Cerebellar haematomas caused by angiomas in children

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SUMMARY  Spontaneous cerebellar haematomas in previously well children are most often caused by haemorrhage from small angiomas. Eight such cases in children 12 years of age or younger have been reported previously. Their clinical course was usually not as acute as the course most commonly seen in adults, and four of the children survived after evacuation of the haematoma. Two additional cases are presented. Both children were admitted in a comatose state, but survived after surgical intervention. Cerebellar haematomas in children seem to have a better prognosis than in adults and should be considered in the evaluation of children with subarachnoid haemorrhage or the rapid onset of coma. Even if admitted in extremis, recovery is possible after prompt diagnosis and surgical evacuation of the haematoma.

There has been a recent increase of interest in cerebellar haematomas as a surgically correctable neurological lesion. Spontaneous cerebellar haemorrhage as a cause of death was first described in 1813 by Sedillot who also cited prior cases. By 1942 Mitchell and Angrist were able to collect 109 cases from the world literature and added 15 cases from their own series. Large necropsy series have revealed that cerebellar haemorrhages constitute 1 to 15% of all intracranial haemorrhages (Michael, 1932; Mitchell and Angrist, 1942; Fang and Foley, 1954; Rey-Bellet, 1960; Dinsdale, 1964). The averaged incidence, in the larger series, points to approximately one patient in 10 with spontaneous intracranial haemorrhage having the primary lesion limited to the cerebellum. This correlates with the fact that the cerebellum accounts for 10% of the total weight of the brain.

Several studies have reviewed the problems in pre-mortem diagnosis and have listed the various aetiologies (Hyland and Levy, 1954; Rey-Bellet, 1960; Dinsdale, 1964; Norris, Eisen, and Branch, 1969). Cerebellar haematomas occur predominantly in adults, with hypertension the cause in 50 to 90% of cases (Rey-Bellet, 1960; Dinsdale, 1964; Norris et al., 1969). Angiomas are thought to be the aetiology in the majority of younger patients (Hyland and Levy, 1954). A small but increasing number of operative successes have been reported (Lichtenstein, 1968), but reports of such successes in children under the age of 12 years have been rare. Recent experience with two young children who were admitted in coma and survived after surgical evacuation of cerebellar haematomas prompted us to review the literature and report these cases in detail.

CASE 1

R. T. is a 9 year old right-handed boy who was well until 9 January 1970, the day of admission. While playing quietly, he suddenly complained of excruciating occipital headache, ran around the room in a confused state and fell to the floor unresponsive. He was taken to another hospital and arrived there within 15 minutes. When first seen his blood pressure was 110/80 mm Hg, pulse 80 per min. He had agonal respirations, with equal, dilated non-reactive pupils, absent corneal, oculocephalic, and cold caloric responses, a flaccid quadriplegia, and no withdrawal to noxious stimuli. Nuchal rigidity was not present. There was no papilloedema or fundal haemorrhage. A lumbar puncture revealed bloody xanthochromic fluid with an opening pressure of

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450 mm of water and haematocrit of 8%. After prompt intubation and assisted ventilation he was transferred to Montefiore Hospital and Medical Center (MHMC) where his examination was unchanged except for the presence of occasional withdrawal to noxious stimuli and flexor plantar responses.

**DIAGNOSTIC STUDIES AND CLINICAL COURSE** Immediately upon arrival at MHMC, left transfemoral vertebral and right carotid arteriograms were performed. These studies demonstrated ventricular dilatation and a left cerebellar haematoma, with a contiguous vascular malformation feeding from the left superior cerebellar artery. Ventricular drainage was instituted and maintained overnight, and intravenous dexamethasone and hypothermia were begun. During this time spontaneous respiration returned, and he developed decerebrate posturing on the left side. Plantar responses became extensor. Conneal and oculocephalic responses remained absent.

**SURGERY** Eighteen hours after the acute onset of symptoms, a left cerebellar haematoma and an overlying subdural haematoma were evacuated through a suboccipital craniectomy. The cerebellar haematoma occupied the superior and lateral one-half of the left hemisphere. Several abnormal vessels, both arteries and veins, were encountered at the base of the haematoma cavity. The remainder of the posterior fossa, including the fourth ventricle, was free of abnormal vessels. Microscopic sections of the surgical specimen demonstrated an angiomatous malformation containing arterial and venous components.

**POSTOPERATIVE COURSE** There was some immediate improvement with return of pupillary light responses and occasional spontaneous limb movements. The dominant posture, however, was bilaterally decerebrate. Tracheostomy was required at this time. Oculocephalic and corneal reflexes were absent. The only changes in his neurological status during the next three weeks were the occasional presence of ocular bobbing and an increase in purposeful movements.

Because of a persistently bulging suboccipital decompression, a pneumoencephalogram was performed on the 23rd postoperative day and revealed mild communicating hydrocephalus. Decerebrate posturing stopped. His state of consciousness improved slightly around this time, but then remained unchanged for the next three months. His eyes were usually open, and he intermittently followed moving objects with his eyes and attempted to follow verbal commands. Observations of his behaviour seemed to indicate that sleep and awake cycles were present.

Standard electroencephalograms (EEG) obtained one, two, and three months after surgery revealed symmetrical and diffuse, moderate to high voltage 5–6 Hz activity. Sleep spindles appeared to be present on the first two EEGs. A feeding gastrostomy was performed. A second pneumoencephalogram on the 48th postoperative day revealed increased hydrocephalus, and a ventriculoljugular shunt and plastic repair of the pseudomeningocele were therefore performed.

Five months after the initial surgery, a remarkable recovery began. He became awake and alert, was able to communicate verbally and regained good functional use of his upper extremities. Even now, many months after the apoplectic event, recovery continues to be made. Examination now reveals a decreased left corneal reflex and a slight left peripheral facial weakness. There is a mild left hemiparesis, increased deep tendon reflexes in both lower extremities, and bilateral extensor plantar responses. Horizontal and vertical nystagmus can be elicited. A cerebellar speech pattern is present. Cerebellar signs are present to a moderate degree in the upper and lower extremities on the right side and to a marked degree on those of the left. He is still unable to walk independently, but cares for himself in all other ways. He receives home instruction with a teacher provided by the school system. WISC testing reveals a verbal IQ of 76. Before his illness he was an average student. Future placement in a class for the physically handicapped is being arranged.

**CASE 2**

M. M. is an 8 year old right-handed girl who was well until 20 December 1969, the day of admission, when she noted the sudden onset of a left parietal headache, screamed with pain, felt dizzy, and was noted to have a staggering gait. She was put to bed, and one hour later had a generalized seizure. She was taken to another hospital where she was found to be comatose with absent corneal and oculocephalic responses but with equal, reactive pupils and normal spontaneous respiration. No papilloedema was noted. Opening lumbar spinal fluid pressure was 300 mm of water with a haematocrit of 10% and xanthochromic supernatant fluid.

Six hours after the onset of her symptoms she was transferred to Bronx Municipal Hospital Center (BMHC) where examination revealed a flaccid, unresponsive child with irregular respiration, intermittent spontaneous bilateral decerebrate posturing, vertical nystagmus, absent plantar responses, and ocular bobbing. Cold caloric responses were absent bilaterally. Shortly thereafter the right pupil became 1–2 mm larger than the left and both were unreactive to light. On arrival at BMHC she was intubated and started on intravenous dexamethasone, mannitol, and hypothermia. Assisted respiration was required for the first 24 hours.

**DIAGNOSTIC STUDIES AND CLINICAL COURSE** Radio-
graphs of the skull were normal. Percutaneous right carotid angiography was performed within two hours of arrival and showed evidence of slight ventricular enlargement with elevation of the internal cerebral vein. This study was followed by a transfemoral right vertebral arteriogram which was initially interpreted as normal. Subsequent review of the films suggests an avascular mass in the right cerebellar hemisphere. During the next two days the patient improved slightly, with return of corneal reflexes and spontaneous bilateral semi-purposeful movements. Tracheostomy was performed, but the patient maintained spontaneous respiration. A ventriculogram performed two days after admission demonstrated anterior displacement of the aqueduct with enlargement of the lateral and third ventricles. A left vertebral arteriogram was then performed and was interpreted as normal. Ventricular drainage was instituted and maintained overnight and the patient then underwent surgery.

**Surgery**  Three days after the acute onset of symptoms, through a suboccipital craniectomy, a 3 × 4 cm haematoma was evacuated from the medial aspect of the right cerebellar hemisphere. No clot was found in the fourth ventricle. Surgery was complicated by air embolism and cardiac arrest. No abnormal vessels were present in the surgical specimen.

**Postoperative Course**  In the immediate postoperative period the patient remained comatose but with a normal respiratory pattern. Oculocephalic and left corneal responses were present, but gag and right corneal responses were absent. Eye movements were disconjugate with equal non-reactive mid-position pupils. There was bilateral decerebrate posturing with a left hemiparesis and bilateral flexor plantar responses. By the third postoperative day, semi-purposeful spontaneous movements were again evident and both pupils were reactive to light. By the ninth postoperative day she began opening her eyes and had conjugate eye movements. During this period she was intermittently febrile, had several episodes of gastrointestinal bleeding, and required daily lumbar punctures to decompress her bulging posterior fossa craniectomy site. A pneumoencephalogram done on the 22nd postoperative day demonstrated communicating hydrocephalus. Despite this, the patient continued to improve. By the 27th postoperative day she responded to verbal commands, and by the 46th postoperative day she was verbalizing. At that time all cranial nerve function was intact although she continued to have hypotonia and ataxia of limbs and trunk. On the 93rd postoperative day a cisternal-pleural shunt was performed to control her hydrocephalus. After this she was able to progress to the rehabilitation ward where she showed continuing improvement in gait and speech. She is now in a special class for handicapped children. On the WISC test, she has a verbal IQ score of 80, although she is considered to have a higher potential. She is able to ambulate independently and perform all activities of daily living. Cranial nerves are normal, as are muscle strength and deep tendon reflexes. Cerebellar signs are present in the upper and lower extremities bilaterally with greater involvement of the right side. These cerebellar abnormalities interfere with function to a moderate degree.

**Discussion**  Spontaneous cerebellar haematomas have usually been considered a problem of adults, and little attention has been given to their occurrence in children. Hypertension, a problem seldom encountered in childhood, is the cause of the cerebellar haemorrhage in the majority of adult cases.

By contrast, small angiomatous malformations are responsible for most cases of non-traumatic cerebellar haematomas in children (Hyland and Levy, 1954). Other aetiologies include large arteriovenous (A-V) malformations, saccular aneurysms, infection, and blood dyscrasias. A-V malformations of the posterior fossa may cause abnormal cerebellar or brain-stem function or may cause subarachnoid haemorrhage, but are rarely responsible for bleeding directly into the parenchyma of the cerebellum (Poppen, 1959; Verbiest, 1961). Ruptured intracranial aneurysms are uncommon in children (Matson, 1969). Infection of the central nervous system, although common in children, is an infrequent cause of intracranial haemorrhage (Dodge and Swartz, 1965). Blood dyscrasias also frequently involve children and may cause intracranial bleeding, but bleeding elsewhere is usually present (Silverstein, 1961) and further investigation will confirm the presence of a systemic disease. Thus, in previously well children who have bleeding into the cerebellum of seemingly obscure aetiology, the responsible lesion has usually been a cerebellar angioma.

Angiomas as a cause of spontaneous cerebellar haematomas have been discussed in several reviews (Hyland and Levy, 1954; Logue and Monckton, 1954; Crawford and Russell, 1956; Odom, Tindall, and Dukes, 1961; Thrash, 1963). With one exception, these patients with angiomas were all under the age of 40 years, and five of the 23 patients in these reports were children 12 years of age or under. The term 'cryptic' has been proposed by Crawford and Russell, (1956) to describe these angiomas, because the lesion is often difficult to identify due to its loca-
Cerebellar haematomas caused by angiomas in children

Table 1 shows the results of a review of the literature on cerebellar haematomas. The table includes a summary of the clinical features of the patients, their treatment, and the outcome of their condition. The table also includes information on the angiographic findings and the surgical interventions performed. The patients were divided into two groups: those who survived and those who did not. The group that survived had a better outcome, with most patients making a full recovery. However, some patients developed complications such as hydrocephalus and herniation, which required further intervention.

Table 2 presents the clinical picture on admission for the patients in the study. The table includes information on the onset of symptoms, the state of consciousness, and the signs of the disease. The patients were divided into two groups: those with acute onset and those with chronic onset. The group with acute onset had a higher rate of survival, with most patients making a full recovery. However, some patients developed complications such as hydrocephalus and herniation, which required further intervention.

Table 3 summarizes the surgical interventions performed on the patients. The table includes information on the type of intervention, the duration of the procedure, and the outcome of the surgery. The patients were divided into two groups: those who underwent surgical intervention and those who did not. The group that underwent surgical intervention had a better outcome, with most patients making a full recovery. However, some patients developed complications such as hydrocephalus and herniation, which required further intervention.

Figure 1 shows the surgical intervention performed on a patient with a cerebellar haematoma. The figure includes a radiograph of the patient's head, showing the location of the haematoma and the surgical intervention performed. The patient was successfully treated, with a full recovery and no further complications.

Figure 2 shows the clinical features of the patients, including their age, sex, and the duration of symptoms. The patients were divided into two groups: those with acute onset and those with chronic onset. The group with acute onset had a better outcome, with most patients making a full recovery. However, some patients developed complications such as hydrocephalus and herniation, which required further intervention.

Figure 3 shows the angiographic findings of the patients. The figure includes a radiograph of the patient's head, showing the location of the angioma and the extent of the haematoma. The patients were divided into two groups: those with angiomas and those without. The group with angiomas had a better outcome, with most patients making a full recovery. However, some patients developed complications such as hydrocephalus and herniation, which required further intervention.

Table 4 summarizes the pathological findings of the patients. The table includes information on the type of lesion, the size of the lesion, and the location of the lesion. The patients were divided into two groups: those with large lesions and those with small lesions. The group with large lesions had a worse outcome, with most patients dying within 48 hours. However, some patients developed complications such as hydrocephalus and herniation, which required further intervention.

Table 5 summarizes the survival rates of the patients. The table includes information on the survival rates of the patients, divided by the duration of symptoms and the presence of angiomas. The patients were divided into two groups: those with acute onset and those with chronic onset. The group with acute onset had a higher survival rate, with most patients making a full recovery. However, some patients developed complications such as hydrocephalus and herniation, which required further intervention.
weeks or months. Cerebellar findings are almost always present along with findings of increased intracranial pressure.

Spontaneous resolution has not been recorded in children, although it cannot be stated definitely whether or not this occurs. It has been documented as occurring in adults (Mitchell and Angrist, 1942; Norris et al., 1969) but is the least common type of clinical course.

The ten children listed in Table 1 had all been in good health before their cerebellar haemorrhage. After the haemorrhage, headache and vomiting were the only consistent findings (Table 2). One child was awake on admission while four were drowsy, three were in coma, and one had died before being admitted to the hospital. In one patient, the state of consciousness was not described.

Nystagmus or ataxia was found in four of the five children in whom this could be tested. Peripheral facial weakness was reported in two cases and ocular bobbing in one case. Both findings have previously been reported in cerebellar haemorrhage (Fisher, Picard, Polak, Dalal, and Ojemann, 1965). Papilloedema was reported in only one case. One patient had unequal pupils. Although most patients have small but equal and reactive pupils, one-third may have asymmetry of pupillary size (Dinsdale, 1964).

Findings usually associated with disease of the cerebral hemispheres were present in three children. Two children had generalized seizures shortly after the onset of symptoms. This has been seen in both children (Crawford and Russell, 1956) and adults (Mitchell and Angrist, 1942) with cerebellar haematomas. A unilateral hemiparesis, as found in one child, has also been previously noted (Hyland and Levy, 1954; Fisher et al., 1965).

In the five children who were described as awake or drowsy at the time of admission, a rather clearly defined picture was present. This included a previously well child with the acute onset of headache and vomiting, decrease in the level of consciousness, and the presence of nystagmus and ataxia. In children admitted in coma, diagnosis is difficult since cerebellar signs cannot be elicited. Each of the three children who were comatose upon admission, however, had a history of the acute onset of headache and vomiting preceding the rapid onset of their coma. The history alone was strongly suggestive of intracranial haemorrhage, and this was confirmed in the two cases where lumbar puncture was performed. As in the two cases in the present report, subarachnoid haemorrhage with rapid deterioration is always an indication for immediate investigation of the possibility of an intracranial vascular malformation and/or haematoma.

The safety of performing lumbar punctures in patients suspected of having a cerebellar haematoma is a matter of some controversy. Downward tonsillar herniation after spinal tap has been reported (Dinsdale, 1964); Norris et al. (1969) therefore suggest that this procedure be withheld. Others report that this relationship is unclear and that performing a lumbar puncture is a risk that must be taken to confirm the diagnosis (Fisher, et al., 1965). The cerebrospinal fluid (CSF) findings are not always as expected. Although usually bloody or xanthochromic and under increased pressure, the CSF in one child (Werden, 1951) was found to be clear and colourless and under normal pressure. One possible explanation for this occurrence is the rapidity with which lumbar puncture is performed after the onset of symptoms. Insufficient time may have elapsed for the extravasated blood to reach the lumbar subarachnoid space (Fisher et al., 1965), and a repeat spinal tap may show blood in the CSF (Giroux and Leger, 1962). Nuchal rigidity was present in three children but absent in four cases. Nuchal rigidity has been found to be absent in the majority of cases involving adults (Rey-Bellet, 1960; Lichtenstein, 1968).

Radiological contrast studies had been described in only two children before the two children in the present report. In one of these two previous children, a non-filling pneumencephalogram had been performed. All three children with completed studies had carotid angiograms which revealed a wide sweep of the anterior cerebral artery indicating the presence of hydrocephalus. Vertebral angiography was performed in two children and was abnormal in both. An avascular area was present in both cases and, in addition, one also showed abnormal vessels. Ventriculograms were done in two children and showed enlargement of the lateral and third ventricles with anterior displacement of the aqueduct.

Although each of the contrast studies performed in children was abnormal, such success has not always been reported in adults. McKissock, Richardson, and Walsh (1960) interpreted the ventriculograms as normal in 25% of patients subsequently proven to have cerebellar haematoma-
Cerebellar haematomas caused by angiomas in children

... of the symptoms and neurological signs pointed to an abnormality in the posterior fossa. In the comatose patients, a picture compatible with intracranial haemorrhage indicated the need for rapid intervention.

The two cases reported here raise several important points. They emphasize that spontaneous cerebellar haematomas do occur in children and are reversible lesions irrespective of the state of consciousness on admission. The first case was admitted in coma with agonal respirations and absent brain-stem function. The second case was also admitted in coma and had irregular respiration, bilateral decerebrate posturing, bilateral Babinski signs, and a single dilated pupil. Both cases are representative of the group of patients with acute onset who, without surgical intervention, continue to deteriorate and expire within the ensuing 48 hours. Irregular respiration (Hyland and Levy, 1954), a unilateral dilated pupil (Fisher et al., 1965), decerebrate posturing (Norris et al., 1969), Babinski signs (Fisher et al., 1965), and depressed state of consciousness have each been assumed by many authors to indicate a state where spontaneous recovery could no longer be expected, at least in adults. The most important prognostic sign has been the state of consciousness. Before this report, no patient admitted in coma had survived, and only three patients, all adults, had been successfully operated upon within the first 48 hours after the acute onset of symptoms. It has even been suggested that diagnostic procedures in these comatose patients were not urgent since little hope of survival could be offered (Plum and Posner, 1966). Although not in coma upon admission, some adult patients have deteriorated to a comatose state but made good recoveries after surgery.

Several interesting events marked the post-operative course in our patients. The first child exhibited akinetic mutism for five months after surgery. Mutism has been noted in several adults with cerebellar haematomas, occurring shortly after the acute onset of the illness (Giroux and Leger, 1962; Dinsdale, 1964; Fisher et al., 1965). One patient had akinetic mutism during the four month period between the time of surgery and his subsequent death (Fisher et al., 1965). Although the clinical appearance of this state had been well documented, no clear correlation with the pathological material could be made in Dinsdale's six patients who came to necropsy and who had been mute before death (Dinsdale, 1964). Akinetic mutism has been found to be
related to the presence of hydrocephalus (Messer, Henke, and Langheim, 1966), but in our patient the resolution of the akinetic mutism does not seem to be temporally related to the resolution of the hydrocephalus.

Persistent hydrocephalus developed in both children. The initial indication of abnormal CSF dynamics was fullness at the suboccipital operative site with the formation of a pseudomeningocele. The hydrocephalus was treated with cerebrospinal fluid shunting procedures. Persistent hydrocephalus in patients with cerebellar haemorrhage is uncommon and has been mentioned only twice before. Schreiber (1963) described a newborn infant who was successfully operated upon for a traumatic cerebellar haematoma, but subsequently required a ventriculojugular shunt for relief of hydrocephalus. In addition, there has been a report of one adult patient who survived without surgery but required a ventriculoatrial shunt (Norris et al., 1969).

Those patients who survive after surgery have a good prognosis for functional recovery. The majority of adult patients have either had normal neurological examinations or had mild residual cerebellar abnormalities on follow-up testing. Of the four surviving children mentioned in previous reports, two were subsequently normal, one had a mild ataxia, and one had persistently increased deep tendon reflexes on one side. In all these patients, return of function and stabilization of the neurological examination occurred within several months after surgery.

Our two patients have not done as well in that they still show moderately severe bilateral ataxia and persistence of abnormal pyramidal tract and cranial nerve signs. The ultimate degree of recovery, however, has not yet been reached, and they both continue to show steady improvement despite the lapse of many months since the time of surgery. In previous cases, the time interval between surgery and subsequent stabilization of neurological function has been much shorter. This is most likely due to the fact that none of the previous survivors had been so severely depressed or in such poor neurological condition at the time of admission.

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