24.9 Brainstem syndromes 6006 David Bates

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ESSENTIALS Brainstem syndromes typically cause ipsilateral cranial nerve lesions and contralateral long tract signs. They are commonly due to brain- stem ischaemia, but can also be caused by neoplasia, demyelination, infective, and hamartomatous lesions. Imaging, ideally with mag- netic resonance imaging rather than computed tomography, is ob- ligatory and only then—and possibly following other investigations to identify systemic abnormality or cerebrospinal fluid changes—can appropriate therapy be introduced. Particular brainstem syndromes These include: (1) Thalamic syndrome—typically presents with unpleasant dysaesthetic burning pain, often following a hemi- plegic and hemianaesthetic stroke. First-line treatment is with centrally acting analgesic agents (e.g. amitriptyline). (2) Midbrain syndromes—characterized by contralateral long tract signs and ip- silateral defects of the third and fourth cranial nerves. (3) Pontine syndromes—present with ipsilateral cerebellar signs, abnormal- ities of ocular movements, and contralateral paralysis and impair- ment of sensation, with details of the deficits depending on the location of the lesion within the pons. (4) Medullary syndromes—
(a) medial—causes paralysis of the tongue on the side of the lesion, with contralateral paralysis of the arm and leg, and impaired pro- prioception over the contralateral half of the body; (b) lateral—causes ipsilateral Horner’s syndrome, vocal cord paralysis, limb ataxia and sensory loss, oscillopsia and nystagmus, and contralat- eral impairment of pain and thermal sensation. Introduction The classic presentation of brainstem syndromes, including the long tracts and deficits of cranial nerve nuclei, commonly causes crossed cranial nerve and motor or sensory long tract deficits; the cranial nerve lesions are ipsilateral to the lesion and the long tract signs are contralateral. It is important to assess the extracra- nial vascular supply to the posterior circulation, especially to listen for bruits over the subclavian vessels and to record the pulse and blood pressure in both upper limbs, remembering that the vertebral arteries arise from the subclavian vessels. Apart from the cranial nerve and long tract deficits, there may be ataxia, vertigo, the pres- ence of an internuclear ophthalmoplegia and unreactive pupils, the symptoms of diplopia and oscillopsia, and the finding of nystagmus or ocular paresis. The circulation to the brainstem is supplied by the vertebral ar- teries, which are the main arteries to the medulla, and then the basilar artery, which supplies the pons and midbrain. The vertebral arteries are frequently asymmetrical and commonly give rise to the

large posteroinferior cerebellar arteries shortly before they join to form the basilar artery. The vertebral arteries are susceptible to trauma within the cervical spine, but the most common lesion affecting the vertebral arteries is dissection, which is probably underrecognized, and thrombosis. The basilar artery branches are paramedian, supplying the area of the pons close to the midline, the short circumferential branches that supply the lateral two-thirds of the pons, the long circumfer- ential branches that are the supero and anteroinferior cerebellar arteries, and several interpeduncular branches that arise at the bi- furcation of the basilar artery and supply the subthalamic and high midbrain regions. Strokes in the posterior circulation account for 20% of all is- chaemic cerebrovascular disease. Although most brainstem syn- dromes are due to vascular or inflammatory demyelinating lesions, an increasing proportion are shown on magnetic resonance imaging (MRI) to be due to cavernomas, which may or may not calcify and which were impossible to diagnose before modern imaging. The management of such cavernomas is difficult and may involve sur- gery or stereotactic radiotherapy. A small proportion of lesions in the brainstem are infective and inflammatory; occasionally a causa- tive organism, commonly an adenovirus, can be identified, and may be seen as a complication of AIDS; when no identifiable infective agent is identified, the condition is termed ‘Bickerstaff’s encephalitis’. The brainstem syndromes Diencephalic syndrome Commonly seen in children under the age of 3 years, Russell’s syn- drome consists of emaciation with increased appetite, euphoria, 24.9 Brainstem syndromes David Bates

 vomiting, and excessive sweating. There may also be motor hyper- activity. The differential diagnosis includes hyperthyroidism, dia- betes mellitus, a tumour in the region of the fourth ventricle, such as a malformation of the great vein of Galen, and a hypothalamic tumour. There can often be optic atrophy and rarely nystagmus. Investi gation may show an elevated serum growth hormone and an MRI will show a hypothalamic mass lesion. Although contraindicated if there is a significant structural pathology, the cerebrospinal fluid may contain malignant cells or an excess of human chorionic gonadotropin in the case of germinoma. Thalamic syndrome Originally described by Dejerine and Roussy in 1906, thalamic pain has a particularly distressing quality. Sometimes spontaneously, but commonly after a recognized hemiplegic and hemianaesthetic stroke, the patient develops altered sensation in a hemisensory distribution together with unpleasant dysaesthetic burning pain (thalamic pain). The pain may be worsened by stimulation and is as- sociated with hemianaesthesia, sometimes proprioceptive loss, and some evidence of hemiparesis. Anatomically the lesion is usually in the ventroposterolateral nucleus of the thalamus and is commonly caused by either a vascular event or a tumour. The investigation required is imaging and therapy is with centrally acting analgesic agents, such as amitriptyline, carbamazepine, gabapentin, and pregabalin. When pain is intractable, deep brain stimulation to the ventroposteromedial nuclei may be considered. Tectal deafness There is a rare syndrome associated with damage at the level of the inferior colliculi, due to either neoplasia or vascular lesions, which results in bilateral deafness with associated difficulty in coordin- ation, weakness, and vertigo. The condition must be differentiated from bilateral conductive hearing loss, cochlear disorders, bilateral cranial nerve VIII lesions, and pure word deafness. Brain imaging will identify the lesion. Thalamic stroke syndrome Lesions affecting the thalamus are commonly vascular and arise from infarction within the distribution of the posterior commu- nicating artery, the basilar, and the anterior and posterior chor- oidal arteries. There is usually hemiparesis with hemianopia, hemianaesthesia, and sometimes hemiataxia. There is often con- fusion and disorientation, and there may be language disturbance. On occasion there may be vertical gaze ophthalmoplegia, loss of pupillary reflexes, and an inability to converge the eyes. There may also

be memory impairment and, on occasions, visual perceptual disturbances are recorded. Midbrain syndromes Damage to areas of the midbrain is characterized by long tract signs contralateral to the lesion with defects of cranial nerves III and IV ipsilaterally. They may occur with lesions in the brainstem or as the evolution of symptoms of rostrocaudal deterioration associated with supratentorial brain swelling (Figs. 24.9.1 and 24.9.2). They are characterized by ipsilateral cranial nerve III and IV palsies together with contralateral hemiparesis, loss of vibration, proprioception, and stereognosis, contralateral loss of pain and temperature, and an ipsilateral Horner's syndrome. Ataxia may occur and there can be eyelid ptosis, diplopia, supranuclear horizontal-gaze paresis, and an internuclear ophthalmoplegia. The association of an ipsilateral oculomotor palsy with a crossed hemiplegia due to a lesion at the base of the midbrain is called Weber's syndrome. Claude's syndrome causes an ipsilateral oculomotor palsy with contralateral cerebellar ataxia and tremor, and is due to a lesion in the tegmentum of the midbrain involving the red nucleus and nerve III nucleus. Benedikt's syndrome also involves the tegmentum of the midbrain, resulting in an oculomotor palsy with contralateral cerebellar ataxia, tremor, and corticospinal signs; it can be regarded as a combination of Claude's and Weber's syndromes. Nothnagel's syndrome occurs with unilat- eral or bilateral involvement of nerve III nucleus together with the superior cerebellar peduncle and causes bilateral ptosis, paralysis of gaze, and cerebellar ataxia. Damage in the region of the dorsal midbrain results in Parinaud's syndrome in which there is paralysis of upward gaze due to damage to the supranuclear mechanisms for upward gaze, loss of accommo- dation, and fixed pupils. Although this may be seen with ischaemic lesions, it is more common with pineal tumours. Third nerve Cerebral aqueduct Middle lateral midbrain syndrome Substantia nigra Spinothalamic tract Medial lemniscus Periaqueductal gray Middle medial midbrain syndrome Medial longitudinal fasciculus Brachium of superior colliculus Midbrain reticular formation Third nucleus Edinger-Westphal nucleus Superior colliculus Pineal Medial geniculate Red nucleus Cerebral peduncle Fig. 24.9.1 Midbrain at the superior colliculus level, showing the medial and lateral territories involved with occlusive stroke in this region. Corticospinal tract Fourth nucleus Inferior lateral midbrain syndrome Medial lemniscus Spinothalamic tract Midbrain reticular formation Medial longitudinal fasciculus Inferior medial midbrain syndrome Substantia nigra Lateral lemniscus Nucleus of inferior colliculus Middle cerebellar peduncle Fig. 24.9.2 Midbrain at the inferior colliculus level showing the medial and lateral territories involved with ischaemic stroke syndromes in this area. Reprinted with permission from DeArmond SI, Fusco MM, Dewey MM, 1976, Structure of the human brain, 2nd edn. Oxford University Press, New York.

section 24 Neurological disorders 6008 Pontine syndromes Lesions in the pons and medulla are commonly identified as involving either the medial or the lateral aspect of the brainstem, depending upon whether the paramedian or short circumferential vessels from the basilar have been involved. In the pons the fol- lowing three levels of damage can be identified and the basal syn- drome can occur at any level. Superior pontine syndrome The medial superior pontine syndrome results in ipsilateral cere- bellar ataxia, internuclear ophthalmoplegia, and palatal and pha- ryngeal myoclonus with contralateral paralysis of face, arm, and leg, and sometimes loss of sensation contralaterally. The lateral superior syndrome causes ataxia of the limbs and gait with dizziness, nausea, and vomiting; there is horizontal nystagmus, paresis of conjugate gaze towards the side of the lesion, loss of optokinetic nystagmus, and sometimes skew deviation of the eyes. There may also be an ip- silateral Horner's syndrome, and there is contralateral loss of pain and thermal sensation on the face and limbs with impaired touch, vibration, and position sense (Fig. 24.9.3). Midpontine syndrome The medial, midpontine syndrome causes ipsilateral ataxia of the

limbs and gait with contralateral paralysis of the face, arm, and leg, deviation of the eyes away from the lesion, and variably impaired sen- sation contralaterally. The lateral syndrome at this level causes ataxia of the limbs on the side of the lesion together with paralysis of the muscles of mastication and impaired sensation over the face on the same side due to damage to cranial nerve V (Figs. 24.9.4 and 24.9.5). Inferior pontine syndrome The medial syndrome causes paralysis of conjugate gaze to the side of the lesion, nystagmus, ataxia of limbs on the same side, and double vision on gaze to that side. Contralaterally there is paralysis of the face, arm, and leg, with impaired touch and proprioception over the opposite side of the body. The lateral syndrome involves ip- silateral, horizontal, and vertical nystagmus with vertigo and nausea, ipsilateral facial paralysis, paralysis of conjugate gaze to the side of the lesion, deafness, tinnitus, and ataxia on the side of the lesion, with impaired sensation of the face on that side. On the opposite side there is impaired sensation over half the body (Fig. 24.9.6). Basal pontine syndrome (locked-in syndrome) Bilateral lesions of the paramedian vessels from the basilar, com- monly seen in patients with hypertension, result in infarction of the basal pontine and cause quadriplegia with loss of the ability to speak. The ascending reticular activating system is intact and Lateral superior pontine syndrome Medial lemniscus Spinothalamic tract Medial longitudinal fasciculus Corticospinal tract Medial superior pontine syndrome Lateral lemniscus Superior cerebellar peduncle Fig. 24.9.3 Superior pontine level, showing the medial and lateral territories involved with occlusive stroke in this region. Reprinted with permission from Adams RD, Victor M, 1993, Principles of neurology, 5th edn. McGraw-Hill, New York. Fifth motor nucleus Medial lemniscus Spinothalamic tract Medial longitudinal fasciculus Corticospinal tract Middle lateral pontine syndrome Lateral lemniscus Fifth nerve Middle medial pontine syndrome Middle cerebellar peduncle Fifth sensory nucleus Superior cerebellar peduncle Fig. 24.9.4 Midpontine level, showing the medial and lateral territories involved with ischaemic stroke syndromes in this locality. Reprinted with permission from Adams RD, Victor M, 1993, Principles of neurology, 5th edn. McGraw-Hill, New York. (b) (a) Fig. 24.9.5 MRI of a midpontine infarction.

 consciousness is therefore preserved. Vertical eye movements and eye closure are all that are possible and under voluntary control in the ‘locked-in syndrome’. Pseudobulbar palsy Bilateral lesions of the long descending tracts in the brainstem can result in pseudobulbar palsy, although this condition is more com- monly seen with lesions higher in the cerebrum. The symptoms are those of spastic dysarthria, dysphagia, bilateral facial weakness with quadriparesis, and emotional lability. Medullary syndromes The medial medullary syndrome may occur with occlusion of the ver- tebral artery or a branch of the lower basilar artery; it causes paralysis and atrophy of the tongue on the side of the lesion with contralateral paralysis of the arm and leg but sparing the face, and impaired tactile proprioceptive sensation over the contralateral half of the body. The lateral medullary syndrome, eponymously called Wallenberg’s syndrome, occurs most commonly with dissection or occlusion of the vertebral artery, resulting in ischaemia into the posteroinferior cerebellar artery; this causes pain, numbness, and impaired sensa- tion of the ipsilateral half of the face with ataxia of limbs on that side, the symptoms of vertigo and nausea, double vision, and oscillopsia, and the signs of nystagmus. There is an ipsilateral Horner’s syn- drome, often dysphagia with paralysis of the vocal fold ipsilaterally, and loss of sensation on the arm, trunk, and leg. There is contralat- eral impaired pain and thermal sensation over half the body and possibly the face (Fig. 24.9.7). A syndrome involving ipsilateral cranial nerve VII and VI pal- sies with a contralateral hemiplegia is called the Millard–Gubler syndrome; the involvement of cranial nerve X, causing paralysis of the soft palate and vocal fold with contralateral hemianaesthesia, is termed Avellis’s syndrome and is due to a lesion in the tegmentum of the

medulla. Investigations and treatment Brainstem syndromes occurring on a vascular basis are increas- ingly easy to diagnose because of MRI and MR angiography. Once a brainstem lesion is suspected, its detection and identification will require neuroimaging, preferably by MR techniques including diffusion weighted imaging and perfusion studies. The use of newer techniques in computed tomography (CT) imaging with dual energy and iterative reconstruction makes CT perfusion more usable and both MRI and CT complement interventional radiology. Interventional angiography may be used to direct the delivery of fibrinolytic agents, attempt the rescue of an embolus, or stent a narrowed portion of the vertebral or basilar artery but such techniques remain the subject of controlled trials and benefit is not yet established. Other investigations (e.g. to search for systemic disease including vasculitis), immunodeficiency, or sources of emboli, may also be re- quired. Vascular lesions within the brainstem often carry a remark- ably good prognosis, but, if the syndrome appears to be evolving, the possibility of anticoagulation must be considered. In those lesions in which damage affects the medulla, it may be important to protect the airway and avoid aspiration during the early phases of the illness. FURTHER READING Adams RD, Victor M (1989). Principles of neurology, 4th edition. McGraw-Hill, New York, NY. Caplan LR (1988). Vertebrobasilar system syndromes. In: Vinken PJ, Bruyn GW, Klawans HL (eds) Handbook of clinical neurology, pp. 390–3. Elsevier, Amsterdam. Chimowitz MI (2013) Endovascular treatment for acute ischaemic stroke: still unproven. N Engl J Med, 368, 952–5. Hurley MC, et al. (2012). Neuroimaging in acute stroke: choosing the right patient for neurointervention. Tech Vasc Interv Radiol, 15, 19–32. Renard D, et al. (2008). MRI-based score for acute basilar artery thrombosis. Cerebrovasc Dis, 25, 511–6. Medial lemniscus Cerebellum Medial longitudinal fasciculus Spinothalamic tract Corticospinal and corticobulbar tracts Sixth nerve Inferior lateral pontine syndrome Inferior medial pontine syndrome Seventh nerve Eighth nerve Descending tract and nucleus of five Dorsal cochlear nucleus Seventh nucleus Eighth nuclei Paramedian pontine reticular formation Sixth nucleus Fourth ventricle Fig. 24.9.6 Inferior pons at the level of nerve VI nucleus, showing the medial and lateral territories involved with occlusive stroke in this area. Reprinted with permission from Adams RD, Victor M, 1993, Principles of neurology, 5th edn. McGraw-Hill, New York. Medial lemniscus Medial longitudinal fasciculus Inferior olive Twelfth nerve Descending nucleus and tract of five Spinothalamic tract Lateral medullary syndrome Nucelus ambiguus Vestibular nucleus Tractus solitarius with nucleus Pyramid Twelth nucleus Medial medullary syndrome Sympathetic tract Tenth nerve Fig. 24.9.7 Cross-section of medulla at the level of the inferior olivary complex, showing the medial and the more common lateral territory involved with ischaemic stroke in this brainstem site. Reprinted with permission from Adams RD, Victor M, 1993, Principles of neurology, 5th edn. McGraw-Hill, New York.

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