|  |  |  |  |
| --- | --- | --- | --- |
| Testo di partenza  \*non tradurre le parti evidenziate in giallo | Testo tradotto dal candidato | Spazio a disposizione del correttore | Punteggio |
| A 22-year-old woman with a history of sickle cell disease and migraine headaches  presented to the emergency department with weakness that had caused her to fall  that day. She reported a gradual progression of weakness over a period of 6 months,  which had led in the previous few weeks to difficulty standing, climbing steps, and  lifting her child. |  |  |  |
| Her weakness was symmetric in both her arms and her legs, and she  had hand stiffness and dull, aching pain in both thighs. She reported no cognitive  impairment, paresthesia, numbness, or urinary incontinence. |  |  |  |
| The differential diagnosis for muscle weakness is broad. Weakness in the proximal  muscles of the arms and legs arouses suspicion for peripheral nervous system  dysfunction such as a myopathy, neuromuscular junction disorder, motor neuropathy  or neuronopathy, and polyradiculoneuropathy. |  |  |  |
| The patient has normal mentation,  absence of sensory symptoms, and intact bladder control, findings that  lessen suspicion of processes in both hemispheres of the brain, the brain stem,  or the cervical spinal cord as the cause of her weakness. The gradual progression  suggests inflammation, infection, and metabolic and neoplastic processes more  strongly than a vascular process. |  |  |  |
| The patient had no history of pain crises from sickle cell disease and was being followed  by a hematologist for chronic leukocytosis and iron overload resulting from  monthly transfusion therapy. She had received a diagnosis of moyamoya syndrome  at 5 years of age after she had had a stroke, and she had recovered without residual  deficits. |  |  |  |
| Magnetic resonance imaging and magnetic resonance angiography of the  head 3 years before the current admission showed hyperintensity in the left basal  ganglia and adjacent white matter, occlusion of the left terminal internal carotid  artery and middle cerebral artery, and a stenotic right middle cerebral artery with  collateral circulation. |  |  |  |
| Sickle cell disease can lead to moyamoya syndrome, which is characterized by  stenosis or occlusions of arteries around the circle of Willis in both hemispheres  of the brain, with prominent arterial collateral circulation. |  |  |  |
| Affected patients are at  risk for ischemic strokes. However, if recurrent ischemic strokes caused this patient’s  weakness, one would expect stepwise decline and asymmetric involvement  rather than gradual progression of symmetric weakness. |  |  |  |
| A review of systems revealed that the patient had  had an unintentional weight loss of 18 kg during  the previous 6 months despite having a good appetite  and eating regular meals. She had had intermittent  choking episodes and difficulty swallowing  over the previous 2 months, but reported no  difficulty chewing. |  |  |  |
| She also reported shortness of  breath with minimal exertion. Outpatient evaluation  had revealed hypoxemia, and administration  of supplemental oxygen through a nasal cannula  was initiated at a rate of 2 liters per minute. |  |  |  |
| Results  of computed tomographic (CT) angiography  of the chest and transthoracic echocardiography  performed during the outpatient evaluation were  normal. She reported no fever, chills, night sweats,  diarrhea, nausea, or vomiting during the previous  2 months. |  |  |  |