

cord compression except for an MRI with brief case description in the review by Pfister et al.¹ The respiratory arrest that is said to accompany acute tonsillar herniation was obviated in our patient by the preceding mechanical ventilation.

The quadriplegia in our patient was associated with a prolonged flaccid state and areflexia, a feature commented on in other cases. The main differential diagnostic consideration was 'critical illness polyneuropathy.' However, the minor motor nerve conduction abnormalities were not consistent with a neuropathetic or myopathic illness of this degree of severity. Although the process was associated with persistently absent F-waves that are typical of polyradiculopathy, we interpreted this finding as a physiologic reflection of suprasegmental damage to motor tracts that corresponded to spinal shock. Transection of the corticospinal tracts probably occurred in the region of the decussation within the foramen magnum and the sensory level indicated an upper level of damage just below the foramen magnum and sparing the spinal trigeminal tracts.

In some children, herniation has been thought to have a temporal relationship to lumbar puncture,⁵ but others disagree.⁶ Many papers fail to distinguish between transtentorial and transforaminal magnum herniations, making interpretation difficult. Even the term 'coning' is used ambiguously in this literature. We are uncertain of the role played by lumbar puncture in the descent of the tonsils in our case, but ICP was greatly elevated when first measured and a contribution from lumbar puncture was possible, though not necessary. Even the risk of tonsillar herniation certainly does not obviate lumbar puncture in these circumstances, but the appearance of flaccid quadriplegia soon after the procedure might justify decompression at the foramen magnum or aggressive medical treatment of brain swelling.

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Spontaneous cerebral haemorrhage without hypertension in non-mosaic 45X Turner's syndrome

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Summary Cerebral haemorrhage without hypertension, arteriosclerosis or clotting defect has not been reported in patients

with Turner's syndrome before. In a 51 year old female patient with non-mosaic Turner's syndrome, acute aphasia and right-sided hemiplegia occurred, due to left-sided basal ganglia haemorrhage. The history for hypertension was negative, blood pressure was normal throughout hospitalisation as well as during 24 h monitoring, and all tests for secondary hypertension were negative. There was no indication of arteriosclerosis or a clotting defect. Since there were hypermobile joints, hyperextensible skin and ectatic ascending aorta and brachiocephalic trunk on angiography, a general connective tissue defect was assumed, making arteries more vulnerable to physiologically increased blood pressure and rupture of intracerebral arteries with consecutive bleeding. © 2000 Harcourt Publishers Ltd

Keywords: Turner's syndrome, chromosomal aberrations, cerebral bleeding, angiography

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INTRODUCTION

Vascular abnormalities are a common finding in Turner's syndrome and comprise aortic dilatation, aortic dissection, aortic coarctation, aneurysms, haemangiomas, arterio venous malformations and teleangiectasias.^{1,2,3} Connective tissue defects are responsible for these vascular abnormalities.⁴ The presence of a general connective tissue defect in these patients is supported by the following case of spontaneous cerebral haemorrhage without hypertension in a patient with Turner's syndrome.

CASE REPORT

A 51 year old female paediatrician was admitted because of acute onset right-sided hemiplegia and aphasia. On admission, blood pressure was 175/100 and 130/80, respectively. The electrocardiogram demonstrated sinus rhythm, tall S-waves and biphasic T-waves in the right precordial leads. Coagulation function tests were normal. The initial computed tomography (CT) scan of the brain showed a left basal ganglia haemorrhage with compression of the left lateral ventricle and a midline shift of 7 mm to the right (Fig. 1) without indication for neurosurgical intervention.



Fig. 1 CT scan of the brain on admission shows left sided intracerebral haemorrhage with pericerebral oedema.



Fig. 2 Digital subtraction angiography of the aorta shows elongation of the aorta, the common trunk, the vertebral arteries and the space basilar artery, dilatation of the ascending aorta and a slight stenosis between the left common carotid artery and the left subclavian artery just before the isthmus.

Measuring the blood pressure regularly, the patient had no history of hypertension and all other risk factors for cerebral bleeding were negative. The patient was not taking any medication before admission. Past medical history was remarkable for Turner's syndrome diagnosed upon typical morphological features, including short stature, delayed menarche, irregular menses, secondary amenorrhoea since age 20 years and cytogenetic investigations. On clinical neurological investigation, hemianopsia to the right, paresis of vertical gaze and horizontal gaze to the right, central facial palsy, sensory-motor aphasia, right-sided hemiplegia, hemihyphaesthesia, apraxia, exaggerated deep tendon reflexes and positive pyramidal signs were found. Phenotypic features attributable to Turner's syndrome included short stature (146 cm), hypertelorism, reduced mandible, low set ears, low nuchal hair line, broad neck, pterygium colli, shield chest with widely spaced nipples, hyperextensible skin, hypermobile joints, bilateral brachydactyly of the 4th and 5th fingers and toes and pretibial hypertrichosis.

Except for one occasion after emotional stress, blood pressure was within the normal range throughout her hospitalisation. Twenty-four hour blood pressure monitoring revealed no abnormality. Thyroid function tests were normal. Except for slightly elevated vanilylmandelic acid, the other urine catecholamine levels were normal, as well as serum lipids, sexual hormone levels and all clotting tests. Optic fundus investigation was not indicative of hypertensive disease. A cerebral magnetic resonance imaging (MRI), 2 months after the bleeding, showed advanced resorption of the haemorrhage but no visible developmental abnormality of the brain, arteriovenous malformation, telangiectasia or aneurysm.

Transthoracic echocardiography demonstrated severe aortic valve stenosis with a valve area of 0.7 cm², slight aortic insufficiency, moderate hypertrophy of the left ventricle and interventricular septum (15 mm) but normal systolic and diastolic function. Cerebral angiography, 3 weeks after the bleeding, showed elongation of the aorta, the cervicobrachial trunk, the vertebral arteries and the basilar artery and dilatation of the ascending aorta (Fig. 2). Transcranial Doppler sonography was normal. The karyotype on cytogenetic analysis of blood lymphocytes and skin fibroblasts showed a complete monosomy X (45, X).

DISCUSSION

The most common causes for cerebral haemorrhage are hypertension, a clotting defect, atherosclerosis or a general connective tissue defect. Although arterial hypertension was made responsible for cerebral bleeding in the only previously reported patient with Turner's syndrome,⁵ there are several arguments against hypertension as the cause in our patient: 1) the history of hypertension was negative; 2) except for the very first measurement and one situation of emotional stress, blood pressure was normal in the acute phase of the bleeding and during rehabilitation; 3) 24 h blood pressure monitoring was normal; 4) all investigations for secondary hypertension were negative; 5) the patient did not need antihypertensive drugs, neither before nor during hospitalisation 6) funduscopy was normal and 7) the patient had severe aortic valve stenosis, which is associated with hypotension rather than hypertension. Despite these arguments, it cannot be definitively excluded that occasional episodes of high blood pressure occurred which remained undetected to the applied blood pressure monitoring. Arguments for the presence of hypertension in our patient are the location of the bleeding and the moderate left ventricular hypertrophy, which could also be due to the aortic valve stenosis. Concerning other aetiologies, all clotting tests were normal and there was no evidence of atherosclerosis. No signs of atheromatosis could be found on cerebral angiography and the patient did not complain of angina pectoris, claudicatio spinalis or intermittens. Risk factors for atheroma like diabetes, hypertension and hyperlipidaemia were all negative.

The assumption of a general connective tissue defect in patients with Turner's syndrome is based on the following findings: 1) the incidence of vascular abnormalities, attributable to connective tissue defects, like aortic dilatation, aortic dissection, aneurysms, arteriovenous malformation and telangiectasias, is higher in patients with Turner's syndrome than normal; 2) there are Turner's phenotypes that resemble Ehlers-Danlos and Marfan syndrome^{3,6} and 3) cystic media necrosis may be occasionally found in some patients with Turner's syndrome.⁴ The basic abnormality, made responsible for the assumed connective tissue defect, is a general elastic fibre disruption.⁴ Such a defect may favour rupture of intracerebral arteries already at physiologically increased blood pressure. Arguments against a general connective tissue defect in our patient are that Ehlers-Danlos syndrome is more frequently associated with carotid artery dissection and subarachnoidal haemorrhage than with intracerebral haemorrhage, and that no aneurysms, arteriovenous malformations or telangiectasias were found on cerebral MRI and angiography. However, patients with Turner's syndrome should carefully monitor their blood pressure and should avoid emotional stress, competitive sports and sports associated with the risk of trauma.^{3,7}

It is concluded that spontaneous intracerebral haemorrhage may occur in patients with non-mosaic Turner's syndrome without hypertension. In such patients a general connective tissue defect, also affecting the cerebral arteries, could be responsible for arterial rupture already at physiologically increased blood pressure.

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INTRODUCTION

Acute intracranial haemorrhage (ICH) accounts for 15–45% of cerebrovascular accidents worldwide.¹ This often presents catastrophically with major symptoms at onset, including severe headache, loss of consciousness or functional limb deficit. The onset is usually not instantaneous but rather a smooth crescendo of symptoms and rapid plateau.² Pathologically and often clinically, intracranial haemorrhage can be further subdivided into intraparenchymal and subarachnoid haemorrhage. Intraparenchymal haemorrhage accounts for approximately 10% of first ever strokes in the West, and subarachnoid haemorrhage provides for the remaining 5% of such strokes. Intraparenchymal haemorrhage can be classified as primary (de novo) or secondary (occurring in an area of previous infarction).

The most common site of occurrence of intraparenchymal haemorrhage is in the basal ganglia, with the next most frequent site being intralobar followed by brainstem bleeding. Once bleeding has occurred, extension through the cerebrum into the subarachnoid space or ventricles is possible.¹ Pathologically, lipohyalinosis of the smaller blood vessels or amyloid change may be noted; often, however, the bleed results in obliteration of the underlying vessel abnormality and it is difficult to ascribe the exact cause of the haemorrhage.¹

Clinically, smaller, less acute ICH may present with milder symptoms, such as altered level of consciousness, confusion or decline in rehabilitation capability during recovery from ischaemic stroke whilst the patient is on anticoagulation.³

Facial pain is not described as a common presentation of ICH. Review of the literature via Medline back to 1966 revealed no previous reported cases of such a presentation. We report a case of acute ICH with the first symptom being facial pain. Speculation as to its pathophysiology will be outlined.

CASE PRESENTATION

A 53 year old Caucasian female who was previously well, developed acute onset facial pain extending from the right orbit to the maxilla. The pain started suddenly and rapidly plateaued over a few seconds. The pain was then at a peak and remained stable. She described the pain as a toothache involving the right side of the face. Within 10 minutes, she started to vomit and on alighting from the car she was noted to be ataxic. Her level of consciousness declined prior to attending the Emergency Department.

On presentation to hospital, she was noted to be in atrial fibrillation, had mild pyrexia and difficulty communicating. Though able to obey commands, she had difficulty with fine finger movements on the left. She had normal sensation and was normoreflexic with an extensor left plantar response.

Facial sensation could not be adequately assessed and it was impossible to define an anatomical localisation to her pain with no demonstrable sensory loss.

Computed tomography (CT) scan imaging of the brain revealed a 4 × 3 cm right temporal lobe haemorrhage with some mass effect and oedema. There was also some blood noted in the subarachnoid space in the middle cranial fossa (Figs 1,2). Symptomatically, she continued to complain of right facial pain for 3 days. The pain was boring in nature and not limited to any subdivision of the trigem-

Facial pain as a presenting feature of intracerebral haemorrhage

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Summary A literature review from 1966 using Medline with keywords 'cerebral haemorrhage' and 'facial pain' failed to reveal any cases in which facial pain was the initial feature of intracranial haemorrhage. The following case describes ipsilateral facial pain which is previously undescribed as a presentation of intracranial bleeding.

A 53 year old female who was previously well, with no significant history of headache, developed right facial pain from the orbit to the maxilla. Ten to 15 min later she developed nausea and vomiting with unsteadiness and confusion. She had difficulty with left-hand fine finger movements, with normal sensation and reflexes but an extensor plantar response on the left. Facial pain persisted for 3 days. Initial imaging revealed a 4 × 3 cm right temporal lobe haemorrhage with mass effect and oedema extending into the subarachnoid space. Angiogram revealed a right temporal lobe arteriovenous malformation.

The basis of the pain remains speculative but includes sensation from the torn vessel wall being referred to the face and subarachnoid blood irritation of the meninges in the middle cranial fossa. Another possibility is irritation of somatosensory cortex II, but why this should result in only ipsilateral pain is unclear. Facial pain should be an alerting symptom to the neurologist when it appears with no apparent cause. © 2000 Harcourt Publishers Ltd

Keywords: facial pain, stroke, presentation, intracerebral haemorrhage

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