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Sudden Death in Case of Syringomyelia

GÖRAN SKÖLD
Department of Forensic Medicine, University of Lund (Sweden)

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Summary. Report of a case of sudden death in a 28 year old man, with known disease of the spinal cord. Autopsy showed hydromyelia, syringomyelia and a moderate intoxication by ethyl alcohol, which in combination are considered to have caused the death.

Zusammenfassung. Bericht über den plötzlichen Tod eines 28jährigen Mannes mit einer klinisch bekannten Erkrankung des Rückenmarks. Bei der Autopsie wurden eine Hydromyelie und Syringomyelie festgestellt. Es bestand eine mittelgradige Alkoholisierung. Die Kombination dieser Gegebenheiten wird als Todesursache angesehen.

Key-Words: Sudden death, syringomyelia — Syringomyelia, sudden death.

In the investigation of the cause of a sudden death in a person following a fall down a staircase or steps while under the influence of alcohol a search is first made for a fatal traumatic injury, above all of the skull, brain, spine or spinal cord. Occasionally, however, the signs of injury that can be demonstrated cannot explain the death which must be ascribed to previous known or unknown disease. Such a case is described below.

Case Report

While on a visit at a friend's flat a man, aged 28, was obviously under the influence of alcohol. Immediately after he had left the flat his friend heard a thud and found the man lying on the next landing. Since he had heard the man go down a few of the steps the latter had presumably fallen from the middle of the flight of steps to the next landing. The man appeared dead, and death was confirmed by the doctor who appeared within a short time. The only anamnestic data available at autopsy were those given by the person's father.

Ten years previously the man had injured the back of his head in a motorcycle accident. Some years later he had begun to be nervous and queer. He had also complained of pain in one arm, which was partly paralysed. At examination the physician had reported that a "cartilaginous mass in the back of the head" had compressed a nerve and that this compression explained the paralysis of the arm. One year before death the man had been operated upon in the back of the head, but after the operation the arm had become completely paralysed. Because of the symptoms of the arm the man had been depressed and had occasionally been admitted to a mental hospital. For some time before death he had drunk heavily.

Autopsy

Gross-examination showed some small abrasions of the skin and subcutaneous haematomas on the head, left arm and trunk, but no signs of injury of the type or severity capable of explaining the death. The most striking finding was that the medulla oblongata and the cervical segment of the spinal cord was markedly swollen, while the upper three fourths of the thoracic part of the spinal cord were moderately swollen. In these areas the tissue was ab-

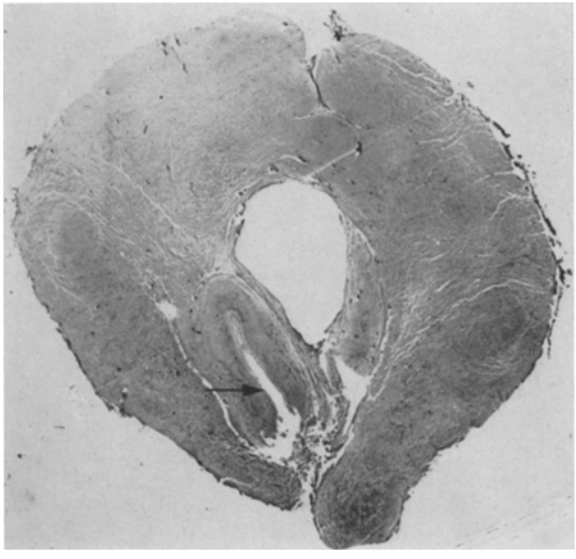

Fig. 1. The enlarged central canal in the center, the syringomyelial cavity to the left (arrow)

vormally soft. The right middle cerebral artery had a small aneurysm without signs of prenious or recent rupture. Other organs showed only stasis.

Examination of the fixed brain and spinal cord revealed two canals in part of the medulla oblongata and the widened part of the cord. Microscopic examination showed that one of the canals extended from the level of the crossing of the pyramidal pathways, down through the entire cervical, and the major part of the thoracic part of the spinal cord. It varied somewhat in thickness, and the wall was made up of mature glia cells with some Rosenthal-like fibers. Outside the actual wall were some protoplasmatic glia which extended out to the lateral funiculi.

The other gross cavity involved only the upper part of the cervical cord. In some parts the wall resembled that of the first, mentioned above, while some areas in the upper part were lined by ependyma-like cells and was seen to merge with the central canal of the spinal cord.

The former cavity was regarded as a syringomyelial cavity; the latter, as an enlarged central canal (hydromyelia).

Chemical examination showed $1.7^0/_{00}$ ethyl alcohol in the blood and $2^0/_{00}$ in the kidney. No barbituric acid derivatives, methaqualone, salicylic acid derivatives or meprobamate could be demonstrated in the blood.

After autopsy the man's hospital records were procured from the neurosurgical and neurological clinics where he had been treated because of his paralysis of the arm. It was found that 18 months before death the man had sought advice for three years' gradually increasing weakness and numbness of the left arm. Subsequent investigation had shown an intramedullary expanding process involving the upper part of the cervical spinal cord. Physical examination, including ECG, had shown nothing else remarkable. The man had been operated upon with extirpation of osteophyte-like formations on the arch of the second cervical vertebra but without any appreciable effect on his symptoms. At review 9 months after the first examination the symptoms of the left arm were largely the same, but weakness of the right shoulder girdle and of the weight-bearing capacity of the right leg had developed. The review revealed also increase in the volume of the cord which now comprised the entire cervical and the major thoracic part of cord. The man flatly refused further neurosurgery. A tumour of the spinal medulla was suspected, but the possibility of syringomyelia had also been considered.

Since the last admission to hospital before death, (about 3/4 of a year) the condition of the man had apparently not appreciably changed.

Discussion

The circumstances under which the accident had occurred suggested that death had been due to a fracture of the cervical spine. This cause of death had also been suspected by the doctor called to the scene.

But autopsy and microscopic examination revealed no such traumatic injury or any other gross injury by the fall down the steps.

Gross and microscopic examination, however, clearly showed a picture of syringomyelia with involvement of the medulla oblongata and the entire cervical, and the major part of the thoracic, spinal cord. In addition, the examination revealed hydromyelia of the upper part of spinal cord.

Syringomyelia is a chronic disease where the symptoms may remain stationary for years or progress only very slowly (Boman *et al.*). Rapid progress has, however, also been reported (McIlroy *et al.*). The death is usually caused by complications of paraplegia or of bulbar paralysis if the cavity extends up to the brain (Hopkins; Merritt; Ralston *et al.*).

Syringobulbia is often associated with dysphagia, dysphonia, respiratory difficulties and fasciculation of the tongue (Duffy *et al.*, Nielsen). Disturbances in the rhythm and depth of respiration may occur with consequent hypoxia, increased pulse rate and changes in blood pressure (Ralston *et al.*). None of these symptoms appear to have occurred in the man in question and the cavity did not extend so far up. A search of the literature available revealed no such cases where syringomyelia alone or in combination with hydromyelia and/or abuse of alcohol resulted in sudden death in the way it did in the present case. It is well known that symptoms of syringomyelia can be suddenly aggravated by bleeding into the cavity (Henneberg *et al.*, Wells *et al.*). Cases with bleeding into the cavity in the medulla oblongata have been described with death occurring within 1 day (Wells *et al.*). Haemorrhages into a syringomyelic cavity can occur spontaneously or after trauma (Perot *et al.*). But no signs of bleeding in the cavity were found in the present case (so that this mechanism can hardly explain the sudden death). Changes in the pressure of the fluid in the cavity are believed to be of significance in the causation of symptoms of syringomyelia (Hauser *et al.*), and could not be excluded in the present case.

It is also known that orthostatic hypotension can cause sudden death. Such hypotension has been described in patients with diseases of the CNS, including syringomyelia (Ebert; Ellis *et al.*, Rosecan *et al.*, Schirger *et al.*).

Orthostatic hypotension is usually characterised by dizziness or feeling of fainting, when standing, impotence and partical loss of sweating. The man in question had probably not been examined for such symptoms, but he had denied dizziness and double vision. The blood pressure had been measured as 150/90, 130/80 and 160/100 on various occasions, though probably always in the supine position. It is very difficult to decide whether hydromyelia had contributed to the clinical picture or the sudden death. Cases with combined hydromyelia and syringomyelia are well known and some authors believe both conditions to be of common origin (Lassman *et al.*, Gardner).

It is thus possible that the man had suffered from orthostatic hypotension secondary to syringomyelia and that this, possibly in combination with the alcohol, can explain the sudden death.

The concentration of alcohol in the blood was not so high as to be fatal in a normal person. It must be regarded as probable that in the present case death was due to alcohol in combination with syringomyelia.

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Dr. Göran Sköld
Department of Forensic Medicine
University of Lund
S-Lund