Spontaneous spinal epidural haematoma.

Etiological considerations

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Summary

Four cases with the typical clinical picture of spinal epidural haematoma are reported. The exact cause of the haemorrhage is generally unknown. A survey is presented of the various causative factors as put forward in the literature so far. The authors point out the possible etiology of this entity and emphasize the need for routine histological examination.

Key words: Spinal epidural haematoma, spontaneous haematoma, etiology.

Introduction

In spite of the fast-growing number of reports in the last ten years, spinal epidural haematoma (SEH) remains an unusual condition of mostly unknown origin.

Bruyn and Bosma[1] found 174 cases in the literature up to 1974 and more than 75 cases have been reported since. In a review of 124 cases of SEH, Piotrowski[2] found a total of 47 cases in which no etiological factor could be identified. It was presumed that the bleeding was spontaneous. The term "spontaneous" has been extended to include those haematomas that occur after minor trauma and physical exertion[1,3]. Other factors that have been related to spontaneous SEH include pregnancy, ankylosing spondylarthritis, arterial hypertension and atherosclerosis[3]. "Spontaneous" excludes, however, those cases in which a significant trauma, for instance a fracture or dislocation of the vertebral column, was responsible[4,5].

Some epidural haematomas develop from systemic disorders, disturbance of blood coagulation, due either to the disease itself[6,7] or anti-coagulant therapy[8-11] and vascular malformation[5]. When iatrogenic, they may represent complications as a result of lumbar puncture, epidural spinal anaesthesia, spinal surgery or anti-coagulant therapy[12].

The purpose of this paper is to report four more cases of apparently spontaneous SEH and to review and analyse the existing literature for etiological factors that might be of clinical importance.

Case report

Case 1

A 6-year-old girl was in good health until February 1983, when she experienced an excruciating pain in the back of her neck. After a while, the pain subsided and later on disappeared completely. Four days later she was awakened from her sleep at 2 a.m. by acute onset of severe nuchal pain radiating into both upper limbs with paresthesias in the fingers. She was able to get out of bed, but within half an hour she could not move either of the lower limbs. In addition, disturbance in micturation and numbness in the legs were noted. There was

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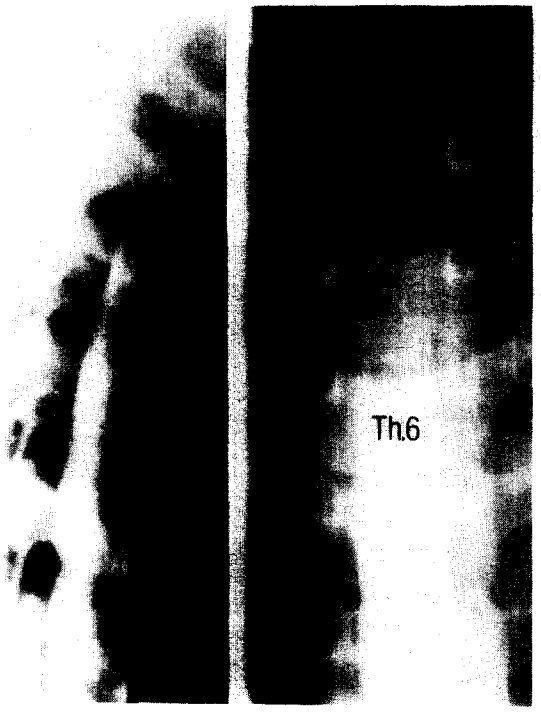

Fig. 1.
Ascending myelography: complete extradural block at Th5.

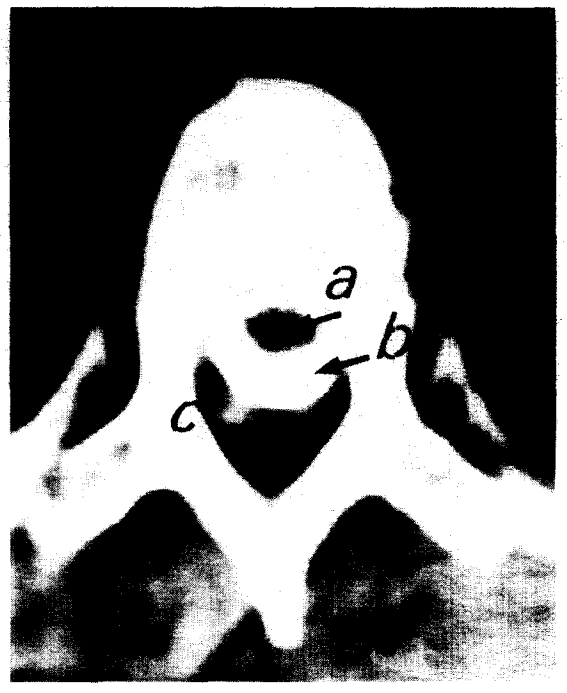

Fig. 2.
CT scan T6 – (a) spinal cord, (b) intrathecal amipaque contrast, (c) dorsally placed extradural mass (haematoma)

no previous history of trauma. Nine hours after the onset of the symptoms, she was admitted to a local hospital. Neurological examination on admission revealed a profoundly flaccid quadriparesis; movement was entirely absent in the legs and hands, and in both forearms only feeble rotation was possible. Sensory testing indicated bilateral anaesthesia below Th2 with hypalgesia and hypesthesia below C6; knee, ankle and plantar reflexes were absent. Plain radiographs of the cervical and thoracic spine were unremarkable. Lumbar puncture revealed the presence of a complete manometric block on jugular compression and amipaque myelography showed a complete extradural block at Th5 (Fig. 1).

Computerized tomography (CT), performed immediately after myelography, revealed a dorsally placed extradural mass at several levels of the upper thoracic and lower cervical spine, with a smooth anterior concave border, which appeared to extend to the C6 level (Fig. 2).

A laminectomy from C6 to Th5 was performed approximately 18 hours after the onset of neurological deficit. A large extradural haematoma was found on the posterolateral aspect of the spinal cord, extending from the inferior margin of C5 to the inferior margin of Th6. The lesion was removed and the dural sac immediately resumed normal pulsations.

Histological examination of specimens obtained during surgery, showed no evidence of tumor or arterio venous malformation, whilst hematological investigation in the post-operative period, excluded a bleeding disorder diagnosis.

There was no immediate post-operative neurological improvement and three months of follow-up showed only functional improvement of wrist and hand flexors.

Case 2

A 75-year-old man, whilst eating, was suddenly struck by a severe pain in the back of his neck, radiating into his right arm. Within an hour, he could not move his legs and there was weakness in his right arm. Apart from a laminectomy for a lumbar herniated disc in 1981, there was no significant medical history of recent trauma or record of drug-taking. Two hours after the onset

of the symptoms at 1 p.m. on 12th February 1983, he was admitted to a local hospital. On neurological examination, he was alert and well orientated. There was a flaccid paraplegia; knee, ankle and plantar responses were absent. The upper extremities showed mild weakness of the left triceps and hand intrinsics, moderate weakness of the right forearm and hand, in addition to severe weakness of the right triceps. The biceps appeared to function normally. A sensory deficit was evident below C6. General examination was unremarkable as were the usual laboratory tests.

Radiographs of the cervical spine disclosed marked osteoarthritic and spondylotic changes, particularly at the C6-C7 level. Cervical myelography revealed an incomplete extradural block, extending from C5 to C8. Following this procedure, the patient suffered a general seizure, due probable to the Amipaque contrast medium which had been injected in to the cysternal subarachnoid space.

After transfer to the Neurosurgical Department, an emergency laminectomy from C4 to Th1 was carried out, approximately nine hours after the onset of the neurological signs. An extradural haematoma was exposed, extending from C4 to Th1. Surprisingly enough, pulsations of the dural sac had not completely disappeared. After removal of the clot, the dural sac regained its normal pulsations. No abnormal vascularity or bleeding point was noted.

On the third post-operative day, the patient was transferred back to the local hospital and there followed a period of dramatic improvement. Neurological examination there revealed only a mild weakness in the right arm and leg predominantly distal. The patient was symptom-free three months following surgery.

Case 3

A 52-year-old man was first admitted to the hospital on 3rd May 1980. Two days before admission, without any apparent predisposing cause, he suddenly complained of severe nuchal pain, radiating into both upper limbs. Some 30 hours after the onset of the symptoms, the patient experienced weakness in the left arm and numbness of the fingers, followed several hours later by weakness in the left leg. There was a previous history of chronic bronchitis and kidney stones. He had also been treated for paroxysmal supraventricular tachycardia and atrial septal defect and had undergone long-term anticoagulant therapy (Acenocoumarol).

He was found to be alert, but held his neck stiffly flexed because the slightest movement of the neck caused intense pain which spread out into the left arm. Neurological examination showed a weakness in the left arm and leg, reduced pain and touch sensations extending below T4 on the right, hypo-active tendon reflexes on the left side, and absent plantar reflex bilaterally.

The results of routine laboratory tests were normal. Prothrombin time was 9% of normal, cisternal CSF was clear, containing 58 WBC, 59 RBC and 15 mg/100 ml of protein.

Amipaque myelography demonstrated an epidural block at the level of C5. Following myelography, a laminectomy was performed from C4 to C7, and on removal of the laminae, a blood clot was found in the epidural space, compressing the dural sac from the left side and at the C5-C6 level from the dorsal aspect. The haematoma was removed. The dura mater was not opened. Histological examination revealed a haematoma devoid of any significant structures. It should be noted, however, that no extensive serial section of the clot was performed. Following surgery, there was gradual improvement of the neurological deficit. Six months after discharge, the patient had regained normal strength in his arm, but some residual weakness of the left leg after walking long distances remained.

Case 4

A 70-year-old woman, whilst lying in bed, felt a sudden sharp interscapular pain radiating into both arms. The pain persisted and in the morning of the following day, she experienced weakness in the left leg. Over a period of several hours, numbness and weakness spread into both extremities.

There was a previous history of vascular hypertension and mild diabetes mellitus. She had been placed on long-term anticoagulant therapy (Acenocoumarol) after a hysterectomy, complicated by venous thrombosis and pulmonary embolism. The next day, after admission to a local hospital, moderate leg weakness pro-

gressed to a complete flaccid paraplegia. There was a diminished pain sensation below T4, absent reflexes and painless urinary retention. General examination was unremarkable. Laboratory tests included a prothrombin (Quick) time 7.5% of normal. Queckenstedt's manometric test revealed a block of the spinal canal. CSF was macroscopically clear. Myelography with Duroliopaque (ethyl-monoiodostearate), introduced cisternally, disclosed a total block at Th2.

Upon transfer to our department on 7th November 1977, three days after onset of the symptoms, an emergency laminectomy was performed from Th1 to Th3. The dura was completely surrounded by a large blood clot which was, however, more prominent on the dorsal and left lateral aspect. After removal, no abnormal vessels or masses were detected. Post-operatively, the patient remained incontinent with severe loss of all sensory modalities below Th5-Th6 and a spastic paraplegia.

Discussion

The large number of reports detailing the features of SEH is indicative of the interest and uncertainty surrounding this subject. From the current data, it seems likely that SEH is a disorder with multiple etiological possibilities. In discussing the possible etiologidal factors contributing towards a reliable classification of SEH, the following aspects deserve consideration from an etiological standpoint.

1. Trauma

Haemorrhage in the epidural space following spinal injury seems a simple etiological explanation of SEH. It may occur in vertebral fractures as a consequence of tissue disruption in the area of the fracture[13], but is more common in cases with no history of significant trauma. An epidural haematoma following injury is rare and is seldom large enough to serve as a space-occupying lesion[4].

It was found in 1 out of 200 cases (0.5%) of spinal injury with fractured vertebrae[13]. In a series of 94 fatal spinal injuries, only 3.8% epidural and 7.5% combined subdural and epidural haemorrhages were reported; almost none were large enough to compress the spinal cord[14].

In an extensive review of 174 cases of SEH, 15 (9%) cases of traumatic origin were reported[1]. Unlike the minor clinical importance of the traumatic cases of SEH mentioned above, the number of SEH in obstetric trauma of the cord appears to be significant. Coutelle[15] in an autopsy study of 155 neonates, noted SEH in 131 cases (85%), and Towbin[16] was of the opinion that epidural haemorrhage in the neonate is a cardinal etiological factor in spinal cord injury. In a considerable number of autopsies, epidural extra-vasation of blood was found, often combined with damage to the adjacent vertebral structures.

Some reports mention that iatrogenic trauma can occur following lumbar puncture and spinal anaesthesia. The large size and thin walls of the epidural veins, make them susceptible to needle puncture, and it is indeed possible that a needle could tear an epidural vein and thus caus a haematoma.

SEH can be caused by a spinal tap alone or in conjunction with anti-coagulant therapy. The risk of bleeding is increased if the lumbar puncture is traumatic. Even if anti-coagulation is initiated within one hour of the spinal tap, there is an increased tendency to bleeding[12].

In patients whose symptoms were preceded by only seemingly unimportant episodes, such as voiding, twisting and lifting weights and other minor traumas, the presumed pathological mechanism would involve transmission of increased intra-abdominal and intra-thoracic pressure to the valve-less, thin-walled epidural veins, thus initiating haemorrhage. It seems unlikely, however, that the elevation of epidural venous pressure associated with normal activities of daily life, is sufficient to break the walls of vessels unaffected by some other pathological process.

2. Vascular malformation

In a number of reported cases of histological examination, a spinal angioma (malformation) or phlebectasia was observed and presumed to account for the haemorrhage[5,17-28]. Pia[29] observed in his own material 2 SEH cases in a total of 54 epidural angiomas. It is possible that angiomas cause these lesions more often than has been suggested in the literature[17,21,30]. Not all reported cases of SEH have been subjected

to pathological examination. The haematomas are generally more extensive than would be expected from the area exposed at laminectomy, thus making it difficult to study them adequately.

Cryptic angiomas are significant in the etiology of intra-cerebral haematomas and were first reported by Margolis[31]; he drew attention to the fact that clear recognition is hampered by the smallness of the malformation and its frequent partial destruction by the haemorrhage. Rupture of these cryptic vascular anomalies, similar to those recognised as forming the etiology of intra-cerebral heamatomas, must be considered a cause of epidural bleeding. Other authors have described this kind of microscopic lesion resulting in a SEH[23,24]. It is only by careful serial sectioning of the blood clot removed during surgery, that we can increase our knowledge of the pathology that lies at the base of the disorder.

Old-age, associated with hypertension and atherosclerosis is sometimes presented as a likely cause of epidural vascular degeneration, with subsequent rupture[19,25], although this does not explain its occurrence in normotensive cases and in the young, nor does it explain the fact that the bleeding is presumed to occur initially in the epidural venous plexus.

3. Increased bleeding tendency

Incidentally, systemic diseases[6,7] have also been associated with SEH. No clues, however, as to the etiology of SEH can be derived from these reports, although it is generally thought that an increased tendency to bleeding itself contributes to the occurrence of the disorder[1]. Anti-coagulant therapy has been recorded in an increasing number of SEH cases[8,9,11]. In an analysis of 45 cases, however, in which the SEH had been attributed to anti-coagulant therapy, no unequivocal causal relationship could be established[1]. In one case of SEH attributed to anticoagulants, it was noted that the haemorrhage occurred 5 days after cessation of the anti-coagulant therapy[2].

Prothrombin time determination has been stressed in the literature; in those cases in which the prothrombim time was available, values ranged from 7.5% to 60%, whilst the majority ranged from 10-15%. These are normal figures and are difficult to correlate with spontaneous haemorrhage. They suggest that SEH is possible in the apparently optimal drug dosage. It is not possible, however, to draw any reliable conclusions from these data on the causative significance of anticoagulant therapy.

4. Idiopathic

In a considerable number of cases, no identifiable cause for SEH was evident; the patients presumably bled "spontaneously". The term "spontaneous", although used widely in the literature, does not always seem to be used in the same sense. By definition, a spontaneous spinal epidural haematoma (SEH) cannot be related to a bleeding disorder, vertebral fracture, or vascular anomalies etc.[1,2] Other associated factors, however, such as hypertension, artherosclerosis, minor trauma, pregnancy and ankylosing spondylitis are generally included in the term "spontaneous"[3]. Tsai[30] introduced the term "idiopathic" to segregate those cases in which there was no obvious cause for the haematoma.

Clinical picture

The clinical picture is mostly of an unusual uniformity. In all our cases, an apoplectiform onset of back and radicular pain was the hallmark and first symptom of SEH. A transverse cord syndrome usually occurred simultaneously with the pain, but may have had a later and more gradual onset. Recovery, following a complete transverse cord lesion, is only possible if there is a minimum of delay between the onset of the symptoms and the start of surgical treatment[32]. Early recognition is the first essential, after which myelographic confirmation and decompressive laminectomy should be carried out within the shortest time lapse possible.

Conclusion

The etiology of spinal epidural haematoma is frequently obscure. From the existing data, it appears likely that SEH is a disorder with multiple etiological possibilities. Severe trauma, anti-coagulants, bleeding diathesis and vascular malformation have all been associated with this kind of haemorrhage; it occurs mostly, however, without any apparent cause.

Expansive epidural haematoma is due only occasionally to injury of the spine, with or without fracture. Only in the neonate can trauma be considered to be an important etiological factor.

In most cases of SEH at present, the etiology of the bleeding remains unclear. The haemorrhage is presumed to occur spontaneously. In the event of a spontaneous SEH, careful consideration should be given to a possible vascular anomaly, and neuro-surgeons dealing with such cases should be constantly on the alert for this.

Careful serial sectioning of the haematoma removed at surgery, should be performed in all cases, in order to identify a possible vascular malformation.

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