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Spontaneous cerebellar haemorrhages in childhood

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Abstract Spontaneous cerebellar haemorrhage is very rare in children and is often associated with blood diseases such as haemophilia A or acute lymphatic leukaemia. During 1995, two paediatric patients with spontaneous cerebellar haemorrhage underwent surgical treatment in the Neurotraumatology Division of "La Sapienza" University hospital in Rome. The authors discuss this unusual pathology with particular regard to the age of the patients described.

Key words Spontaneous · Cerebellar haemorrhage · Paediatric · Surgery

Introduction

Spontaneous cerebellar haemorrhage (SCH) was first described by Mitchell in 1942 [11]. Its incidence has been estimated as between 6% (21) and 13% (11) of all parenchymal haemorrhages, and has steadily increased since the introduction of CT into clinical practice [2, 3, 5, 12, 13, 17, 20]. Factors that predispose to and/or accompany onset of SCH are the same as those for cerebellar haemorrhages [17]. The main difference is that cerebellar haemorrhages are more rapidly fatal, with the risk of mortality estimated at approximately 73.5% by McKissock [10].

In their study, Rosemberg and Kauffman [18] demonstrated the total lack of pathognomic signs useful for early diagnosis; this characteristic, coupled with the lack of specific radiological investigations in their series (CT, angiography of the cerebral vessels), significantly influenced the mortality, as observed by Fisher [4]. The following signs were considered predictive by Ott [16] for early diagnosis: endocranial hypertension (nausea, vomiting, headache), inability to remain standing, brain stem signs (less frequent), deficits of the cranial nerves and alterations of respiratory function.

In 1978, on the basis of CT findings, Little et al. [8] established the parameters of operability in their series of patients: they divided patients into two categories: those who presented brain stem compression and those who did not. These workers advocated surgical treatment for patients with a blood clot over 3 cm in diameter, acute hydrocephalus or clinical signs of worsening brain stem involvement.

The concept of "tight posterior cranial fossa" (TPCF) was first suggested by Weisberg [22] to explain and/or classify cases that displayed rapid involvement of the basal cisterns, hydrocephalus and distortion of the IV ventricle.

In 1988, Amacher [1] was able to identify a definite relationship between the various clinical stages and gradual involvement of the basal cisterns and/or the eventual presence of brain stem compression. Kobayashi et al. [7] introduced a new parameter for determining which patients should undergo surgery; this took into account the size of the haematoma and the patient's neurological condition on admission.

Patients and methods

Between January 1990 and December 1995, we observed two pediatric patients (late childhood; 13 and 15 years of age) with SCH. Both were surgically treated in the Neurotraumatology Division, Department of Neurological Sciences, Rome, "La Sapienza" University.

On admission, both patients were evaluated neurologically and according to the Glasgow Coma Scale (GCS). Appraisal of CT and MR images paid particular attention to the following aspects: extent of the haemorrhage and whether it involved the ventricles and brain stem; hydrocephalus; brain stem compression; compression or obliteration of the basal cisterns, evidence of a tight posterior cranial fossa (p.c.f.). The last parameter was evaluated in accordance with the classification proposed by Weisberg [22], namely: stage 1, basal cisterns in the p.c.f. not visible; stage 2, enlargement of the III ventricle, temporal horns and lateral ventricles; stage 3, IV ventricle not visible (inconstant).

In addition, a search was made for any other pre-existing and/or concomitant pathologies (diabetes mellitus, arterial hypertension, blood disease, liver disease, etc.).

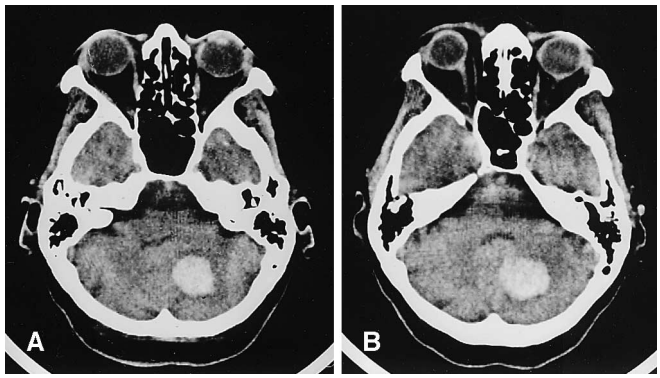

Fig. 1A, B A preoperative CT brain scan showed a large haematoma in the left cerebellar hemisphere, involving the posterior cisterns and with mild brain stem compression

Case reports

Case 1

This 13-year-old boy with acute lymphatic leukaemia was already receiving treatment in the Haematology Division of our hospital, since systemic disease had been diagnosed 1 year earlier: he had already commenced chemotherapy. He was hospitalized for the acute onset of severe nuchal headache accompanied by vomiting and mental confusion. Neurologically, he appeared apathetic but still cooperative. A CT brain scan detected the presence of an intracerebellar haematoma of the right hemisphere, which initially invaded the posterior basal cisterns without compressing the IV ventricle and brain stem; there was no apparent hydrocephalus. Via a right paramedian suboccipital craniectomy and a Y-shaped dural incision, the blood clot was removed after corticectomy. About 1 week after the operation the patient was discharged: he was completely autonomous and neurologically intact and had a KPS score of 90; the only disturbance was a slight instability when moving from the sitting to the standing position.

Case 2

This 15-year-old boy suffered from haemophilia A, with severe impairment of coagulation. He was admitted to the Emergency Department of our hospital because he presented with violent headache and vertigo and was unable to remain standing. Prior to the onset, he had felt slightly unwell and presented an uncharacteristic asthenia and also some signs of arterial hypotension. His neurological condition was as those reported above, with only a mild impairment of consciousness and persistent nuchal headache. Blood tests performed on admission revealed delayed coagulation times and normal platelet values. A CT brain scan (Fig. 1) documented an enormous haemorrhage in a left intracerebellar site, initial involvement of the posterior cisterns, mild brain stem compression, a normal posterior cranial fossa and initial distension of the supratentorial ventricular system.

The haematoma was surgically removed via a left suboccipital craniectomy and homolateral hemispheric corticectomy. The patient remained in hospital for 11 days after the operation and on discharge was able to walk supported on one side. Clinical-radiological follow-up examinations confirmed the good surgical outcome and the patient's return to normal family and social activities. He is still an outpatient at the Haematology Clinic of our hospital.

Results

Both patients described here presented a severe cephalalgic syndrome on admission, predominantly or exclusively nuchal. The predisposing and/or comcomitant pathologies identified were disease of the blood in 1 case and coagulation disorders in the other. The site of the haematoma was hemispheric in both of them. Indications for surgery were: haematoma larger than 30 mm, tight/p.c.f. and initial distension of the ventricular system.

Follow-up continued for about 12 months. At the end of this period, 1 patient was completely autonomous (KPS >90) while the other occasionally required assistance with his daily activities (KPS of 70).

Discussion

It has been estimated that about 50% of patients with spontaneous cerebellar haemorrhage die within the first 24 h and as many as 75% within the first week after haemorrhage [6]. Spontaneous cerebellar haemorrhage almost always affects middle-aged or elderly patients, and in other studies reported [1, 7, 9] the decades most frequently affected were the 5th, 6th and 7th. As emphasized by other workers [2, 4, 5, 14, 15, 19] spontaneous cerebellar haemorrhage is extremely rare and is usually favoured by the presence of blood disorders (proliferative diseases, coagulopathies) [15]. As already observed in elderly patients, the age of the two young patients described here did not influence the clinical evolution, nor was it an impediment to surgical treatment, as demonstrated by our previous study too.

However, as previously emphasized by Norris [15], both the natural history of the disease and the outcome of

surgical treatment were negatively influenced by the presence of other pre-existing and/or concomitant pathologies.

According to Muller [12], in 50% of cases the haematoma is complicated further by ventricular invasion, which aggravates the already severe neurological conditions. Prognosis differs according to the site of the haematoma. A medial site (vermian) seems to be a less favourable prognostic factor than a lateral one (hemispheric or paravermian): the studies reported by Kobayashi [7] and Luparello [9] also seem to confirm this.

In our view, rapid clinical and neurological deterioration is due to a tight p.c.f leading to rapid involvement of the posterior basal cisterns, as previously suggested by Weisberg [22]. According to Kobayashi [7] and Luparello [9], the simultaneous presence of hydrocephalus worsens the prognosis in such patients although, in our patients, early surgical treatment did not modify the quality of life.

We disagree with Neubauer's assertion [13] that all patients with GCS <9 and a haematoma of 3 cm or more in diameter should be surgically treated without further selection. In contrast to Rosenthal, we do not advocate surgical treatment for patients in arreflexic coma when the basal cisterns in the p.c.f. are not visible and the haematoma is 20 cc or more in volume, because it is unlikely that neurological recovery will be possible.

Conclusions

In the light of the two cases described here and our previous findings [17], we are able to make the following observations:

1. Childhood did not significantly influence the clinical evolution, nor did it represent a contraindication to surgical treatment when necessary.

2. The diameter of the haematoma was not predictive of clinical evolution, as long as there was no ventricular or brain stem involvement.

3. Hypertensive hydrocephalus, when treated early, did not modify the quality of life in the long term.

4. A GCS <8 was an unfavourable prognostic factor.

5. Distortion or obliteration of the basal cisterns was the deciding element indicating surgical treatment in borderline cases.

6. Patients with a GCS <13, haematoma >30 mm, normal-sized p.c.f., without acute hydrocephalus or any of the concomitant or predisposing pathologies mentioned above should be treated conservatively.

7. It is essential to keep any systemic disease that favours development of the haematoma, as in our cases, under control.

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