Rhinology, 29, 223-230, 1991

A clinical study of spontaneous CSF rhinorrhoea

N.S. Tolley

Royal National Throat, Nose and Ear Hospital, London, United Kingdom

SUMMARY

A retrospective study of 10 patients presenting with spontaneous (normal pressure) CSF rhinorrhoea was performed. The epidemiology, role of radiology and surgical treatment are discussed. Nose blowing was found to cause onset of rhinorrhoea in 30% of patients studied. The presence of congenital dehiscences and the formation of small meningoceles are the most likely aetiological basis for this condition. The Valsalva like effect of nose blowing may cause dural rupture at sites of dehiscence thereby leading to rhinorrhoea.

Contrast CT imaging identified the site of leakage in 70% of cases and further identified the side of leak in 20%. Contrast CT imaging is, therefore, the best modality for investigating these patients.

Extracranial methods successfully repaired 88% of leaks compared with 50% for craniotomy. Extracranial techniques should be the primary surgical option since they have higher rates of successful repair and less morbidity than craniotomy.

INTRODUCTION

CSF rhinorrhoea may be classified as either spontaneous or traumatic. Spontaneous rhinorrhoea may further be described as primary where no underlying cause can be found, or, secondary to either intra or extracranial pathology (Troland, 1960). It was Miller who first described this condition in 1826, although credit should be given to Sir St. Clair Thomson whose treatise on the subject in 1899 allowed its wider clinical recognition (St. Clair Thomson, 1899).

Intracranial causes such as olfactory neuroblastomas, meningiomas, encephalocoeles, gliomas and particularly pituitary tumours have all been described as presenting with spontaneous CSF rhinorrhoea. Extracranial causes include: angiofibromas, ethmoid osteomas, chronic suppurative sinus disease, syphilis, leprosy and influenza (McCoy, 1963).

Spontaneous rhinorrhoea may be caused by fistulous communication from the anterior and middle cranial fossae or the posterior fossa via the Eustachian tube (Brockbank et al., 1989). The radiological investigation of this condition must, therefore, assess all cranial fossae including the inner ears (Freeland, 1973; Kaufman et al., 1977). Radiology has an extremely important role in the management of this condition for the following reasons. Firstly, in cases of spontaneous rhinorrhoea intra and extracranial causes must be excluded. Secondly, the site, side and nature of the fistula is of prime importance in planning the surgical approach. Thirdly, if no leak is demonstrated, more conservative approaches may be prudent. This is based upon the fact that 30% of patients undergoing surgery without identification of the fistula site will require additional procedures (Ahmadi et al., 1985).

A detailed study of the case records of 10 patients collected over a 10 year period from 1980–1990 presenting with primary CSF rhinorrhoea was performed. Particular note was made of the incidence of meningitis and common cold symptoms, the radiological techniques used to demonstrate the site of the fistula and their correlation to operative findings. Finally, the success and morbidity of the surgical approach used was examined. The results are discussed in terms of possible mechanisms responsible for this rare and curious clinical condition.

ANATOMICAL AND PHYSIOLOGICAL CONSIDERATIONS

A comprehensive knowledge of the relevant anatomy and its variation is a sine qua non to the understanding of fistulous routes and their surgical treatment. The site of fistulous communication in primary spontaneous CSF rhinorrhoea involves dehiscence in the region of the cribriform plate, fovea ethmoidalis, sphenoid sinus and rarely the frontal sinus alone or in combination. The fovea ethmoidalis is dehiscent in approximately 14% of bones (Ohnishi, 1981). Similar dehiscences have been described in the sphenoid sinus (Peele, 1957; Morley and Wortzman, 1965; Hooper, 1971). Persistence of the embryonic lateral craniopharyngeal canal has also been proposed as a route for middle fossa leaks (Hooper, 1971). Dehiscence of the frontal sinus has been reported as an unusual anatomical variant and, in 10% of bones, the cribriform plate has been reported to extend into the frontal recess, lateral to the middle turbinate (Lang, 1989). This anatomical point is of importance in both the diagnostic localization and surgical treatment of fistulae. Furthermore, Ohnishi states that multiple dehiscence may occur.

Excessive pneumatization of the sinuses may be one reason for the presence of bony dehiscences of the skull base (Stupka, 1938). Recesses of the sphenoid sinus are common, in particular inferolateral recesses may be seen in 25–36% of sinuses (Lang, 1989). Morley and Wortzman (1965) reported lateral extensions in 28% of skulls, 16% were bilateral. Rarely, such recesses have been described as

pneumatizing the greater wings of the sphenoid reaching as far as the apex of the temporal bone and foraminae of ovale and rotundum (Peele, 1957). Such over-pneumatization will increase the total surface area of the sinus and make an association with bony dehiscence more likely. The finding of recesses radiologically may therefore be significant, as their presence may indicate fistulous communication from the middle fossa. It remains unproven however, whether patients presenting with spontaneous CSF rhinorrhoea have a higher incidence of excessive sphenoid pneumatization and recesses compared with a controlled population. There are several reasons for this: firstly, few large series have been reported in the literature due to the rarity of this condition. Secondly, the presence of sphenoid recesses are common in the population in general, large numbers would therefore be required to reveal significant differences. Thirdly, as far as I am aware, this question has not been previously addressed.

Hartz (1911), had demonstrated that small communications exist between right and left sides of the nose. In addition, CSF has been shown to communicate with the lymphatics of the nasal mucosa along perineural clefts of the olfactory filaments (Naumann and Naumann, 1977; Messerklinger, 1978). This nasomeningeal tract is confined mainly to the olfactory and cribriform plate regions. Tears of these membranes will undoubtedly lead to CSF rhinorrhoea offering an explanation for their association with nose blowing.

RESULTS

Ten patients with primary CSF rhinorrhoea were studied. The age range was 19–75 years, with the mean age being 46 years. Four patients presented in their 5th decade, two in their 6th and one patient each in the 2nd, 3rd and 8th decades. The female/male ratio was 2 : 1. No patient gave a history of significant head trauma prior to onset of their symptoms. The presence of minor trauma occurring some time previously or which had been subsequently forgotten by the patients could not, however, be totally excluded as an aetiological factor.

Five patients presented with right sided, four with left sided and one with bilateral rhinorrhoea. In eight patients, symptoms of rhinorrhoea were ipsilateral to the side of the fistula demonstrated radiologically, in one patient the side of the rhinorrhoea was contralateral to the bony defect.

Three patients gave a history of a cold immediately preceding the onset of rhinorrhoea. Two patients (20%), gave a history of meningitis, one of whom had had four episodes over a five year period before the diagnosis was finally made. Of these patients, nine underwent surgery having a total of 14 operations, follow-up varied between one month to nine years, with a mean of 3.5 years. Six patients required only one operation; of these, five were extracranial and one was by a craniotomy procedure, the latter contracting post-operative epilepsy. Two patients required two operations, one patient had an initial unsuccessful

craniotomy repair which was later successfully dealt with by an extracranial technique. Another patient required two attempts at extracranial repair before the leak stopped. One patient has had four unsuccessful surgical procedures, three extracranial and one craniotomy repair. She continues to leak, but has not contracted meningitis and remains well. One patient was managed conservatively with bed rest and a lumbar drain, whereupon his rhinorrhoea stopped. He remains well and symptom free after six years follow-up. Extracranial repair techniques had an overall success of 88% compared to that of 50% for craniotomy. Contrast CT scanning was able to precisely identify the site of leak confirmed at operation in seven patients (70%). In two patients contrast scanning lateralized the leak but could not identify the site. In one patient, the contrast scan was unhelpful, this being the patient managed conservatively on bed rest. Contra CT scanning with metrizamide was helpful therefore, in nine out of the ten patients (90%).

Figure 1 shows a defect in the cribriform plate in one of the patients studied. The precise site of fistulous communication in each patient is shown in Table 1.

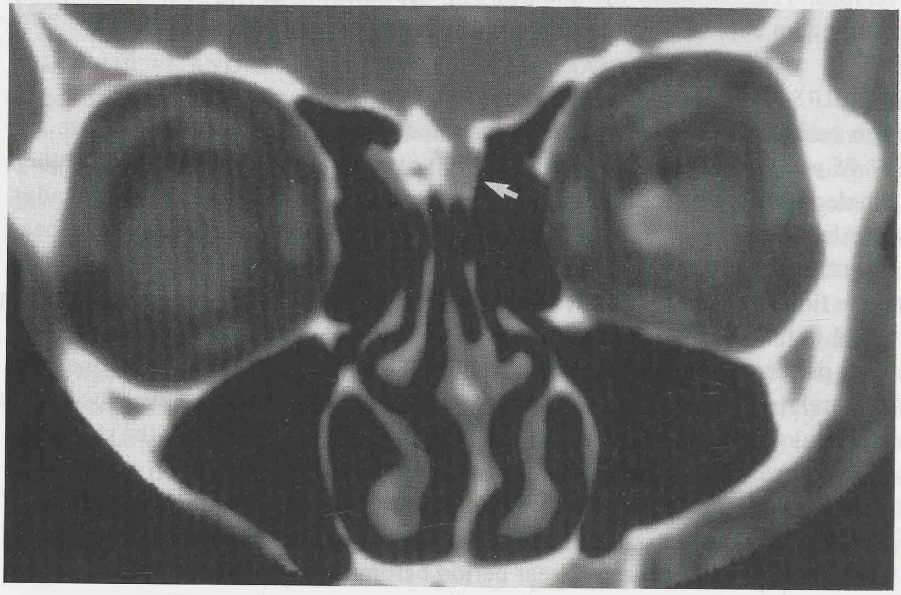

Figure 1. Coronal CT scan of the nose and paranasal sinuses. On the left side, a defect of the cribriform plate can be seen (arrow). At operation this was confirmed to be the site of the CSF leak. CT scanning with larger window settings would, however, have been able to reveal finer anatomical details than shown here. This illustrates, that dialogue between surgeon and radiologist before scanning is often worthwhile in the management of such cases.

Table 1. Precise location of the fistula site found at operation.

case number	cribriform plate/fovea ethmoidalis
1	defect in the right cribriform plate and fovea ethmoidalis
2	defect in the right cribriform plate
3	defect in the left cribriform plate
4	defect in the left cribriform plate
5	defect in the right cribriform plate
	sphenoid sinus
6	defect in the sphenoid roof
7	defect in the right superolateral sphenoid roof
8	defect in a left sided sphenoid recess
9	defect in a left sided sphenoid recess
10	defect in a left sided sphenoid recess

DISCUSSION

A review of the literature shows this to be one of the largest series reported. In 1960 Troland reported that only 33 cases had been described in the literature (Troland, 1960). Since that time many more cases have been reported. Hubbard and colleagues reported on 28 cases of spontaneous CSF rhinorrhoea, of these however, probably only nine cases had primary spontaneous CSF rhinorrhoea (Hubbard et al., 1985). Even in 1969 it had been estimated that the total number reported in the literature was still less than 150 cases (Brisman et al., 1969). The literature therefore contains few large series.

Primary spontaneous CSF rhinorrhoea in this series was found to be twice as common in females. This is consistent with previous reports in the literature (Troland, 1960; Hubbard et al., 1985). Amongst the theories held responsible, either congenital dehiscence or bony atrophy produced by the normal pulsatile CSF pressure pulse are the most likely (Kaufman et al., 1977; Calcaterra, 1980). If the latter theory was the whole explanation it is difficult to explain the fact that it appears to be a condition most prevalent in middle and not old age. In addition, as previously stated bony dehiscences of the skull base are relatively common.

Of even greater significance, 30% of patients in this series, reported that their rhinorrhoea was associated with common cold symptoms and nose blowing. It can be easily understood how such a "Valsalva like" manoeuvre by increasing CSF pressure might lead to dural tears at the site of dehiscence. Whether a female preference for nose blowing compared with the male for sniffing might explain the sex distribution is an interesting but unproven suggestion. The incidence of meningitis in this series was 20% which was less than in that reported by Hubbard and colleagues. Surprisingly, however, the benefit from antibiotic prophylaxis remains unproven.

The role of radiology in the management of such cases is crucial. Metrizamide CT imaging has a low morbidity and may therefore be repeated as required. In this series, contrast CT correctly identified the fistula site in 70% and was helpful in a further 20%. This compares with the usefulness of skull radiography (21%), conventional tomography (53%), in addition, some 33% of radioisotope scans may give false positive results (Ahmadi et al., 1985; Hubbard et al., 1985; Bleach et al., 1988). Leakage of CSF can be contralateral to the site of leakage as shown in one patient in this series.

Despite reports advocating their use (DiChiro and Grove, 1966; Salar et al., 1978; Lantz et al., 1980), radioimaging is not to be generally recommended because of its high rate of false positive results. In addition, it is often unable to identify the precise site of leak, which is so important for planning the surgical approach.

Intrathecal injection of dyes such as fluorescein are also useful for both the preoperative diagnosis (Oberascher, 1988) and peroperative localization of CSF leak (Montgomery, 1973). More recently the non-invasive detection of beta-2-transferrin has been proven as a highly sensitive and specific test for diagnosing CSF leaks, its detection is possible in as little as 1/50th of a drop of CSF (Oberascher, 1988).

Dohlman (1948), reported the first extracranial repair. The various techniques employed in repair have been previously reviewed (Montgomery, 1973; Calcaterra, 1980). The success of extracranial repair techniques used in this category of patients was 88% compared with 50% for craniotomy approaches. It is acknowledged that numbers are small, but being a rare condition large series are difficult to accumulate. There is little doubt, however, that craniotomy carries with it a higher morbidity compared with extracranial procedures. The findings from this study support other reports in the literature advocating a primary extracranial approach (Montgomery, 1973; Calcaterra, 1980; Von Haacke and Croft, 1983; Hubbard et al., 1985; Robson et al., 1989). Ommaya and colleagues (1968) advocated a primary neurosurgical approach. Their main argument against extracranial methods was that sufficient exposure to effect repair of the fistula would not always be possible. This argument does not hold true in practice however. Craniotomy is associated with significant morbidity, the approach to repair is intra not extradural, it causes permanent anosmia and still has a significant failure rate (Ray and Bergland, 1967).

In conclusion, the fact that onset of rhinorrhoea was associated with nose blowing in 30% of the cases, would support the hypothesis that congenital bony dehiscences and meningocoele formation are the most likely predisposing factors. The role of radiology in the surgical management of CSF rhinorrhoea is vital. Finally, the otolaryngologist has much to offer in the diagnosis and surgical treatment of this condition.

REFERENCES

1. Ahmadi J, Weiss MH, Segall HD, Schultz DH, Zee CS, Giannotta SL. Evaluation of cerebrospinal fluid rhinorrhoea by metrizamide computed tomographic cisternography. Neurosurgery 1985; 16: 54–60.
2. Bleach NB, Stanworth PA, Stansbie JM. Spontaneous cerebrospinal fluid rhinorrhoea. An investigative problem. J Lar Otol 1988; 102: 633–635.
3. Brisman R, Hughes JEO, Mount LA. Cerebrospinal fluid rhinorrhoea and the empty sella. J Neurosurg 1969; 31: 538–543.
4. Brockbank MJ, Veitch DY, Thomson HG. Cerebrospinal fluid in the rhinitis clinic. J Lar Otol 1989; 103: 281-283.
5. Calcaterra TC. Extracranial surgical repair of cerebrospinal rhinorrhoea. Ann Otol 1980; 89: 108–116.
6. DiChiro G, Grove AS. Evaluation of surgical and spontaneous cerebrospinal fluid shunts by isotope scanning. J Neurosurg 1966; 24: 743–748.
7. Dohlman G. Spontaneous cerebrospinal fluid rhinorrhoea. Acta Otolaryngol (Stockh) 1948; 67: 20–23.
8. Freeland AP. Non-traumatic CSF rhinorrhoea associated with congenitally malformed ears. J Lar Otol 1973; 87: 781–786.
9. Hartz HJ. The lymphatics of the nose and nasopharynx, with consideration of the general lymphatic system. Trans Am Acad Ophthal Oto-lar 1911; 16: 119–139.
10. Hooper AC. Sphenoidal defects. A possible cause for cerebrospinal fluid rhinorrhoea. J Neurol Neurosurg Psychiat 1971; 34: 739–742.
11. Hubbard JL, McDonald TJ, Person BW, Laws ER. Spontaneous cerebrospinal fluid rhinorrhoea. Evolving concepts in diagnosis and surgical management based on the Mayo Clinic experience from 1970 through 1981. Neurosurgery 1985; 16: 314–323.
12. Johnson ED. Acute frontal sinusitis. Archs Otolar 1948; 47: 165–176.
13. Kaufman B, Nulsen FE, Weiss MH, Brodkey JS, White RJ, Sykora GF. Acquired spontaneous non-traumatic normal pressure cerebrospinal fluid fistulas originating from the middle fossa. Radiology 1977; 122: 379–387.
14. Lang J. Clinical Anatomy of the Nose, Nasal Cavity and Paranasal Sinuses. New York: Thieme Medical Publishers Inc., 1989.
15. Lantz EJ, Forbes GS, Brown ML, Laws ER. Radiology of cerebrospinal fluid rhinorrhoea. Am J Neuroradiol 1980; 1: 391–398.
16. McCoy G. Cerebrospinal rhinorrhoea. A comprehensive review and a definition of the responsibility of the rhinologist in diagnosis and treatment. Laryngoscope 1963; 73: 1125–1157.
17. Messerklinger W. Endoscopy of the Nose. Baltimore-Munich: Urban and Schwarzenberg Publ., 1978.
18. Miller C. Case of hydrocephalus chronicus with some unusual symptoms and appearances on dissection. Trans Med-Chir Soc Edinburgh 1826; 2: 243–248.
19. Montgomery WW. Cerebrospinal fluid rhinorrhoea. Otolaryngol Clin North Am 1973; 6: 757–771.
20. Morley TP, Wortzman G. The importance of the lateral extensions of the sphenoidal sinus in post-traumatic cerebrospinal rhinorrhoea and meningitis. Clin Radiol Aspects 1965; 22: 326–332.
21. Naumann HH, Naumann WH. Kurze Pathophysiologie der Nase ihrer Nebenhohlen (unter Ausschluß des Riechorgans). In: Berendes J, Link R, Zöllner F. Hals-Nasen-Ohren-Heilkunde in Praxis und Klinik, Bd. I: Obere und untere Luftwege I. Stuttgart: Thieme 1977.
22. Oberascher G. A modern concept of cerebrospinal fluid diagnosis in oto- and rhinorrhoea. Rhinology 1988; 26: 89–103.
23. Ohnishi T. Bony defects and dehiscences of the roof of ethmoid cells. Rhinology 1981; 19: 195–202.

24. Ommaya AK, DiChiro G, Baldwin M, Pennybacker JB. Non-traumatic cerebrospinal fluid rhinorrhoea. J Neurol Neurosurg Psychiat 1968; 31: 214–225.
25. Peele JC. Unusual anatomical variations of the sphenoid sinus. Laryngoscope 1957; 67: 208–237.
26. Ray BS, Bergland RM. Cerebrospinal fluid fistula. Clinical aspects, techniques of localization and methods of closure. J Neurosurg 1969; 30: 399–405.
27. Robson AK, Clarke PM, Dilkes M, Maw AR. Transmastoid extracranial repair of CSF leaks following acoustic neuroma resection. J Lar Otol 1989; 103: 842–844.
28. Salar G, Carteri A, Zampieri P. The diagnosis of CSF fistulas with rhinorrhoea by isotope cisternography. Neuroradiology 1978; 15: 185–187.
29. Stupka W. Die Mißbildungen und Anomalien der Nase und des Nasenrachenraumes. Wien: Springer Verlag 1938.
30. Thomson St. Clair. The Cerebrospinal Fluid. Its Spontaneous Escape from the Nose with Observations of its Composition and Function in Human Subjects. London: Cassell & Co Ltd, 1899; 8.
31. Troland CE. The surgical treatment of spontaneous cerebrospinal rhinorrhoea. Archs Otolar 1960; 72: 254–255.
32. Von Haacke NP, Croft CB. Cerebrospinal fluid rhinorrhoea and otorrhoea. Extracranial repair. Clin Otolaryngol 1983; 8: 317–327.

N.S. Tolley, M.D., F.R.C.S.
Senior Registrar
Royal National Throat, Nose and Ear Hospital
Gray's Inn Road
London WC1X 8DA
United Kingdom