Cerebro-spinal otorrhoea

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Perilymph or cerebro-spinal otorrhoea following rapidly on ear surgery, though still a rare phenomenon, has recently become well recognized and has received much attention in otological literature. The discharge is characteristically short-lived and seldom is associated with serious intracranial infection. In most cases the discharge continues no more than a few days and ceases spontaneously, but if prolonged meningitis becomes a considerable risk. This is particularly the case when the fluid passes through a facture line in the petrous bone into the middle ear when it is unequivocal cerebro-spinal fluid otorrhoea.

In the following case the presence of a congenitally abnormal labyrinth has been demonstrated radiologically together with marked enlargement of the cochlear aqueduct.

Case report

Mrs. C. G., aet. 30

This patient was admitted to the National Hospital for Nervous Diseases on 7 April 1970 with the main complaint of watery discharge from her left ear for $2\frac{1}{2}$ years and the following relevant history:

History of left chronic suppurative otitis media

The patient had had left ear trouble in the form of recurrent discharge and deafness since the age of four years.

First episode of meningitis

On 31 October 1967 when she was 35 weeks pregnant. Mrs. C. G. was admitted to another hospital with acute meningitis confirmed by a purulent cerebro-spinal fluid from which was cultured staphylococcus pneumoniae. A swab taken of her left aural discharge was found to culture a moderate growth of coagulase positive staphylococcus pneumoniae and pseudomonas pyocyanea. The patient responded to conservative treatment.

Left radical mastoidectomy

On 11 November 1967 at the same hospital the left mastoid and middle ear were opened and found to contain pus, granulation tissue and cholesteatoma, the tegmen was found to be eroded and granulation tissue present on the middle fossa dura and a radical mastoidectomy was performed.

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Recurrent otorrhoea after mastoidectomy

For the following period of just over two years the left mastoid cavity was never completely dry for more than a month or so at a time.

Second episode of meningitis

On 21 January 1970 Mrs. C. G. was admitted again for meningitis. A lumbar puncture showed turbid C.S.F. and the left aural swab showed a few gram positive cocci on the smear and a profuse growth of pseudomonas pyocyanea and a moderate growth of coagulase negative staphylococcus. However, even on discharge from the hospital, the patient still continued to complain of watery left otorrhoea which was worse when she lay with her left ear undermost.

Further investigation and treatment of cerebro-spinal fluid otorrhoea at the National Hospital

On admission the patient was found to have clear watery otorrhoea and a clear watery fluid was seen to issue from a small opening in what appeared to be a fibrous membrane over the medial wall of the radical mastoid cavity in the region of the promontory. This fluid was shown to contain sugar by means of the Clinistix test and was considered to be cerebro-spinal fluid. The leak became more profuse whenever the patient lay down, especially with her left ear undermost, and whenever the left internal jugular vein was compressed.

VIIIth nerve function tests showed no hearing in the left ear with the right ear masked and the caloric response was absent on the left.

Congenital abnormalities noted were a type I cleft of the soft palate, absence of abduction of the eyes bilaterally owing to previous operations for congenital squint and a large opening for the cochlear aqueduct on X-ray.

X-ray report—skull. Apart from the abnormal petrous bones and the upper cervical anomaly, the skull is normal.

Petrous temporal bones. Tomograms of the petrous bones are abnormal. There is a surgical defect with absence of all the mastoid air cells on the left. In the A.P. views the operation cavity apparently has a thick roof but in the lateral oblique views there is some doubt about this, so there could be a defect in the tegmen. There is evidence of a developmental anomaly of the left bony labyrinth, the vestibule is continuous with an imperfectly formed cochlear and this single cavity also joins a short, wide lateral semi-circular canal (arrows, Fig. 2). There is no sign of a superior semi-circular canal. The floor of the vestibule is deficient and there is a wide opening in the region of the cochlear aqueduct which is most probably the bony opening of the aqueduct (arrow, Fig. 4).

Chest. There appear to be twelve rib-bearing vertebrae. There is a slight mid-dorsal scoliosis, concave to the right, and some articulation of the right first and second ribs anteriorly. The heart and lungs are normal.

Cervical spine (Fig. 4). The adontoid is not seen in the lateral projection. It is noted that on the tomograms of the petrous bones there is a small, separated, midline bone remnant which may represent it. The atlas appears rather high. There is fusion of the next three vertebral bodies, including their lateral masses so that five cervical vertebrae (allowing for the fusion) can be seen. The angle of the first rib is rather steep. Thus there only appear to be exit foramina each side, for three cervical nerve roots.

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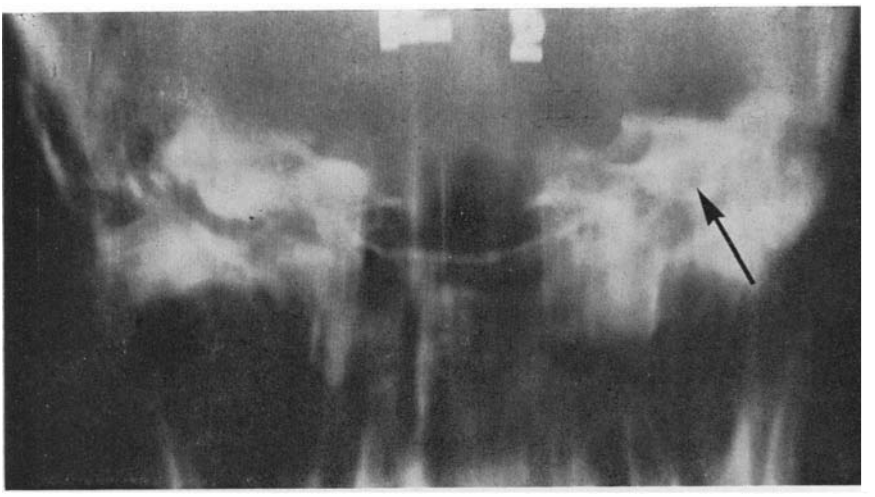


FIG. 1.

Tomogram of skull (fronto-occipital view). Arrow indicates imperfectly formed left labyrinth.

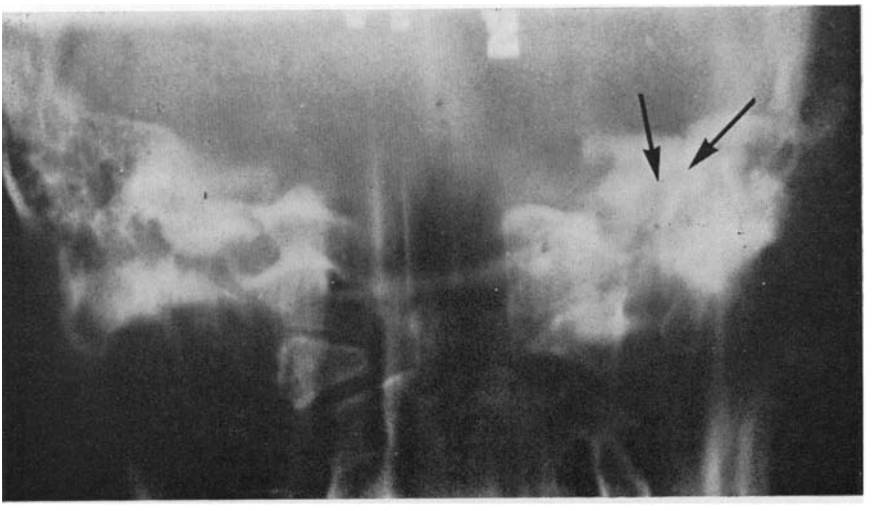


FIG. 2.

Tomogram of skull (fronto-occipital view). Arrows indicate imperfectly formed left labyrinth with deficient floor.

The left mastoid was reopened 10 April 1970 to check the site of origin of the cerebro-spinal fluid leak.

A post-auricular incision was made and the cavity lining stripped. A large area of middle fossa dura had been exposed at the previous operation and now appeared healthy. There were cells containing granular tissue in the mastoid process and above the transverse portion of the facial nerve. The facial spur was lowered, the middle ear cleared of debris, and its posterior wall taken back to

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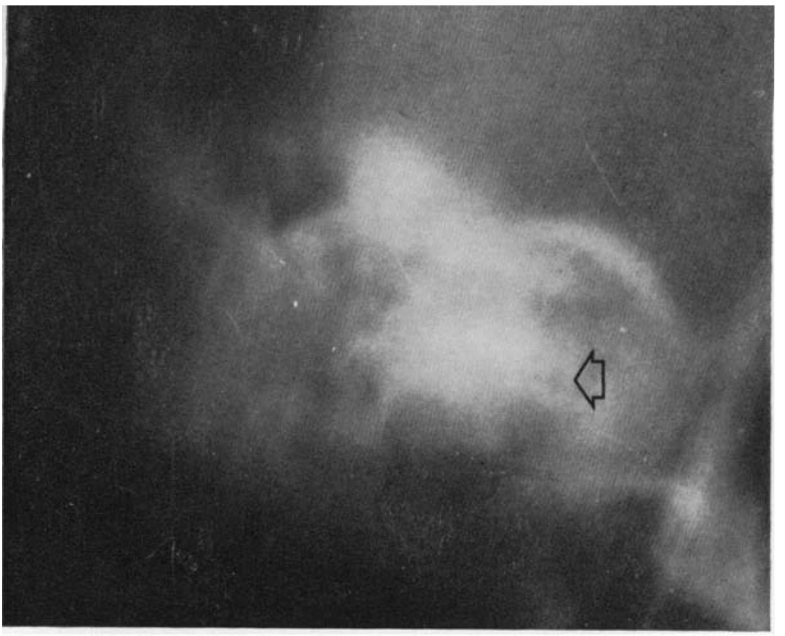


FIG. 3.
Arrow indicates normal appearance of the cochlear aqueduct.

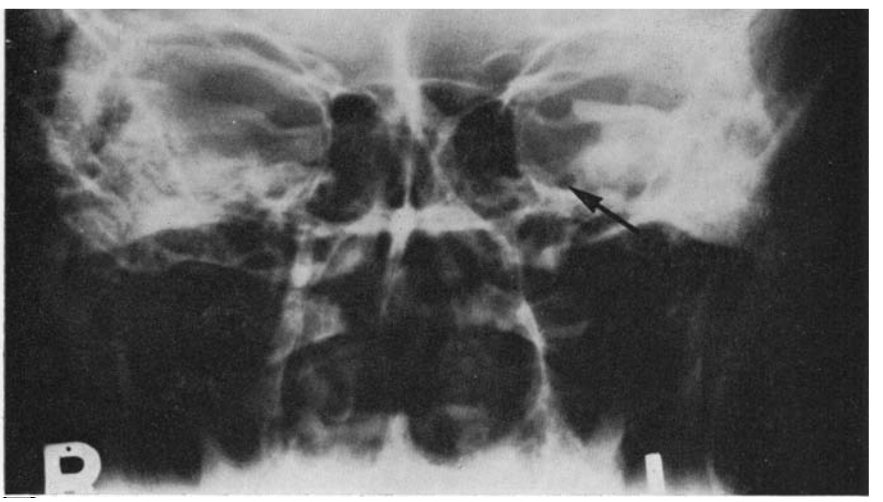


FIG. 4.
Tomogram of skull (fronto-occipital view). Arrow indicates the abnormally wide opening of the cochlear aqueduct in the present case.

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the descending portion of the facial nerve. The C.S.F. was seen to be coming from the oval window in considerable quantity, the promontory was opened widely and the inner ear filled with muscle tissue from the temporalis and held in position by iodoform gauze. Most of the cavity lining was replaced.

The cavity settled rapidly and has remained dry and satisfactory.



FIG. 5.

Radiograph of cervical spine and base of skull (lateral view) showing congenital abnormalities.

Discussion

If there were a free direct path via cochlear aqueduct to perilymph spaces, an infection of the labyrinth might occur with every attack of meningitis. Palva (1969) says that this is prevented by swelling of the soft tissues of the duct during an inflammation of the meninges, but Schuknecht (1970) has recently shown that of seven patients dying of pneumococcal meningitis four showed suppurative labyrinthitis of meningitic origin which appeared to have reached the inner ear via the cochlear aqueduct or the internal auditory meatus. (If the aqueduct is large the reverse, that is, infection of the meninges during attacks of labyrinthitis certainly seems to occur.) Also a profuse leak would occur each time the stapes footplate was opened, for the C.S.F. pressure in a supine patient may be as much as 200 mm. of water, enough to produce a considerable flow into the middle ear.

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The many investigations of the function of the cochlear aqueduct, which was first described by du Verney in 1684, have been diverse in their approach to the problem and similarly show a lack of unanimity in their conclusions. Early workers were: Weed (1914), Wittmaack (1919), Kalefors (1924), Guild (1927), Meurman (1930), and Karbowski (1930).

After examination of the inner ears of patients dying of subarachnoid haemorrhage, Crowe in 1930 considered that blood cells might reach the labyrinth by five routes—along the sheaths of the facial, vestibular and cochlear nerves, along the sheaths of blood vessels entering the labyrinth or through the cochlear aqueduct. Perlman and Lindsay (1939) and later, Holden and Schuknecht (1968) have confirmed Crowe's observations. They showed that out of twelve patients with subarachnoid haemorrhage, in six the cochlea aqueduct was large enough to allow red blood cells to enter the inner ear and thus was large enough to allow direct communication between cerebro-spinal fluid and perilymph. Other investigators have injected test fluids and radio-isotopes into the labyrinth, into the cerebro-spinal fluid or into the blood stream. In the guinea-pig and in rabbits and monkeys (Altmann and Waltner, 1950; Hughes and Chou, 1963), the cochlear aqueduct is normally patent, but it is likely that the human cochlear aqueduct does not allow active communication between cerebro-spinal fluid and perilymph under normal conditions. In the guinea-pig Nakamura (1967) found a 'passive' hydrops from a fall in perilymph pressure after destruction of the cochlear aqueduct. Kley (1951) showed that a negative pressure of 50 mm. Hg. is necessary to draw cerebro-spinal fluid through the *human* cochlear aqueduct, and also that with the aqueduct blocked perilymph continued to form. He injected fluorescein intravenously and found that with the aqueduct blocked it still entered the perilymph. Schuknecht's work (1963) on the cat appeared to confirm these findings yet five days after cochlear aqueduct obstruction in cats Silverstein (1969) found a doubling or trebling of the protein content of the perilymph which returned to normal after four months, the sodium, potassium and glucose content remained unchanged. He concluded that perilymph arose from the cerebro-spinal fluid, perilymphatic vessels and from the VIIIth nerve sheaths, but in the cat cerebro-spinal fluid flowed through the aqueduct and water was absorbed by the veins of the walls of the perilymph space. Chevance (1960) demonstrated that in humans perilymph proteins are like those of the blood, but in the cerebro-spinal fluid they are quite different, but Palva (1969) considers the albumen of cerebro-spinal fluid and perilymph are alike.

The capillaries of the spiral ligament are adequate in numbers to keep up the supply of perilymph and as they have no epithelial layer, permit easy transudation (Graf and Poretti, 1950), a significant finding. Lim (1970) studied the ultrastructure of the perilymph space of the cochlear and showed that free communication takes place between

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perilymph and the underlying vessels. Yet Crifo (1971) has recently shown experimentally that in the horse the free amino acids of the perilymph differentiate this fluid from C.S.F. and blood serum, and that perilymph is not a simple plasma ultrafiltrate.

The consensus of opinion is thus that in the human the cochlear aqueduct is largely vestigial (Ritter, 1965) but that in many cases it allows some exchange between cerebro-spinal fluid and perilymph, but the normal size of 6–7 mm. by less than a millimetre (Palva, 1969 and 1970; Ritter, 1965), is too small to allow the profuse discharge which has occasionally been experienced after injury to the stapedial footplate. For this reason most of these cases with profuce flow are considered to be associated with a congenitally large cochlear aqueduct or an injury of the medial wall of the cochlea. Several authors suggest an association between the enlarged cochlear aqueduct and a congenital abnormality of the labyrinth (Alberti, 1961; Wolferman, 1964; Bennett, 1966; Rice, 1967; and Crook, 1967). Kramer (1971) found that eight out of nine children reported with spontaneous cerebrospinal otorrhoea had major anomalies of the inner ear, but considered this is seldom the case in adults. Waltner (1948) considers that the normal aqueduct is never fully open, that it is very narrow near the scala tympani and that there is a net-like structure preventing rapid flow under normal conditions, thus a diffusion but no direct flow can occur. In the present case there appears to be a clear relation between the large cochlear aqueduct and the profuse flow of cerebro-spinal fluid from the oval window, but in the guinea-pig in whom the aqueduct is normally large a prolonged free flow does not occur. In a series of six guinea-pigs, the foot-plate was widely opened and a flow of perilymph similar to that noted in the similarly treated normal human ear during stapes surgery. The flow slowly decreased over three to four minutes and ceased, nor recurring if the animal was held in the vertical position with its head downwards. In all the guinea-pigs re-opening of the oval window after two, five and ten days produced only a short-lived fluid leak. It would thus appear that in the profuse cerebro-spinal fluid leak which sometimes occurs after stapedectomy not only must there be a wide connection between cerebro-spinal fluid and perilymph spaces, but also absence of the normal connective tissue barrier to free flow which appears to be present in animals and man.

In the present cases there is evidence that the patient's hearing loss on the left side was the result of chronic infection in the ear; there is no evidence that the congenital mixed deafness described by Farrior (1971) was present and for which a patent cochlea aqueduct was postulated as the cause. There nevertheless remains a constant danger of infection of the labyrinth and hence of the meninges if the case is complicated by a chronic suppurative otitis media, and the risk as in the present case, is not eliminated by mastoidectomy. It appears that there was either a

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congenital defect of the stapes footplate (Bennett, 1971), that the footplate was injured at the operation in 1967, or that the perilymph pressure produced a leak from the footplate (Farrior, 1971). Unfortunately, the quantity of C.S.F. leak at the second operation concealed the exact pathology in the fenestra ovalis and the stapes footplate could not be preserved.

Summary

The clinical and operative findings are described in a patient who was shown radiologically to have an enlarged cochlear aqueduct. The similarly large aqueduct of the guinea-pig does not freely transmit C.S.F. as is demonstrated at operation on this animal.

The function of the cochlea aqueduct is discussed in animal and man. In treatment of a long continued C.S.F. leak associated with recurrent complications as meningitis accompanied by severe deafness certain closure of the fistula should be assured at the first operation by wide opening of the promontory and packing of the labyrinth with muscle.

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