Vermis, solid	Medullary infarct following reoperation, 3 years
5	1973	43	F	(R) cerebellar, cystic	Sudden death, 3 years
6	1974	41	F	(R) cerebellar, cystic	Sudden death, 12 years
7	1976	42	M	(L) cerebellar, solid	Aspiration pneumonia following reoperation, 1 year
8	1980	51	M	Upper cervical cord, solid	Aspiration pneumonia, 4 months
9	1981	34	F	Floor of 4th ventricle, solid	Sudden death following revision of shunt, 1 year
10	1981	11	F	(R) cerebellar, cystic	Medullary infarct following reoperation for upper cervical solid tumour, 6 years.

Eight of the 15 patients who remain alive have sustained neurological improvement following surgery. Six out of these eight patients had cystic tumours. In five of the 15, the neurological picture was unchanged by surgery. Two were made worse, one requiring a feeding gastrostomy for bulbar palsy.

TABLE V

Results of surgery in 10 series of posterior fossa haemangioblastomas

	Percentage of all cases						
	Total cases	*Cystic*	*Solid*	*Multiple Tumours*	*Post-op death*	*Alive and well 3/12 post-op*	*Incapac-itated*
Cushing and Bailey (1928)	11	63	37	0	37	54	?
Davis (1946)	22	100	0	0	23	32	?
Perlmutter et al (1950)	25	100	0	0	8	68	20
Olivecrona (1952)	70	79	21	4	14	60	7
Silver and Hennigar (1952)	40	85	15	10	20	?	?
Krayenbuhl and Yasargil (1958)	45	86	14	?	9	64	8
Mondkar et al (1957)	108	70	30	9	15	86	?
Stein et al (1960)	19	80	20	0	30	52	0
Jeffreys (1973)	67	70	30	7	15	80	7
Chee and Bailey (1985)	25	60	40	4	0	60	8

DISCUSSION

In this study posterior fossa haemangioblastomas affected female patients more commonly than males, unlike any other series reported where there was a male to female preponderance of 2:1.[1, 4, 9, 11, 13, 14, 15, 16, 18, 20] Some comparisons with other series are shown in Table V. Clinical symptoms may appear at any age group but predominantly between the fourth and sixth decades of life. There was no significant difference in their ABO and Rhesus blood group distribution compared with that of the general population.[17, 19]

The clinical manifestations can usually be explained on the basis of the tumour location and slow growth characteristics. A triad of headache, vomiting and ataxia or ataxia, papilloedema and nystagmus[20] were found in a large proportion of these patients and any such symptoms should arouse suspicion of this type of lesion. The non-specific triad of headache, vomiting and papilloedema was present in only one-third of the patients. On average, 13 months elapsed between a patient first experiencing a symptom referrable to the nervous system and arrival at hospital compared with a range of seven to 12 months in other series.[1, 4, 11, 14, 15]

Various conditions have been observed in association with posterior fossa haemangioblastomas. Von Hippel-Lindau complex was found in 24% of our cases. One patient also had neurofibromatosis. Two patients with solid tumour had erythrocytosis but, to our surprise, five patients had persistently elevated peripheral white cell counts pre-operatively for which no other cause was found, highlighted by one woman having a leucocytosis of more than 25,000 per ul for six months before the cerebellar tumour was found.

Since the introduction of computed tomographic scans, the pre-operative diagnosis and localisation of this posterior fossa lesion has been made easier and more accurate. The presence of a posterior fossa cyst with a mural tumour nodule which enhances after injection of contrast is almost pathognomonic of a cystic cerebellar lesion.[21] However, in certain cases, angiography is still advisable to outline the vascular supply of the lesions, the mural nodule and the anatomy of

main vessels. Angiography in our series gave 100% correct localisation of the tumour, similar to that reported by Jeffreys.[1] Electroencephalography and radioisotope scans were of limited value in making a correct diagnosis and localising these tumours. Plain skull X-ray may be useful but did not reveal any abnormality in 10 cases. Ventriculography has now been superseded by CT scanning as a primary method of investigation. Solid tumours accounted for 40% of our cases — a higher incidence than usually found. Seven cases (64%) were located in the floor of the fourth ventricle and upper cervical cord.

The post-operative death rate in other series was 8% to 37%. Our results compare favourably in that no patient died during operation or the early post-operative period. This may reflect the introduction of the operating microscope for all cases, a more conservative approach to solid tumours (especially those involving the floor of the fourth ventricle and upper cervical cord) and advances in neuroanaesthesia and resuscitation. Nonetheless, 8% of the patients were incapacitated following the operation although 60% were alive and well at three months. In our experience, cystic tumours have a better outcome after surgery than solid tumours which is in agreement with other reported series.[22, 23]

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