The Cerebrospinal Fluid Fistula. Rhinorrhoea, Otorrhoea and Orbitorrhoea

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History

The historical survey of cerebrospinal fluid leakage started with Galens investigations in the second century. He assumed, that the intracranial fluid was excreted from various parts of the brain and that it was stored in the ventricles and escaped through the nose. Thomas Willis published in 1676 (28) the book "Cerebri Anatome", in which he described two cases of cerebrospinal fluid rhinorrhoea. At the end of the 17th century Govaart Bidloo, a Dutch professor in anatomy and a medical adviser to King William III, reported a post–traumatic discharge of fluid from the nose. The first description of what was certainly a case of non–traumatic cerebrospinal fluid rhinorrhoea, was presented by Miller (19) in 1826. This case concerned an infant, whose skull showed an abnormal increase in circumference. At the age of ten months a leakage of cerebrospinal fluid through the nose occurred. After some months the baby died. Necropsy revealed an internal hydrocephalus and a small opening in the lamina cribrosa.

The term "cerebrospinal fluid rhinorrhoea" was introduced by Sir St. Clair Thomson in 1899 (24) in a monograph in which he reviews all such cases recorded up to that date.

In 1935 Britt (4) wrote: "Apparently brain tumours are the most frequent cause of rhinorrhoea. The escape of cerebrospinal fluid is undoubtedly due to the pressure of the increased intracranial fluid upon the lamina cribrosa". Nowadays cerebrospinal fluid leakage is mainly traumatic because of the increase in traffic accidents which tend to produce this certain kind of injury.

It was Dandy, who in 1926 (9) was the first one to repair an anterior fossa dural tear, caused by a head injury. Dandy used a transcranial extradural approach. Nowadays the operative treatment of a cerebrospinal fluid leak has become a routine neurosurgical operation.

Classification of cerebrospinal fluid leakage

Several causes of cerebrospinal fluid leakage are known. The first classification was given by Ommaya et al. in 1968 (21). They divided the causes of cerebrospinal fluid leakage into two main groups: the traumatic and the non–traumatic cerebrospinal fluid leakage. Our classification (Fig. 1) is based on the one given by Ommaya et al. (21). We decided to define as cryptogenic (spontaneous) cerebrospinal fluid leakage in cases of non–traumatic origin with unknown aetiology, in agreement with other authors (8, 10, 20, 26). In the literature traumatic, non–traumatic, and cryptogenic cerebrospinal fluid leakage are often confused (2, 7, 12, 22). The most common cause of cerebrospinal fluid leakage is a basal skull fracture, in which the dura is torn. Several papers on traumatic rhinorrhoea have been published (13, 15, 16, 25). Non–traumatic CSF–rhinorrhoea is mostly caused by an empty sella syndrome or by direct tumour growth (3, 17).

Pathophysiology and diagnostic measurements

It is well–known, that the anterior cranial fossa is separated from the nasal cavity and the paranasal sinuses by only a thin layer of bone, in some places with an adherent dura. A fracture of the bone and a laceration of the adherent dura does not always result directly in a cerebrospinal fluid leakage; interposing blood, fragments of bone or brain tissue may close the opening of the fracture. However the cerebrospinal fluid leakage may still result from coughing, sneezing or nose–blowing. A meningitis can be the first sign of an open connecting passage between the anterior cranial fossa and the nasal cavity. One should always suspect a cerebrospinal fluid fistula after the development of meningitis in a case of trauma that occurred a long time previously or an attack of meningitis shortly after a recorded accident.

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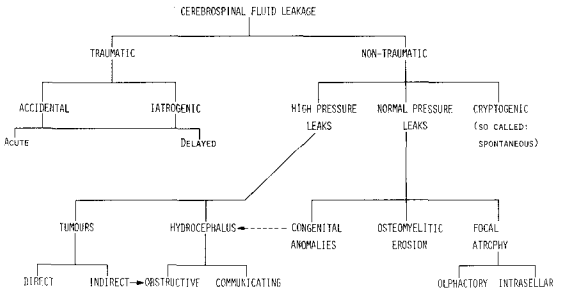

Fig. 1. The classification of cerebrospinal fluid leakage by Ommaya (modified).

In case of negative intracranial pressure a pneumatocele may develop, and this also increases the risk of meningitis.

In case of rhinorrhoea the most important symptom is a discharge of clear fluid from the nose. The amount of discharge can be increased by flexion of the head or by jugular compression. By examining the sugar content of the fluid discharge, cerebrospinal fluid may be suspected. In case of blood–stained fluid the demonstration of a clear halo surrounding a central blood–stain on a handkerchief or piece of cotton wool, can also be taken as evidence that the discharge contains cerebrospinal fluid.

As traumatic cerebrospinal rhinorrhoea often goes with injuries of the diencephalic and orbital areas of the brain, a pronounced organic cerebral psychosyndrome may be found. Furthermore it is important to check if there is a partial or complete anosmia.

In comparison to rhinorrhoea, otorrhoea is not so frequent, but it is also mainly caused by accidents. In cases of otorrhoea cerebrospinal fluid escapes through the ruptured dura and the bone fracture into the mastoid air cells and through the rupture of the tympanic membrane to the outer world. Also possible is an outflow of cerebrospinal fluid through the Eustachian tube into the pharynx. Otorrhoea seldom requires operative treatment because it seals spontaneously and permanently.

Although orbitorrhoea is quite rare it has been described in the literature (14).

For the direct demonstration of a cerebrospinal fluid fistula dyes like methylene blue or a photoluminescent substance like fluorescein are injected intrathecally. In this way it is easy to establish if the secreted fluid is cerebrospinal fluid. Often it is even possible to detect the site of the leakage.

A more modern method is the cisternal injection of a radionuclide like ^{99m}Tc (technetium–labelled human serum albumin). In the case of a fistula in the anterior cranial fossa the isotope can be seen in the region of the fracture or even in the pharynx. Sometimes the amount of secreted cerebrospinal fluid is low and the demonstration of a leak can be achieved by measuring the radioactivity of sponges, left in the nose for 24 hours, or by increasing the intracranial pressure by subarachnoid infusion.

Patients

Between 1969 and 1979 we operated on 79 patients, 76 showing a rhinorrhoea, two an otorrhoea and one orbitorrhoea (case history). There were 61 male and 18 female patients. According to our classification we found traumatic and non–traumatic cerebrospinal fluid leakage. The different causes are listed in Tab. 1.

The cerebrospinal fluid rhinorrhoea had an iatrogenic cause in 11 patients, including one case in which the fistula resulted from thermocoagulation of the gasserian ganglion, four cases with a leak after trans–sphenoidal hypophysectomy and six cases with a leak after ENT operations.

The first symptom in 66 (84%) patients with cerebrospinal fluid leakage was rhinorrhoea, that often stopped spontaneously. Meningitis occurred without CSF leakage in eight cases, intracranial air in two cases and CSF otorrhoea in two cases. One patient developed a CSF leak into the orbit.

Meningitis following cerebrospinal fluid discharge occurred in 29 (37%) patients, before they were admitted for an operative closure of the fistula. Some of these patients had had meningitis up to four times.

There was a lot of variation in the time of onset of symptoms (Tab. 2). The longest period in which a patient showed no symptoms of a fistula was 24 years. This patient was 22 years old when he suffered brain injury, from a gunshot in his anterior cranial fossa. After 24 years a meningitis occurred and a cerebrospinal fluid fistula was diagnosed.

Tab. 1. Different causes of cerebrospinal fluid leakage

Non-Traumatic:	Direct Tumour Growth	1	*Traumatic:*	Fighting	3
	Congenital Anomaly	1		Sports Injury	4
	Cryptogenic	1		Domestic Accident	5
	Empty Sella	2		Other Accident	9
				Iatrogenic	11
				Industrial Accident	12
				Traffic Accident	30

Twenty–five (32%) of our patients displayed symptoms of impairment of their sense of smell.

The routine test for confirmation of the CSF leak was radiological examination, performed in 61 patients, and radionuclide tests, performed in 58 patients. Positive radiological changes were found in 46 patients (75%). Possible but unconfirmed changes in 11 patients were noted and in the case of four patients, no fractures were observed. Isotope tests were found to reveal positive evidence in 39 patients (76%), uncertain results in 11 patients and negative results in eight patients.

Of the 79 patients operated on in our clinic, 74 patients were cured after the operation while five patients suffered from a recurrent leak and a second operation was necessary. Four of these patients were cured after the second operation. In one patient the result of the operation could not be assessed because the patient died of postoperative complications.

Case history (orbitorrhoea)

A one–year–old child fell 2 metres downstairs. After a couple of hours a swelling of the right eyelid occurred with a fluid discharge out of the corner of the eye (Fig. 2). X–rays revealed a large skull fracture extending into the right orbit. The tentative diagnosis of orbital cerebrospinal fluid leakage was confirmed by an isotope test (Fig. 3). During the operation we found a frontal brain contusion with a skull fracture and a frontal dural tear. The torn dura was repaired. One week after the operation there was no longer any swelling of the eyelid (Fig. 4). Up to the present, five years after operation no recurrence of cerebrospinal fluid discharge has occurred.

Tab. 2. Time of onset of CSF leakage after the trauma

Immediately	to	7 days after trauma	39
7 days	to	1 year after trauma	16
1 year	to	5 years after trauma	8
5 years	to	15 years after trauma	3
More than		15 years after trauma	8

Fig. 2. Swelling of the right eyelid. The picture is taken just before operation.

Fig. 3. The positive isotope test in the case of the orbitorrhoea.

Fig. 4. No swelling of the eyelid after operation.

Discussion

Our material shows, in agreement with the literature, that traffic accidents are the most frequent cause of CSF leaks. In our view spontaneous CSF leakage is of non–traumatic origin with an unknown aetiology, so we have slightly modified the classification given by Ommaya et al. (21).

Three patients were below the age of five years. One of these patients has been described in this paper. Rhinorrhoea is rare at this age because the paranasal sinuses are at an early stage of development and also because of the relative elasticity of the skull (5, 13, 14).

In CSF leaks the clinical symptoms are very variable. The most common finding in our patients was rhinorrhoea (84%) followed by meningitis (37%). In 53% the first symptoms occurred within seven days of trauma, but in almost 11% even later than 15 years after the trauma. The late appearance of a fistula is also mentioned by other authors (11, 15, 16, 25). In our opinion, this is mainly caused by cerebral atrophy which leads to a rupture of cicatricial tissue.

Twenty–nine of our patients had pre–operative meningitis. Excluded are the eight patients in whom the meningitis was the very first symptom. Other investigators placed the risk of pre–operative meningitis at between 9.3% and 65% (11, 15, 27).

In our patients 32% showed a disturbance of the sense of smell. Here also different percentages are mentioned, ranging from 17% up to 78% (11, 16).

Concerning the timing of operation, Adson and Uihlein (1) wrote in 1949, that it was wise to wait for a period of eight weeks, before operating on a traumatic cerebrospinal rhinorrhoea, because it frequently ceases spontaneously. Taking present knowledge into consideration it is still advisable to postpone an operation until some time after the head injury, in order that the frontal brain swelling can subside, but the operation is mostly postponed only a couple of days because of the heightened risk of meningitis which may cause permanent defects. If there is a compound frontal depressed fracture, immediate decompression is necessary. If the swelling of the frontal lobe prevents the sealing of the dura, a second operation must follow. In cases in which the patients suffer from meningitis, urgent treatment is required as soon as possible. Nowadays CAT scan can give further information on localized areas of the brain, for example, areas of swelling.

It often happens that, during operative treatment, no dural defect can be seen in spite of a positive radionuclide investigation where the site of the CSF leak has been established by sponge activity. In this case it often concerns olfactory filaments that were torn out when the injury occurred. This leak can be stopped by grafting muscle on to the ethmoidal cells with adhesive fibrin.

Our material shows that a CSF leak appears one or more years after the initial trauma in 26% of the skull injuries. This leads us to the opinion that CSF leaks which cease spontaneously, also need operative treatment.

We dismiss other methods of closing traumatic CSF fistulas, such as regular lumbar puncture or the insertion of an external or spinal CSF drainage although other investigators seem to have controversial opinions (6, 18, 23). In our opinion it increases the risk of an ascending infection leading to meningitis or arachnoiditis.

Summary

Between 1969 and 1979, seventy–nine patients suffering from a cerebrospinal fluid (CSF) leakage, were operated on in the neurosurgical department of the University Clinic Essen (West Germany). A modified classification, the onset of symptoms and the diagnostic procedures are described and some data are compared with the results of other investigators. Other methods of closing a CSF fistula and the timing of operation are briefly discussed. Also the case history of a patient with orbitorrhoea is described.

Key words:
Cerebrospinal fluid fistula – Rhinorrhoea – Otorrhoea – Orbitorrhoea.

Zusammenfassung

Zwischen 1969 und 1979 wurden in der neurochirurgischen Universitätsklinik Essen 79 Patienten an einer Liquorfistel operiert. Eine veränderte Einteilung, der Beginn der Symptome und die diagnostischen Untersuchungsmethoden werden besprochen und die Resultate anhand der Literatur miteinander verglichen. Ebenfalls wird ein Patient mit einer orbitalen Liquorrhore vorgestellt.

Schlüsselwörter:
Liquorfistel – Rhinogene Liquorrhoe – Otogene Liquorrhoe – Orbitale Liquorrhoe.

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