Intramedullary migration of spinal cord lipoma

Spinal cord lipomas may grow with changes in body fat, and can produce neurological manifestations due to nervous tissue compression or cord tethering. It is very unusual for the tumour to migrate from one part of the cord to another, thus to cause neurological symptoms at the migrated level. This, however, is a report on such a case. To the best of our knowledge, this has not previously been described in the literature.

Case illustration

A 45 year old man presented to a different neuroscience centre with a history of deformity of the right foot since childhood. In his 20s, he developed intermittent intercostal pain and was found to have reduced sensation in the left T4 to T12 region. In October 1998, an MRI scan of the spine showed a low abnormality was found; the cord appeared normal. A needle aspiration was performed but no abnormal tissue was identified. Following the surgery, the intercostal pain resolved.

Nine months before referral to our unit, the patient developed numbness in the C8 distribution of the left hand. Examination revealed normal strength in the upper limbs with diminished reflexes in the left arm. There was reduced pinprick sensation from C8 to L4 on the left and from T4 to S4 on the right. Proprioception was affected in the right foot. Clonus was present in the right ankle. The right lower leg was wasted and there was pes cavus deformity. There was grade 3 weakness of right ankle eversion. Spinal examination was normal except for the surgical scar.

Review of the original MRI scan (fig 1) suggested the presence of a dermoid cyst at L2/3 intimately related to the lipomatous tissue. A further MRI scan in 2001 (fig 2) showed that the lipomatous tissue lying within the cord had extended to the C6 level, but that the amount of lipomatous tissue in the lower cord had reduced as compared with the first scan. The nature of this tissue was confirmed as fat by a fat suppression MR sequence. In the lower region of the spinal cord it was now possible to see a syrinx cavity, the presence of which had been obscured by fat on the initial MRI scan.

As the patient’s clinical deterioration had stabilised at this stage, we decided to adopt a conservative policy.

Discussion

Tethered cord is known to be associated with spinal cord lipomas. Terminal lipomas are spinal cord lipomas that are inserted in the end of the conus and incorporated in the filum. They may contain excessive amounts of non-adipose mesenchymal derivatives such as cartilage, bone, and fibrous septums. Tethering of the spinal cord can be associated with syringomyelia, probably due to pathological intramedullary pressure changes during movements of the spine. In this case, we suggest that there was a spontaneous rupture of the possible dermoid cyst into the intramedullary syrinx, allowing migration of fat into the syrinx. This fat was then transported cranially with the CSF flow, remaining there by virtue of its viscosity. It further enlarged and extended the syrinx cranially, and accumulated at the cranial end of the syrinx.

After a careful literature search, we could find no previous description of this. However, Klekamp and Samii illustrated a very similar case in their textbook. Intramedullary lipomas do have a potential to change size with the body fat and hence cause neurological symptoms. This is the first case report of possible migration of lipomatous tissue within the cord.

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Figure 1  (A) MRI of the lumbar spine in 1998 showing tethered cord with dermoid opposite the L2 and L3 bodies, with intramedullary lipoma extending to the T10 region. (B) MRI of the cervical spine in 1998 showing no evidence of involvement of the cervical and upper thoracic spinal cord.
Economy class stroke syndromes: vertebral artery dissection revisited

Economy class syndrome was first reported in 1988 and refers to an association between air travel and venous thromboembolism. Recently, three cases of stroke in young adults without risk factors other than a patent foramen ovale have been reported, presumably the result of paradoxical embolism. We now present an unusual case of medial medullary infarction caused by vertebral artery dissection associated with abnormal neck posture during a long haul flight—another health related reason to travel first class.

A 56 year old right handed insurance broker on a 7 hours economy class long haul flight returning to the United Kingdom fell asleep with his head uncomfortably twisted to the right. On waking he noticed pain in the left side of his neck and paraesthesia in the right arm. The episode resolved after 15 minutes, but recurred with a right hemiparesis affecting the arm and leg. He smoked 40 cigarettes a day and drank 24 units of alcohol a week. There was no significant past medical history and no family history of vascular disease.

On examination, conscious level, cognition, speech, and the cranial nerves were entirely normal, there was no facial weakness, and the tongue was normal. There was pyramidal weakness MRC grade 3 affecting the right arm and leg with right sided hypereflexia and a right extensor plantar response. Joint position, vibration, and light touch sensations were normal, but pain and temperature sensation were reduced over the right side. Blood pressure was 150/90 and cardiopulmonary examination was otherwise normal. Fasting cholesterol was 3.5 mmol/l, but other routine blood tests and coagulation studies, autoantibody screen, treponemal serology, urinalysis, ECG, chest x ray, and transthoracic echocardiography were all normal.

Magnetic resonance imaging (MRI) of the brain (fig 1, panels A and B) showed signal abnormality of the left pyramid, part of the olive, and the lemniscal region of the medulla, confirmed on diffusion weighted imaging to represent an acute infarct. Doppler ultrasoundography of the neck vessels was normal. Initial time-of-flight magnetic resonance angiography (MRA) showed a probable minor irregularity of the left vertebral artery in its proximal portion distal to the C2 vertebral body. Subsequent contrast enhanced magnetic resonance angiography (fig 1C) showed definite irregularity of caliber of the left vertebral artery from C2 to its junction with the basilar artery. Fat suppression axial MR of the upper cervical spine and lower posterior fossa confirmed left vertebral artery dissection at the level of C2 and along its length to its junction with the basilar artery (ordinary T2 weighted image shown in fig 1, panel D). The patient was treated with aspirin, clopidogrel, and sinvastatin. With rehabilitation he made a full recovery and was discharged home.

Comment

Medial medullary infarction accounts for less than 1% of cases of symptomatic vertebral stroke and is usually thought to result from atherosclerosis of the vertebral artery, anterior spinal artery, or medullary perforating arteries. In the upper medulla the anteromedial medullary perforating arteries arise from the vertebral artery in the cephalad limits of the anterior spinal artery above their point of fusion anterior to the lower medulla. They supply the ipsilateral medial portion of the pyramidal tract, the medial lemniscus, and the medial longitudinal fasciculus and extend dorsomedially to reach the hypoglossal nerve and central reticulotegmental formation. The distribution of the signal change in the axial T2 weighted sequence (fig 1A) corresponds exactly to the vascular territory of the anteromedial perforating branches of the left limb of the anterior spinal artery.

Classically, medial medullary infarction, or Dejerine’s syndrome, results in contralateral hemiparesis sparing the face, with lemniscal sensory loss and ipsilesional lingual palsy. Several series have reported a greater heterogeneity of clinical features than previously documented, including nasea, vertigo, headache, somnolence, mild ipsilateral pupil and rotational nystagmus, dysarthria, dysphagia, mild contralateral facial palsy, ipsilateral or contralateral decrease in sensation (most commonly pain and vibration), ipsilateral limb ataxia, and contralateral truncal lateropulsion. Dissociation of medial lemniscal sensory modes such as vibration and proprioception has also been described. Notably, although pain sensation is carried in the lateral lemniscus, contralateral reduction in pain sensation in the arm and leg has been well documented after medial medullary infarction.

Three main clinical pictures have emerged: sensorimotor stroke with ipsilesional lingual palsy, sensorimotor stroke without lingual palsy, and a stroke syndrome. Two retrospective reviews of MRI proven medial medullary infarction reported 11 cases among 4200 (0.26%) and 2310 (0.52%) consecutive patients with ischaemic stroke. The proportion of cases of medial medullary infarction with ipsilateral lingual palsy has ranged from 11% to 64%.

In most series, medial medullary infarction is attributed to atherothrombosis of vertebral artery, anterior spinal artery, or their medullary penetrating branches. Cardiovascular risk factors are usually present. Dissection of the vertebral artery leading to medial medullary infarction has only been postulated in four of over 80 cases of this condition reported previously, but included three of seven cases reported by Bassetti et al. Whatever the mechanism, dissection of the anteromedial perforating branches of the medulla causing the medial medullary syndrome, it seems unlikely that it would often be the result of embolism from a distal site, as the branches usually arise from small vessels off either the anterior spinal artery or the vertebral artery. Local occlusion of the anterior spinal artery or the vertebral artery by either dissection or atherothrombosis is a more appealing explanation. In the present case, the dissection is likely to have affected the vertebral artery along its entire distal length and to have included the origin of the anterior spinal artery. It seems probable—particularly in young adults in the absence of cardiac embolism or widespread atheroma—that dissection is a more common cause of medial medullary infarction than previously acknowledged. We should therefore suggest that all MRI sequences including axial images and contrast enhanced MRA of the vertebral

Figure 2. (A) MRI of the lumbar spine in 2001 showing lipomatous dilatations in the low lying cord. However, there is less lipoid tissue in this region than in the 1998 scan. (B) MRI of the cervical spine in 2001 showing extension of the lipoma up to the C6 level and involvement of the lower cervical and upper thoracic region.

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arteries should be included in the investigation of the aetiology of medial medullary infarction. However, it may prove difficult to distinguish between dissection and atherothrombosis, especially once occlusion of the vessel has occurred.

Anticoagulation has been advocated to prevent thromboembolic complications of acute carotid or vertebral artery dissection; however, there are no reported controlled trials proving its efficacy. In the context of the rapid neurological recovery after initial treatment with antiplatelet agents, and in the absence of good evidence that anticoagulation started postacutely is effective, antiplatelet treatment alone was continued in the present case.

Our case adds to the literature of stroke precipitated by minor trauma, including “beauty parlour stroke”, “bottoms up dissection”, and hyperextension or rotation of the neck associated with chiropractic manipulations, practising yoga, painting a ceiling, prolonged telephone calls, roller coaster rides, coughing, vomiting, sneezing, routine anaesthesia, and the act of resuscitation. Apart from three cases recently reported of arterial stroke after air travel in association with a patent foramen ovale, presumably caused by paradoxical embolism, a Medline search over the last 35 years revealed only two cases of thromboembolic stroke, and no cases of vertebral artery or carotid dissection associated with air travel. Nevertheless, exhausted long haul passengers with cardiovascular risk factors should obviously request first class accommodation in which to fall asleep!

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Adult paraneoplastic opsonolus-myoclonus syndrome associated with antimitochondrial autoantibodies

Paraneoplastic opsonolus-myoclonus syndrome (OMS) is a rare complication of cancer characterised by chaotic, synchronous eye movements (opsoclonus), spontaneous muscle jerks (myoclonus), and ataxia. In children OMS is almost exclusively associated with neuroblastoma, whereas in adults small cell lung cancer (SCLC) and breast cancer are the most frequent tumours associated with OMS. Some breast cancer patients with OMS have an antineuronal antibody called anti-Ri in the serum and CSF. In most patients with SCLC-associated OMS, antineuronal antibodies are not detected. Here we report the first case of SCLC-associated OMS with high titre antimitochondrial antibodies (AMA) in serum and CSF.

A 58 year old Caucasian woman developed a flu like syndrome, followed by eye movement disturbances three days later. On admission to our hospital, physical examination revealed disorientation, opsoclonus, myoclonus predominantly of the upper limbs, and severe limb and trunk ataxia. The deep tendon reflexes were normal and no paresis or sensory deficit could be found. Cranial CT and MRI scans and routine laboratory tests

Figure 1
Axial T2 weighted (A) and coronal FLAIR (B) sequences showing signal abnormality involving the left pyramid, anteromedial olive, and lemniscal region of the medulla. There was corresponding signal change suggesting restricted diffusion, consistent with acute infarction, on diffusion weighted imaging (not shown). Contrast enhanced magnetic resonance angiography (C) showed irregularity of calibre of the left vertebral artery. It was confirmed that this represented dissection on axial images of all pulse sequences, including fat suppression images (not shown), and T2 weighted image (D), which in addition shows the crescent sign of dissection.
were normal. CSF examination revealed lymphocytic pleocytosis, normal total protein, and positive oligoclonal bands. In serum and CSF, antineuronal antibodies by high titre AMA could be detected. High dose steroid therapy was started one week after onset of symptoms, and the patient experienced significant improvement of the OMS within three days of commencing steroid treatment. By four weeks the opsonolus and myoclonus had disappeared, and there was only a mild trunc axatia: the patient could walk unassisted.

CT scan of the thorax showed a mediastinal mass. Bronchoscopy was normal, but a mediastinoscopic biopsy was positive for SCLC cells. Starting at the fourth week after admission, the patient received chemotherapy followed by a complete remission of the SCLC. At present, after a 2 year follow up, the patient is in good health (under intermittent chemotherapy without signs of tumour relapse) and, with exception of a mild ataxia, the neurological status is normal. The patient did not have clinical or laboratory signs of primary biliary cirrhosis (PBC) or any other liver disease at any time.

Using frozen sections of primate cerebellum, neuroblastoma (SK-N-SH) and SCLC cell lines, cells and kidney (Euroimmun, Lübeck) for routine autoantibody tests, no antineuronal antibody was found, but strong staining of the mitochondria could be detected in all tissues by indirect immunofluorescence. The initial CSF titre of the AMA was 1/1000 and CSF ASI was 18.9 (normal value, [AMA(serum)/IgG(serum)]) was calculated. ASI (ASI = [AMA(CSF)/IgG(CSF)]/[AMA(serum)/IgG(serum)]) was calculated. ASI was 18.9 (normal value 3), indicating intrathecal synthesis of the AMA.

Western blot analysis, using pyruvate dehydrogenase (PDH), cerebellum, liver, and kidney as well as protein homogenates of neuroblastoma (SK-N-SH) and SCLC cell lines (N417), was performed as previously described. No reactivity against PDH or any neuronal specific reactivity was found. However, the patient’s serum showed a reactivity to 18 and 21 kD protein exposed in all the above tissues and cell lines. Using the EUROASSAY® technique (Euroimmun, Lübeck), the serum showed no reactivity to the mitochondrial M2, M4 or M9 antigens. Western blot of recombinant NOVA-1/2 proteins showed no reactivity for the mitochondrial M2, M4 or M9 antigens. However, the patient’s serum showed a neuronal specific reactivity was found. In adult onset idiopathic or paraneoplastic sensory, and autonomic nervous function is usually presents with deterioration of motor, sensory, and autonomic nervous function caused by rostrocaudal traction on the spinal cord. The onset of symptoms commonly occurs in childhood,12-14 There have been a few reported cases of adult onset tethered cord syndrome, and the mechanisms of late onset are generally attributed to the degree of

Comment

Tethered cord syndrome with spinal lipoma usually presents with deterioration of motor, sensory, and autonomic nervous function caused by rostrocaudal traction on the spinal cord. The onset of symptoms commonly occurs in childhood.12-14 There have been a few reported cases of adult onset tethered cord syndrome, and the mechanisms of late onset are generally attributed to the degree of disturbance is generally only seen in patients with a relatively short clinical history; therefore early diagnosis and treatment are essential to achieve a good functional recovery.

We present a rare case of a patient who had suffered from refractory diarrhea for years without a definite diagnosis and who was finally found to have tethered cord syndrome after bladder symptoms became apparent.

Case report

A 22 year old woman was admitted to our department of gastroenterology with a complaint of severe diarrhea which had been occurring five to 10 times a day since she was 19. The cause of the diarrhea could not be determined, even after a barium enema and endoscopy. The diarrhea did not improve with drug treatment and diet modification. The patient also began to suffer from a urinary disturbance. Incomplete emptying of her urine had been verified when she was 21 years old. After this history, the patient was admitted to the department of urology, where a urodynamic study showed a neurogenic bladder. There was sphincter dysfunction and hyper trophy of the external sphincter. Lumbar magnetic resonance imaging (MRI) was done and showed a thick tethering film with intradural spinal lipoma in the sacral region (fig 1). There were no abnormal signs over the skin at the lumbosacral region. The spinal cord was untethered (fig 2), after which the refractory diarrhea improved promptly and had almost completely resolved after six months. However the urinary disturbance remained and the patient had to perform self catheterisation.

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Figure 1 T1 weighted magnetic resonance image showing the intradural spinal lipoma (arrow) tethering the spinal cord at the level of the fifth lumbar vertebra.

Adult tethered cord syndrome presenting with refractory diarrhea

Adult tethered cord syndrome presents with various neurological symptoms such as bladder/bowel disturbance, motor and sensory disturbance, and pain.12-14 A common bowel disturbance is constipation and incontinence, which may recover depending on the severity or duration of the symptoms.12-14 In adult tethered cord syndrome, recovery of bladder
tethering and the cumulative stress of repeated microtrauma from exercise, especially any requiring a flexion position. The mechanical stress resulting from a tethered cord is usually localised in the lumbosacral region; thus most sensorimotor deficits appear in the lower limbs or in relation to bladder and bowel function. In urinary disturbances, some cases show a spastic small capacity bladder caused by supranuclear interruption, while others present with a hypotonic, large capacity bladder caused by dysfunction of the sacral autonomic nuclei. This suggests that longitudinal tensile stress within the cord may be transmitted even to remote regions of the cord, and various symptoms may appear that reflect the damaged part of the cord. Bowel dysfunction can also be attributed to this mechanism. Although most previous reports have mentioned either constipation or incontinence, in this case the diarrhoea was possibly caused by irritation of the digestive system related to parasympathetic upregulation or sympathetic inhibition. The surgical procedure of untethering the spinal cord and loosening the longitudinal tensile stress may alter the balance of sympathetic and parasympathetic bowel supply and affect the symptoms of bowel hypermotility. Because it is more difficult to evaluate bowel function objectively than to evaluate urinary dysfunction, bowel disorders—especially diarrhoea— are often regarded as functional disorders (irritable colon) if there is no obvious underlying disease. This is the probable reason why we could find so few reports on bowel dysfunction in tethered cord syndrome.

The improvement in our patient’s diarrhoea after untethering the conus medullaris strongly suggests that the tethered cord was the primary cause of the symptom.

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