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Acute spontaneous spinal subdural haematomas in a patient with essential thrombocythaemia

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Sirs: Spinal subdural haematomas are rare, and most of them develop following trauma [11]. Spontaneous spinal subdural haematomas are far more uncommon and to our knowledge they have never been reported in patients with essential thrombocythaemia.

Essential thrombocythaemia (ET) is a chronic proliferative disorder of the bone marrow that is characterized by increased production of platelets (PLT) [4]. In contrast to thrombocytopenia, a well-known entity predisposing to spontaneous cranial subdural haematomas, thrombocythaemia manifests mainly with thrombotic events [1, 4]. There have been very few reported cases of spontaneous intracranial subdural haematomas complicating ET [2, 3, 9, 12] and only one case of spinal extradural haematoma [14].

We report a patient with known ET who presented with acute extensive spontaneous spinal subdural haematomas.

A 60-year-old female with a history of E. T. was admitted with acute onset of severe headache, midscapular pain, and back pain radiating to both buttocks. No trauma was reported, not even a trivial one. Physical examination revealed neck stiffness, bilateral Laségue sign and increased deep-tendon reflexes in the lower limbs, but otherwise muscle power, sensory and bladder functions were intact. Plantar reflexes were flexor and her level of consciousness was normal. Her platelet count was 619.000, and the INR was 0.87. RBCs, WBCs, and biochemical tests were all within normal limits. A coagulopathy profile including activated protein C, protein S, antithrombine III, factor V, factor II, lupus anticoagulant, and anticardiolipine antibodies was normal. ANAs, anti-dsDNA, ANCA, RF, CRP, C3, C4, values were also negative/normal.

Examination of bone marrow smears disclosed a large number of giant megakaryocytes. Platelet aggregation studies were abnormal showing markedly reduced response to ADP, collagen, and epinephrine. After CT of the brain that did not reveal any abnormal findings, a lumbar puncture followed that produced an haemorrhagic CSF with xanthochromic supernatant after centrifugation. Eight days after the onset of symptoms MRI of the spine showed a longitudinal mass lesion within the ventrolateral subdural spinal space extending from T3 to L5 (Figs. 1 and 2). The lesion gave a heterogeneous high signal on both T1 and T2 weighted images suggesting subacute haemorrhage.

Her past medical history was remarkable for two incidents of acute

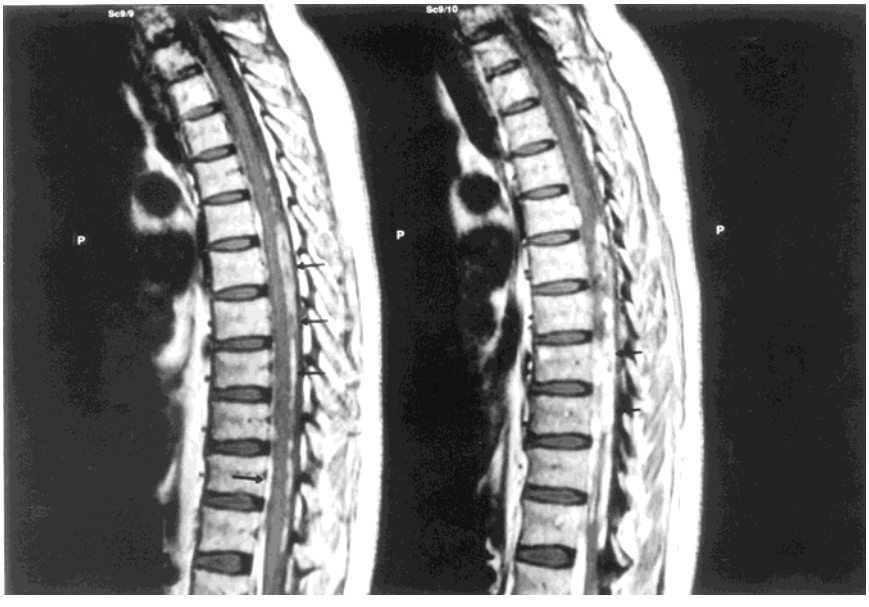
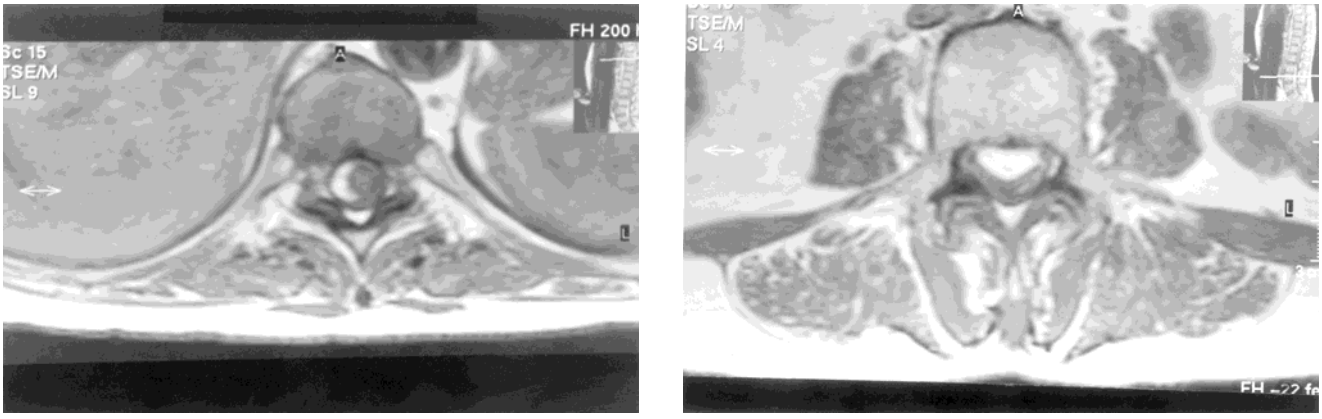


Fig. 1 T1-weighted MRI of the spine eight days after the onset of symptoms, demonstrating a longitudinal subdural mass within the spinal canal (arrows), with an heterogeneous high signal suggesting subacute haemorrhage



(A) (B)

Fig. 2 Transverse T1-weighted MRI scans of the spine, demonstrating a subdural haematoma appearing: (**A**) as a semicircular right ventrolateral collection at the level of T10, (**B**) as a pure ventral collection at the level of L4

pulmonary embolism, the later one four months prior to admission. After the second pulmonary embolism extensive investigations followed that established the diagnosis of ET and the patient had been placed in low-dose aspirin treatment. Two months before the present hospitalization the patient had been admitted at the hospital in a comatose condition. CT of the brain had revealed bilateral frontal acute subdural haematomas. There was no history or physical evidence of trauma. An emergency craniotomy was performed and subdural clots were evacuated. The patient had an uneventful recovery and she was discharged to home without any neurological sequelae. Low-dose aspirin treatment was discontinued.

Spontaneous spinal subdural haematomas are a very uncommon clinical entity. Russell et al. [11] in a review of the literature found 58 cases of spinal subdural haematomas and only five of them were spontaneous. Our case is the first report of spontaneous spinal subdural haematoma in a patient with ET.

More than half of the patients with ET are asymptomatic at the time of diagnosis [4]. Vascular complications like thrombotic and microvascular events are secondary to increased platelet aggregation and may appear in up to 50 % of patients [4]. Transient cerebral ischaemia and stroke are the most frequent CNS manifestations [1, 4]. Histological studies have demonstrated microvascular occlusions with emboli composed of PLT [4]. CNS haemorrhagic complications are extremely rare with only four reported cases of cranial extradural, subdural and intracerebral haematoma. [2, 3, 9, 12]

The pathogenetic mechanism of haemorrhage in ET is not fully understood [1, 4]. However, the major factor in the production of haemorrhage seems to be an intrinsic qualitative abnormality of platelet function, mainly deficient release reaction and deficient platelet factor-3 (PF-3) [5, 13]. This abnormality presumably leads to a haemostatically inadequate platelet aggregation [5]. Indeed aggregation by collagen, epinephrine and ADP that act by inducing the release reaction were deficient in our patient, thus supporting this view.

Spinal subdural haematomas predominate in the thoracolumbar spine, and their extension varies from focal to markedly extensive [11]. In our patient the haematoma was quite extensive spreading from T3 to L5 spinal segments. In spinal extradural haematomas bleeding is believed to be of venous origin [10, 11]. Unlike the spinal epidural space, however, the spinal subdural space does not contain major blood vessels [8].

In those cases the haemorrhage may originate primarily from a vessel within the subarachnoid space when there is a sudden increase of abdominal or thoracic pressure, with a secondary dissection into the subdural space where the major clot develops [10, 11]. The present case supports the theory that spinal subdural haematoma originates in the subarachnoid space, because our patient presented with severe headache and neck stiffness among other symptoms, and the cerebrospinal fluid was haemorrhagic.

MRI is the imaging modality of choice to distinguish a spinal subdural haemorrhagic collection from an epidural haematoma [6]: Epidural haemorrhages are frequently located dorsal to the spinal cord because ventrally the dura is tightly fixed to the posterior longitudinal ligament [15]. In contrast, dorsally the dura is not attached to the vertebral arches so that epidural haematomas are free to expand in this location. The epidural collection appears biconvex, displaces the dura ventrally, and extends over two or three ver-

tebral bodies only [6]. Subdural blood collections are mainly found ventral to the spinal cord, or are semicircular crossing the midline with a major ventral part, and usually extend over a wider range in the rostrocaudal direction [6]. MRI also provides an accurate estimate of the stage of haemorrhage by identifying sequential patterns of transformation of the haemoglobin molecule within the haematoma [7].

In the survey by Russell et al. [11], 92 % of the patients with spinal subdural haematomas presented with paraplegia or major paraparesis. In most of these cases an emergency surgical evacuation was performed resulting in improvement of the neurological deficits. In that respect our case is unusual because motor and bladder functions remained intact, and only pain was the prevailing symptom. Based on the absence of serious neurological signs a conservative treatment was followed in our patient. Moreover, since her platelet count was still high, the risk for further hemorrhagic complications was increased if a surgical evacuation was attempted. The patient received hydroxyurea and started rehabilitation therapy. She made an excellent recovery and after three weeks she was discharged ambulatory at home with no neurological deficits.

In conclusion, patients with ET may be at risk of developing acute spontaneous spinal subdural haematomas. Even in the absence of neurological signs, acute symptoms like severe back or midscapular pain in a patient with ET, should raise the suspicion of spontaneous spinal subdural haematoma and emergency MRI of the spine should be performed.

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