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Spontaneous spinal extradural hematoma in children

Report of three cases and a review of the literature

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Abstract. Three young children who presented with acute backache and rapidly progressive neurological deterioration were investigated and found to have spontaneous spinal extradural hematoma (SSEDH). They were operated on soon after the diagnosis had been confirmed. The present report highlights the fact that in children progressive weakness of nontraumatic origin should be investigated thoroughly and as early as possible. Patients with SSEDH should be operated on immediately, as this is a curable condition. The pertinent literature has been reviewed.

Key words: Spontaneous spinal extradural hematoma – Spinal hematoma – Quadriparesis – Paraparesis

Spontaneous spinal extradural hematoma (SSEDH) is a rare condition. Only 17 cases have been reported in children under 12 years. It is usually found in the cervicodorsal region in children [2, 14]. Back pain and progressive weakness of limbs with disturbance of bladder function is the common clinical picture [15].

Early diagnosis using imaging modalities and surgical intervention leads to a dramatic improvement in neurological functions in children. The management of three children with progressive weakness of nontraumatic origin is discussed in this report.

Case reports

Case 1

An 11-year-old girl was admitted with sudden pain in the nape of the neck radiating to the left shoulder and upper limb, and rapidly progressive weakness of both upper limbs followed by weakness of the trunk and both lower limbs. Two hours later, she developed retention of urine. Her neurological deficit progressed to a total loss of function below the C4 level over the next 16 h.

The baseline investigations, including a complete coagulogram, were normal. Plain X-ray films of the cervical spine did not reveal any abnormality. Iohexol myelography showed a partial extradural block from C5 to C7 posterolaterally on the left side. Cerebrospinal fluid (CSF) examination was within normal limits. Computed tomography (CT) scan showed a high attenuation lesion, most probably an epidural blood clot, in the same location (Fig. 1).

Exploration was performed 24 h after onset of the symptoms. An epidural hematoma was found after removal of C4–C7 laminae and a black jelly-like clot was removed. Histological examination revealed an organizing hematoma. No vascular malformation was seen. The patient showed very little improvement in the immediate postoperative period and only a slow recovery thereafter. Follow-up examination after 8 months revealed grade 4/5 power in both upper and lower limbs. Sensations and bladder function recovered to near normal.

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Case 2

An 8-year-old boy had slipped in the bathroom about 1 month previously. Eighteen hours later, he started having low back pain and developed weakness of both lower limbs and urinary incontinence on the 3rd day.

Examination revealed normal higher mental functions and cranial nerves. Power in the upper limbs was normal. He had grade 3/5 power in the right hip and knee, and grade 0/5 in the ankle and toes and the left lower limb. Joint position sense was impaired below the anterior superior iliac spine. Other sensations were normal. Deep tendon reflexes were absent in the lower limbs, as were plantar reflexes, which could not be elicited. Abdominal reflexes were present. Plain radiography of the dorsolumbar spine was normal. Iohexol myelography showed a partial extradural block at the L3–4 level. CT scan showed a hyperdense epidural mass causing an incomplete block at the D10 level (Fig. 2).

Exploration was performed 50 days after the onset of symptoms. At laminectomy from the D10 to L4 level, an epidural clot was found and evacuated. Histological examination revealed an organizing hematoma. No vascular malformation was seen. There was no significant return of function in the early postoperative period. Follow-up examination after 9 months revealed grade 4/5 power in the lower limbs with near normal bladder control.

Case 3

A 5-year-old boy, who had received a vaccination for diphtheria and tetanus 3 days before admission, presented with inability to bear weight on both lower limbs, retention of urine and progressive weakness of both the wrists and hands for 1 day prior to admission. He also had difficulty in breathing as well as pain in the upper back and neck.

Examination revealed normal higher mental functions and cranial nerves. The patient had grade 3/5 power in both upper limbs and 0/5 power in both lower limbs. He had intercostal muscle

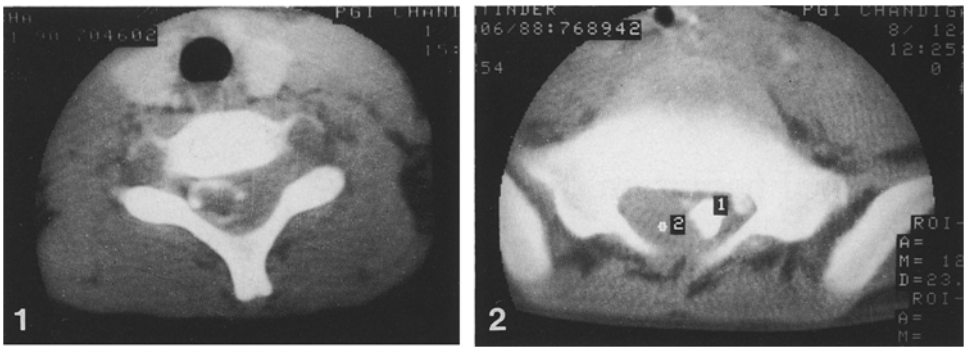

Fig. 1. Iohexol computed tomography (CT) of cervical region showing extradural block from C5 to C7

Fig. 2. Iohexol CT of lumbar region showing incomplete extradural block due to high attenuation lesion

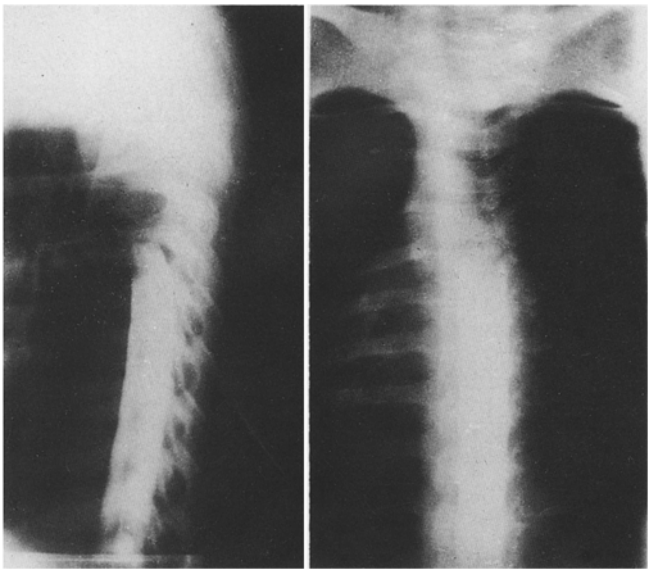

Fig. 3. Iohexol myelogram showing extradural block at D4

paralysis with bladder involvement. Sensations were lost below the D4 level. A diagnosis of acute Landry-Guillain-Barré syndrome was considered and the patient was put on ventilatory support for 5 days. Myelography performed at this time showed an extradural block at the D4 level from the posterior aspect (Fig. 3). As the patient had clinical involvement from C5 to D4 levels, a laminectomy from C5 to D4 was performed. An extradural hematoma, 1.5 cm thick, was seen extending from the C5 to D4 level and was evacuated. Histological examination revealed an organized hematoma. No vascular malformation was seen. The patient improved in the immediate postoperative period. Power in the upper limbs improved to grade 4/5 and in the lower limbs to 2/5. Sensations and bladder function returned to normal.

Discussion

SSEDH is a rare condition and only just over 100 cases have been reported to date [14]. It can occur at any age from 6 months to the 8th decade [2], but is most frequently seen in the middle aged or elderly [14]. The condition is uncommon in children and only 17 such cases have so far been reported in the literature [2]. It is commonly seen in the cervical and dorsal regions in childhood [14].

Though Jackson (1868) is credited as first reporting this entity, it was Blauby who first described this clinical condition in 1809 [9]. Johnston [2] was the first to report SSEDH in a child. SSEDH can be produced by: (1) rupture of valveless veins in the internal vertebral plexus with the slightest change of posture such as during sleep, turning movements, coughing, straining or due to Valsalva's maneuver [2]; (2) bleeding from abnormal vessels, e.g., angiomas [2, 4] or arteriovenous malformations (AVMs), whether cryptic or manifest [2, 11, 13]; (3) anticoagulant drugs, blood dyscrasias, e.g., hemophilia, leukemia, polycythemia vera, macroglobulinemia [2]; (4) metabolic changes during pregnancy [15]; (5) acute infections [2]; and (6) whooping cough [2].

Most authors consider that venous bleeding leads to the formation of SSEDH [14]. Pathological alterations of the vessel wall in vascular tumors and AVMs are known to lead to spontaneous and even multiple bleeding episodes and histological evidence of this bleeding can be seen in the vessel walls of AVM during microscopic examinations [13].

Cooper [3] and Schwartz et al. [12] concluded that "in the absence of gross trauma or bleeding diathesis, it is conceivable that this type of hematoma is related to an organised vascular abnormality or to transmission of increased intra-abdominal or intra-thoracic pressure to the epidural plexus initiating bleeding. However, it seems unlikely that this type of increased pressure would initiate hemorrhage in normal vessels." However, Foo and Rossier [6] found vascular malformations in 6 out of 158 and Müller et al. [11] in 4 of 5 cases of nontraumatic SSEDH.

Bleeding commonly occurs in the dorsal region and may spread up or down or even along the whole length of the spinal canal producing a rapid deterioration of neurological function. Spontaneous resolution of SSEDH, though extremely uncommon, does occur and results in clinical improvement [1, 5, 7]. There is an explanation for spontaneous recovery. Emery and Cochrane [5] have postulated that the oil-based contrast agent which they used acted by increasing the intrathecal volume dispersing the liquid component of hematoma, thereby relieving compression. Alternatively, they suggested that in cases where SSEDH had solid and liquid contents, removal of spinal fluid during myelography may promote dissipation of liquid hematoma into the epidural space.

About 35% of patients undergoing surgery for SSEDH recover their ability to walk [10]. The longer the delay in surgery, the worse are the results. The ultimate recovery depends on the duration, rapidity of cord compression and promptness of surgical decompression [10]. In contrast to the view of Karvounis et al. [8] that no recovery could be expected if decompression was per-

Table 1. Compilation of reports from the literature on patients less than 12 years of age [2]

Author	Age – sex	Site	Cause	Mode of onset	Deficit	Myelo/ CT	Oper- ation	Results
Johnston 1938	5 years-M	D8-12	Spontaneous (hemangioma)	1 year recurrent	Paraplegia	–	–	Autopsy
Shenkin et al. 1945	6 months-F	D7-10	Spontaneous	14 days	Paraplegia	D5-10	+	Cured
Carrea et al. II 1954	4 months-M	D9-L2	Birth injury	Intermittent	Paraplegia	L2	+	No improvement
Jones and Knighton 1956	12 years-M	C6-D2	Hemophilia	12 h	Paraplegia	–	+	No improvement (died after 6 months)
Maxwell and Puletti 1957	4 years-M	D2-4	Spontaneous	Acute	Paraplegia	–	+	Cured
Scott 1958	9 years-F	D1-4	Spontaneous	6 months	Paraplegia	–	+	Cured
Jackson 1963	1 year 3 months-F	D1-5	Spontaneous (scarlatina)	3 h	Paraplegia	–	+	Cured
Lepintre et al. I 1956	8 years-F	C7-D1	Spontaneous	12 h	Quadriplegia	–	+	Cured
Lepintre et al. II 1957	6 years-M	C7-D2	Fall (hemophilia)	2–4 h	Quadriplegia	–	+	No improvement
Lepintre et al. III 1969	1 year 6 months-F	C3-7	Spontaneous (minimal trauma)	7 days	Quadriplegia	–	+	Cured
Rubello and Dastur 1966	11 years-M	C3-7	Spontaneous	Acute	Quadriplegia	C3	+	Cured
Hehman and Norrell 1968	1 year 9 months-M	C3-D9	Spontaneous (minimal trauma)	5 days	Quadriplegia	–	+	Cured
Posnikoff 1968	2 years 6 months-F	C5-D5	Spontaneous	5 days	Quadriplegia	–	+	Cured
Amyot et al. 1969	1 year 4 months-F	C5-7	Fall	Weeks	L arm weakness partial block	–	+	Cured
Valladeres 1972	2 years 6 months-M	D3-5	Spontaneous	12 days	Paraplegia	D6	+	Cured
	2 years 9 months-F	C7-D3	Spontaneous (minimal trauma)	2 days	Quadriplegia	C7-D3	+	Improved
Present series	11 years-F	C4-7	Spontaneous	2 h	Quadriplegia	C5-C7	+	Improved
	8 years-M	D10-L4	Spontaneous (minimal trauma)	50 days	Paraplegia	D10-L4	+	Cured
	5 years-M	C5-D4	Spontaneous	3 days	Quadriplegia	C5-D4	+	Improved

formed 30 h after onset of symptoms, Müller et al. [11] felt that there was no relation between the degree of recovery and localization of the hematoma, patients' age or even the delay in surgery. One of their patients showed a good recovery following surgery 18 days after the onset of symptoms. We had similar experiences in cases 2 and 3 with good recovery following surgery after a delay of 50 days and 55 days, respectively.

Reviewing the available literature on cases of children under 12 years of age, we came to the conclusion that a good recovery occurs in children irrespective of age, duration of cord compression, rapidity of onset of symptoms or the force of compression (length and thickness of clot) (Table 1).

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