Three cases of communicating syringomyelia secondary to midbrain gliomas

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SUMMARY Three cases of midbrain gliomas are described clinically and pathologically. In each case high pressure symptoms were followed by visual disturbance and the onset of syringomyelia symptoms before death. All the patients had hydrocephalus. In one case with concomitant syringobulbia, the syrinx appeared to be due to CSF communicating with the cord cavity through the tissues of the brain stem. In the other cases the communication between the CSF pathways and the syrinx was at the usual site, through the central canal at the obex.

There are many reports of cases in which syringomyelia has been associated with intracranial tumours. Such reports have been prompted by the rarity of the combination, and often by the belief that they shed light on the pathogenesis of syringomyelia. Individual reports have come from Lhermitte and Boveri (1912), Tauber and Langworthy (1935), Kaelber (1952), Kosary *et al.* (1969), Feigin *et al.* (1971), and De Reuck *et al.* (1974). An extensive review is given by Poser (1956), and a reasonable discussion is found in Barnett *et al.* (1973). The different conclusions drawn by various authors serve to illustrate the complexity of the problems. In the cases reported here, there was a clear temporal sequence of events followed by full pathological examination. A probable succession of causative factors is suggested; the time course of the three illnesses is summarised in the Table.

Case 1

This man had no relevant family history and no history of birth difficulties. Headaches developed at the age of 21 years (1963). These became more severe, and at the age of 24 years he was noted to have papilloedema. No neurological signs were found. Ventriculography showed a moderate hydrocephalus with arrest of iophendylate (Myodil; Pantopaque) at mid-aqueduct. In November 1966 ventriculocisternostomy (Torkildsen's operation) was carried out using a small occipital craniectomy.

The patient was well until April 1967 when headaches, aggravated by coughing and movement, recurred. He had attacks of vomiting, drowsiness, and deterioration of vision. These tended to pass off quite suddenly. He also sometimes collapsed without headache; epilepsy was suspected. One such severe attack was followed by diplopia for a week. There was no papilloedema and no other neurological signs. He seemed to improve on anticonvulsant medication, and in August 1967, four months after this treatment started, both kinds of attack ceased.

In November 1967 he was suddenly affected by faintness and a pain in the back of the neck which passed into the head and down into the left shoulder and side of the chest. A second similar attack was followed by pain between the shoulder blades and feelings of prickling and numbness which spread into the left hand, the outside of the left leg, and the whole of the left foot.

Neurological examination showed bilateral optic atrophy, vision J6 both eyes, and fine vertical nystagmus exaggerated by upward and leftward gaze. The remaining cranial nerves, including sensation, were normal. The site of the craniectomy was concave and pulsating, but neck movements in all directions were limited by pain and he preferred to hold the head rotated with the chin to the left. All tendon jerks were brisk without spasticity, the plantar responses were flexor, and sensory testing was normal. Ventriculography showed normal pressure, hydrocephalus, and aqueduct blockage as before. In February 1968 the numbness involved the whole of the left arm and the left side of the chest. On examination, the neck movements had improved but there was an

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Table Time sequence of illness in three cases

Case	Sex	Age at major events (yr)		Ventriculography and first operation	Visual symptoms	Onset of syringomyelia	Death
		Onset of headache					
1	M	21		24	26	26	27
2	F	13		13	13	14	25
3	F	7		7	15	18	18

increase in the nystagmus, which was now rotatory, and impairment of pin prick sensation over the trigeminal distribution, the left side being more affected than the right. There was some wasting and weakness of the left scapular muscles and weakness of the left arm with depressed tendon jerks. The power on the right remained normal with increased tendon jerks. The leg reflexes were brisk and symmetrical with downgoing plantar responses. There was a spinothalamic type of sensory loss from C2 to T9 segments on the left blending into the trigeminal loss. Brain scan with ⁹⁹Tc showed no abnormal uptake; a right carotid angiogram showed no abnormality apart from a persistent right trigeminal artery and hydrocephalus. Pneumoencephalography showed a suboccipital sac and a smooth swelling of the back of the medulla and upper spinal cord. The fourth ventricle filled but air did not go through the aqueduct.

In March 1968 at operation, both cerebellar tonsils were slightly herniated with surrounding arachnoiditis and the cord was soft and swollen. The Torkildsen tube was removed and the spinal cord aspirated. After removal of clear CSF containing droplets of iophendylate, the cord collapsed. The tonsils were then separated, and a catheter was used to maintain a channel from the inside of the fourth ventricle to the subarachnoid space at the C2 cord segment level. Immediately after operation, the nystagmus disappeared and the weakness of the left arm was improved.

In December 1968 the patient had severe backache and weakness of the left leg, without sciatic pain. The inside of the left leg and the perineum became numb, and there was hesitancy of micturition. These difficulties passed with bed rest, but in June 1969 failure of ejaculation occurred. There was no impairment of erection or bladder function and no return of his limp.

In February 1970 a respiratory infection caused several violent sneezes. These made him tingle all over and lose the ability to swallow. On examination there was horizontal and vertical nystagmus. There was some loss of pin prick sensation on both sides of the face, but lower cranial nerve function seemed normal to routine testing; his palate and tongue moved normally, and he could close the larynx; yet he could not swallow.

At operation the previous drainage tube was removed. The median groove of the floor of the fourth ventricle was obliterated by a cystic expansion of the pons and upper medulla. The syrinx was reaspirated and shown to be connected to a large collection of fluid in the brain stem. No communication could be found between the syrinx and the ventricle at the obex. A catheter was passed into the back of the third ventricle through soft unidentified tissue. The wound was closed leaving a large artificial cisterna magna.

After operation his state fluctuated. He was ataxic with episodes of pyrexia and hypertension. The vision varied; sometimes the acuity dropped to J18, and diplopia was sometimes present on looking to the right. Gradually the occipital decompression became tense. Pneumoencephalography showed the tube to be in correct position and patent. A ventriculoatrial shunt was inserted in March 1970, but he deteriorated, and died in April.

At necropsy there was haemorrhage in the posterior fossa. The tube in the aqueduct was then occluded but not removed. The syrinx was inflated using sodium fluorescein in formol saline. As the syrinx filled, fluorescein ran out of the fourth ventricle, into the lateral ventricles, and out of a split in front of the medulla. The Holter valve was patent and sterile, and there was no intracardiac thrombus formation.

After removal and fixation, an injection of diodone was made into the syrinx and radiographs taken (Fig. 1). They showed a communication from the syrinx to a slit in the sagittal plane tracking forward to the front of the medulla. Above this was a cavity in the coronal plane under the floor of the fourth ventricle on the right side. There was a diffuse mass of diodone in the cerebellum in the roof of the fourth ventricle. The fluid passed up to the back of the third ventricle but did not show the fourth ventricle.

Examination of the fixed brain showed hydrocephalus. The hole made by the catheter was just above and to the left of the aqueduct which was indistinguishable in a mass of gelatinous, well-defined grey tumour containing multiple cavities. The tumour invaded the quadrigeminal plate and the upper part of the fourth ventricle and cerebellum. There was a clot in the fourth ventricle and under the

Fig. 1 Case 1. Drawings of postmortem radiographs. The dotted area indicates the cavities outlined after diodone has been injected into the spinal cord. The cavities of the cord were in communication with both the tumour and the aqueduct. There was some escape of diodone from the fourth ventricle but this was not well outlined.

Fig. 2 Case 1. Cross-section of fourth ventricle. The ventricle is full of blood clot and there is soft necrotic tumour in its roof. There is a cavity underneath the ependyma of the floor of the fourth ventricle which communicates with cavities in the spinal cord, and with the tumour and aqueduct.

ependyma of the floor (Fig. 2), forming a cavity on the right side of the pons which tracked downwards and, in turn, communicated with the anteroposterior slit in the medulla. There were multiple irregular cavities all of which were in communication with each other, and were continuous with a well developed syringomyelia cavity which extended transversely across the cord and into the dorsal horn on the left, maintaining this position throughout the cord (Fig. 3). The cavity was smaller, rounder, and more central in the lumbar

cord. There was no communication demonstrable in the usual situation between the syrinx cavity and the lower part of the fourth ventricle.

Microscopic examination showed that the tumour was a well-differentiated infiltrating astrocytoma forming 'young' weakly staining fibrils (Fig. 4). The syrinx had a long established glial wall. It was present as two channels in parallel in some parts while in others there were diverticula from the main cavity which had thinner walls (Fig. 5). At some levels there were ependymal cells in the wall but a separate central canal could be distinguished in the lumbar cord and at some higher levels. There was widespread fluid infiltration of the cord outside the gliotic areas, particularly in the upper cord, with enlargement of the perivascular spaces of Virchow-Robin.

Case 2

A 13 year old girl with a normal birth history presented with attacks of headache and vomiting for a year. These came on always in relation to exertion and lasted for about two hours.

On examination she had a normal intellect. There was papilloedema and optic atrophy. Visual fields were full but acuity was down to 6/35 in each eye. The other cranial nerves were normal. The arms showed dysidiadochokinesia but normal power and tone, except that the left triceps jerk was depressed, and the legs were mildly spastic with downgoing plantar responses. She tended to fall to the right when walking and also to hold the right arm flexed. Sensation was normal elsewhere.

Ventriculography in April 1961 showed gross hydrocephalus with raised pressure. Iophendylate showed appearances compatible with aqueduct stenosis. At operation the cerebellar tonsils were found to be herniated, and a Torkildsen's shunt was inserted. After the operation the vision was unimproved, but the headache and sickness were better until one month later when the occipital decompression became tense. A ventriculoatrial shunt was inserted. She developed a high fever and became completely blind, with drowsiness and increased spasticity. No infection was evident but after antibiotic treatment she gradually recovered perception of light, pupillary reactions returned, and she then went to a school for the blind. In October 1962 she had an unexplained attack of mutism. She recovered from this but it was followed by onset of paralysis of the left arm and weakness of both legs.

The diagnosis was thought to be midbrain glioma but repeat iophendylate ventriculography did not confirm this, and the CSF constituents were normal. Once again she made a partial recovery, and by January 1963 she was able to walk with a caliper. She

Fig. 3 Case 1. Drawings of successive levels from the midbrain downwards.

- a. Midbrain with multiple cavities within the tumour.
- b. At the level of the olive and pyramids—that is, below Fig. 2—showing the main cavity as a vertical slit with a small cavity among the pyramidal tract fibres and another dorsal to the main cavity.
- c. Upper medulla oblongata with cerebellum involved in adhesions around the outside of the medulla and four sizeable cavities within the medulla.
- d. The cavities are adopting a transverse orientation.
- e and f. Multiple cavities apparently of differing ages.
- g, h, and i. Typical syringomyelia cavity with a well formed wall travelling down to the lumbar region.

continued to deteriorate slowly, and by November 1968 there was no movement at either ankle. Pin prick sensation was lost in the right leg up to about mid thigh. In September 1973 she had a severe cold followed by headache and complete paralysis of the legs. Meningitis was diagnosed and treated with penicillin and streptomycin, although again no organisms were isolated. She seemed to be recovering, but in November 1973 she complained of the sudden onset of headache and backache together with numbness of the right hand. Shortly afterwards she became unconscious and died.

At necropsy there was a severe symmetrical hydrocephalus with distension of the third ventricle. There was a tentorial pressure cone and herniation of both cerebellar tonsils around the Torkildsen tube. There was inflammatory exudate and arachnoiditis around the brain stem and medulla. Section of the midbrain showed an astrocytoma invading the quadrigeminal plate, narrowing the upper end of the aqueduct, and blocking the lower aqueduct.

Microscopic examination of the tumour showed abnormal pleomorphic cells with stout glial processes. The nuclei had irregular outlines, pale nucleoplasm, and a fine chromatin network with condensations. In many instances the cells were in pairs or small clusters. The ependyma of the lateral ventricle was largely denuded with the ventricular lining thrown into folds lined by loosely arranged glia showing lymphocytic cuffing of vessels. The lining of the fourth ventricle was interrupted by several nodular proliferations of subependymal glial cells. The medulla oblongata was surrounded by an intense inflammatory cell infiltrate in the pia arachnoid membranes and cuffing of small blood vessels in the subependymal region of the lower part of the fourth ventricle. There was a communication in the usual situation between the fourth ventricle floor and the central canal (Fig. 6).

Section of the spinal cord showed a syringomyelia cavity which was largest in the cervical and sacral segments. From C1–C3 segments the cavity was a distended central canal which had ruptured on both

Fig. 4 Case 1. Tumour from the midbrain.

lateral aspects, producing necrotic extensions involving both lateral corticospinal tracts. From the level of C4 cord segment downwards the cavity ran alongside a normal central canal. In the cervical region a moderately thick gliotic wall was present, but in the lower dorsal and sacral regions the wall was thinner.

Case 3

This girl was 7 years old in 1961 and had a history of six months' unsteadiness of gait, vomiting, and reluctance to use the left arm. On examination there was bilateral papilloedema but normal visual acuity. There was nystagmus in all directions, most marked on the left. A left hemiparesis affected the face, arm, and leg accompanied by ataxia. Ventriculography with iophendylate showed an aqueduct obstruction thought to be from a tumour. At operation the cerebellar tonsils were herniated. At a depth of 15 mm within the cerebellum was a cyst cavity with tumour invading the floor of the fourth ventricle.

She recovered well and was able to go back to school. By July 1969 her gait was worse and she had attacks of impaired vision. There was papilloedema and rotatory nystagmus worse on gaze to the right. A ventriculoatrial shunt relieved the pressure symptoms but she remained dysarthric and ataxic. She died

Fig. 5 Case 1. Part of the wall of the syringomyelia cavity (Fig. 3g). The densest part of the wall shows a transition to a thinner wall apparently dissecting the cord afresh.

suddenly at home in September 1972. For about two months before death she had complained of coldness in the left hand and trunk.

Necropsy showed no significant features outside the nervous system. The Spitz-Holter valve was blocked with fibrinous material. The lateral and third ventricles were dilated. There was a tumour in the midline of the cerebellum and brain stem, mainly on the left side, occluding the aqueduct and most of the fourth ventricle. It had infiltrated the quadrigeminal plate and nodules protruded into the aqueduct. There were areas of necrosis, haemorrhage, and cyst formation particularly in the vermis. There was a syrinx cavity within the cord extending downwards for 13 cm, below which the cord was not examined.

Microscopical examination showed a grade II astrocytoma with numerous microcytic and macrocytic areas. There were numerous Rosenthal fibres

Fig. 6 Case 2, the deep slit in the floor of the fourth ventricle leaving the central canal and providing a communication to the syrinx.

and parts of the tumour were excessively vascular. In the floor of the fourth ventricle, there were three ependymal lined canals of complicated structure with branching and glial nodules protruding into the canal, including one elongated slit which joined the upper end of the syrinx. The upper spinal cord showed a central canal with a large syringomyelia cavity just posterior to it extending into both lateral white columns of the cord. The surrounding cord was oedematous. The canal was sometimes involved in and sometimes separate from the syrinx. At the level of the lower cervical segment there was almost total loss of anterior horn cells and the cavity became multiloculated with extension almost to the surface of the cord.

Discussion

From the histories it is clear that the periaqueductal tumours antedated the syringomyelia in each case. Symptoms of hydrocephalus were present for a long time before they became severe, and it seems probable

that the blockage caused by the tumour was gradual in onset.

In case 1 it seems probable that the Torkildsen's shunt never worked well. Apparently it lowered the pressure but there were only six months free from symptoms, and it seems likely that the intracranial pressure remained somewhat raised, the radiological appearances of bone erosion never resolved, and removal of the Torkildsen's tube in March 1968 did not produce any relapse. There is a distinct failure rate for Torkildsen's operation, even in non-tumourous aqueduct stenosis (Torkildsen, 1960; McMillan and Williams, in press). The second case also had a Torkildsen's operation which became non-functional, and thus had episodes of high pressure.

All these patients had high ventricular pressure which caused some degree of tonsillar herniation. Each patient then had posterior fossa surgery which might be expected to provoke an arachnoiditis. Either tonsillar herniation or arachnoiditis in that region may provide part of the anatomical substrate of communicating syringomyelia.

Unfortunately, although there is no doubt that the syrinx in the first case was communicating, it is unclear exactly where or how CSF was entering the cavity. The failure to demonstrate a central canal at the obex in life or death, macroscopically or microscopically, seems to exclude the common form of communication. The communication between the ventral end of the medullary cavity and the subarachnoid space might be an artefact but seems more likely to be a late event in the disease. It is difficult to visualise how this could be an entry portal for CSF. The presence of cystic cavities in the tumour, and the extent of the subependymal cavitation in the floor right up to and into the tumour, suggest that the entry portal was at the tumour. It seems probable that the hydrocephalus dissected into the upper part of the tumour and the aqueduct, and thence into the tissue planes below. The finding of iophendylate in the syrinx suggests this. The formation of false channels around non-tumourous aqueduct stenosis has been reported (McMillan and Williams, in press), but no report of their extension into a frank syrinx has been found. In most cases syringobulbia is probably formed by fluid breaking upwards through the decussation region from the syrinx to form a cavity in the bulb. There is one case described by John Pearce and quoted by Barnett *et al.* (1973) in which there was cerebellar ectopia but no hydrocephalus. No communication was found between the fourth ventricle and the central canal at the obex. It seems probable that in this case also, the fluid tracked downwards through the syringobulbia into the syringomyelia. She had a communication between a syringobulbia and the upper part of the fourth ventricle at the base

of the superior cerebellar peduncle. The syringobulbia and syringomyelia extended from the thalamus to the lumbar enlargement.

The dissection in our case 1 probably started between April and August 1967. The episode of diplopia was severe and unremitting, and could correspond to a sudden injection of fluid into periaqueductal tissues. The next quiescent period may have been one during which the fluid could not get past the medulla. The decussation of fibres in the medulla makes it a tough zone for fluid to break through, particularly if it is externally supported by a dural constriction or prolapsed tonsils. Few cases of syringomyelia go on to form syringobulbia. Figure 1 indicates the probable zones of hold up. In this case, the fluid seems to have broken through from above downwards in November 1967; pain passed immediately down to the chest. Once in the grey matter of spinal cord, fluid spreads quickly, breaking freely into and out of the central canal (Williams and Weller, 1973). Fluid probably dissected the lumbar region quickly, as was suggested by the further attack later that month which produced numbness in the left foot.

The significance of severe neck pain in syringomyelia and syringobulbia is unclear. It is a common symptom in communicating syringomyelia, not always relieved by operation, and sometimes aggravated by surgery. There are three likely causes: local sensitivity of vessels and arachnoid mater to the displacement of cerebellar ectopia, traction on upper cervical nerve roots associated with downward descent of the cord and, thirdly, tracking of fluid internally into pain evoking regions. Fluid injected into the spinal cord of conscious animals appears to cause discomfort sometimes and occasionally appears to be painful (Williams and Weller, 1973). It is likely, therefore, that neuronal discharges, interpretable as pain, are produced by dissection of fluid into the sensory regions of the cord. In case 1 the cerebellar descent was not great and the upper nerve roots were not stretched. The similarity of the pain to that which he felt in the chest, and its improvement when numbness supervened, suggested an internal disturbance of the posterior grey columns.

Relapses and remissions are a characteristic feature of midbrain tumours as was pointed out by Sarkari and Bickerstaff (1969). They suggested that necrosis and oedema in and around the tumour was a probable cause but the present cases illustrate that, sitting as these tumours do at the confluence of CSF pathways, intermittent CSF obstruction followed by tracking of fluid into the brain tissues *via* the tumour may sometimes play a part.

The mechanism of filling in communicating syringomyelia remains unproven. Although pulsatile pressure changes are almost certainly the most

important, the presence of more continuously raised intracranial pressure is significant in some cases. Newton (1969) reported two cases in which acute hydrocephalic symptoms supervened in the course of syringomyelia. Experimental syringomyelia produced by intracisternal injection of noxious material (Camus and Roussy, 1914; McLaurin *et al.*, 1954) is always accompanied by hydrocephalus. Eisenberg *et al.* (1974) have suggested that the syrinx serves as a conduit for intraventricular CSF to escape to the spinal subarachnoid space. This might have occurred in this case, particularly since there were two breaks into the subarachnoid space. It is also possible that, before such a break opened, the tissues of the spinal cord absorbed some CSF as has been suggested occurs in the brain in hydrocephalus (Milhorat, 1972; Weller and Williams, 1975).

There is some evidence that temporary or permanent pressure differences across the foramen magnum are responsible for filling the syringomyelic cavity, or even causing its production. This craniospinal pressure dissociation may be derived from pulsatile pressure changes (Williams, 1972, 1974). Such pressure differences can readily be envisaged across both the incisura in case 1 and the foramen magnum in all three cases.

It is certain that at some time, in all three cases, the fluid within the brain was under increased pressure, although it need not necessarily have been increased at the time of development of the syrinx. The volume of CSF was certainly increased during the period of development of the syrinx, and since that volume of fluid is incompressible, the intracranial capacitance was altered. The change in capacitance produced by supratentorial cerebral tumour may be concerned in the development of communicating syringomyelia in the cases reported by Kaelber (1952) and Barnett *et al.* (1973). The improvement in occasional cases of communicating syringomyelia after insertion of a ventriculoatrial shunt, even where there has been no evidence of raised intracranial pressure (Krayenbühl and Benini, 1971; Jacobson, 1973, personal communication), seems more likely to be a capacitance effect in as much as shunting lowers the CSF pressure and volume and lessens the pressure on coughing. Capacitance effects can be expected to alter the development of craniospinal pressure dissociation and also the surge of fluid within syrinx cavities. An illustrative fluid analogue and the theoretical concepts are described elsewhere (Williams, 1974).

One feature of case 1 is that the thickness of the glial lining was variable throughout the syringomyelia and syringobulbia. It is notable that the clefts in the medulla were little affected by gliosis. This is the usual finding in a syringobulbia above a syringomyelia and, although age difference may explain

different thickness of the glial lining, in this case it is most probable that the syringobulbia is older than the syringomyelia. A reasonable explanation, therefore, is that the gliosis is proportional not only to the age of the lesion but also to the severity of the forces acting upon the walls. The orientation of the glial fibrils is circumferential, that is disposed as if to withstand a distending force. In communicating syringomyelia the spinal cord is usually flaccid, and distension is likely to be episodic and related to surging of fluid within the syrinx produced by coughing or similar exertion. The magnitude and speed of the pressure pulse in the subarachnoid space has been measured by Williams (1976). It seems likely that this surge is the principal cause of the extension of the syrinx once it passes a certain size (Williams, 1974). Our first patient thought that his sneezes precipitated some change in his condition just before the onset of dysphagia. The distribution of the usual syrinx is consistent with an upward surge which is arrested at about C2 cord segment. Similarly the usual cause of syringobulbia above a syringomyelia, or syringomyelia above a paraplegia, may be related to an upward surge imparting movement to fluid within the cord. There is clearly a zone of increased resistance to upward surge as shown by the narrowing of the communication at the level of the medulla oblongata, and it may be that the syringobulbia is partly protected against upward surge by this.

It is difficult to imagine the validity of Gardner's claim that arterial pulsation originating in the choroid plexuses is responsible for producing distension of a syrinx under any circumstances. Certainly it is unlikely ever to be responsible in three cases such as this with aqueduct stenosis preceding the presentation of the syrinx.

ADDENDUM

Since these cases were prepared for publication the following patient has been diagnosed and treated by Mr J. G. Hamilton.

A 10 year old West Indian girl presented with headache, diplopia, and papilloedema. Investigation showed hydrocephalus although the aqueduct was not well shown; posterior fossa exploration was negative, the cerebellar tonsils were normal in position, and no tumour was found. A ventriculoatrial shunt with a Spitz-Holter valve was used to control the hydrocephalus. Ten months after the initial investigation, she again presented because of progressive weakness of both hands with clawing of the fingers. The legs also became weak, particularly on the right, with absent tendon jerks in all limbs, and marked dissociated sensory loss over the right side of the body. Attempts at myelography with iophendylate were unsatisfactory because the lumbar puncture pressure

was low and entry to the CSF space could not be obtained. Iophendylate ventriculography confirmed the clinical suspicion of syringomyelia by running through the communication at the obex and into a syrinx cavity. This cavity extended down to the conus at L 1-2 vertebral level. The fourth ventricle was low in position and the aqueduct was narrow.

Re-operation on the posterior fossa after some weeks showed a gelatinous tumour in the right cerebellar hemisphere, invading and adhering to the floor of the fourth ventricle. Removal was impossible because of the widespread invasion complicated by arachnoiditis from the previous exploration. Biopsy showed this to be a largely undifferentiated medulloblastoma.

The upward extent of this tumour has not been determined but the similarity to the other two girls is close with the exception that the visual acuity remains J1 in each eye. There is a possibility that the tumour may have gone downwards into the subarachnoid space or even into the syrinx itself. Radiotherapy is being given.

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