The neurological manifestations of dissecting aneurysm of the aorta

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Summary
Six cases of dissecting aneurysm of the aorta, four with cerebral manifestations, one with peripheral neuropathy and one with ischaemic necrosis of the spinal cord are reported. Cerebral manifestations include confusion and stupor, syncope, grand mal epilepsy, ischaemia of the spinal medulla, carotid artery occlusion and cerebral hemisphere infarction.

Ischaemic neuropathy is characterized by severe limb pain not of peripheral nerve distribution, but associated with areflexia and a peripheral pattern of sensory loss.

Ischaemic necrosis of the cord is characterized by flaccid paraplegia and sphincter disturbances with segmental sensory loss exhibiting an upper level on the trunk.

The differential diagnosis of the neurological complications of aortic dissection is discussed and a brief review of advances in management made.

Introduction
Dissecting aneurysm of the aorta is a comparatively common condition. Recent reports (Palmer & Wheat, 1967) indicate an improved prognosis as a result of advances in treatment, and in view of this, recognition of the numerous clinical syndromes associated with aortic dissection has become more important.

There are few case reports of the neurological complications or presentation of aortic dissection (Weisman & Adams, 1944) and because of the paucity of the literature we are reporting six cases illustrating the syndromes encountered.

Case reports
Case 1
A 78-year-old man suddenly lost consciousness: on examination his eyes were deviated to the left and the left pupil was small and did not react to light. There was neck rigidity and a flaccid right hemiplegia, accompanied by left extensor plantar response. Left carotid, brachial and femoral artery pulsations were absent and there was a systolic thrill and murmur over the right carotid artery (not observed in the previous examination). He rapidly deteriorated and died 2 days later.

Necropsy revealed a dissecting aneurysm of the aorta, involving the aortic arch and extending along the left carotid and subclavian arteries; the right innominate artery was not involved.

Case 2
A 54-year-old man with malignant hypertension noticed slight weakness of his left arm followed a few minutes later by a grand mal attack. When examined ½ hr later he was unconscious: there was a left upper motor neurone facial palsy and a spastic tetraplegia with bilateral extensor plantar responses. He rapidly deteriorated and died 24 hr later.

Necropsy revealed a dissecting aneurysm of the aorta involving both the innominate and left carotid arteries.

Case 3
A 59-year-old woman complained of tingling and weakness of both legs and mild chest pain while walking along a street. Within 5 min her speech became incoherent and she became drowsy. On examination she had a left upper motor neurone facial palsy and a flaccid left hemiplegia; tendon reflexes were sluggish and the left plantar response was extensor. Death occurred 5 hr after admission.

Necropsy revealed that a dissection of the aorta, beginning 1 in. above the aortic valve, had almost completely occluded the lumen of the innominate and left common carotid arteries.

Case 4
A 71-year-old man complained of sudden mild abdominal pain which lasted a few minutes. Half an hour later, he noticed tingling numbness and weakness of his legs. On examination he had a flaccid
paraparesis, and impaired sensation to pin prick in both legs; the tendon reflexes and plantar responses were absent. Femoral artery pulsations were delayed and weak, and the blood pressure was unrecordable in the legs. Death occurred 5 hr after admission.

Necropsy revealed that the common iliac arteries were obstructed by thrombus secondary to a dissection of the lower half of the abdominal aorta.

Case 5
A 38-year-old man with acromegaly, gout and hypertension complained of chest, abdominal and back pain followed by weakness and numbness of the legs and incontinence of urine. On examination, the legs were cold and pale and all peripheral arterial pulses in the lower limbs were absent: there was a flaccid paraplegia with sensory loss up to a level on the trunk corresponding to D.10. The patient rapidly deteriorated and died.

Necropsy revealed a dissecting aneurysm of the aorta, with an entry tear above the non-coronary aortic cusp. The aneurysm extended to the external and internal iliac vessels, occupied two-thirds of the circumference of the thoracic aorta and involved many intercostal arteries.

Case 6
An 86-year-old woman was admitted with a 10-hr history of confusion and syncope: there was no history of pain.

On examination she was anaemic (haemoglobin 7.7 g/100 ml), and arterial pulsations and blood pressure recordings in the legs were unequal. The patient deteriorated and died 4 days after admission to hospital.

Necropsy revealed that a dissecting aneurysm confined to the abdominal aorta had leaked retroperitoneally.

Discussion
The neurological complications of dissecting aneurysms of the aorta usually result from ischaemia of the brain, spinal cord or peripheral nerves. Interference with the blood supply to nervous tissue is most commonly brought about by extension of the dissection to, or severance of, contiguous arteries, but thrombus formation secondary to arterial narrowing may occur and obstruct branches, either by direct extension or by embolism.

Cerebral manifestations
The cardinal features of patients with cerebral manifestations of dissecting aneurysms of the aorta are coma and syncope. Patients may, however, present with severe headache or confusion and stupor (as in Case 6), sometimes alternating with lucid periods.

Cases 1, 2 and 3 all showed manifestations of carotid artery occlusion and ischaemia of the spinal medulla (Mergliano & Mongia, 1963). In Case 2 there was bilateral involvement of the carotid arteries but it is well recognized that cerebral blood flow via the basilar artery may be sufficient to sustain life. With the exception of one report (Shennon, 1934) major epilepsy (Case 2) has not previously been described and the left-sided meiosis found in Case 1 was probably due to Horner’s syndrome secondary to involvement of the sympathetic plexus overlying the occluded carotid artery.

Rapid onset of loss of consciousness may conceal a history of pain which is otherwise only rarely completely absent in patients with aortic dissection (Condon & Tanner, 1967). Occasionally because of severe pain, neurological symptoms and signs may be overlooked. Pain is uncommon in the great majority of cases of syncope with the exception of myocardial infarction, pulmonary embolism and acute abdominal catastrophes. These conditions have a clinical picture which can be easily confused with that of aortic dissection but neurological manifestations are rare although cerebral ischaemia may result from an embolus or hypotension.

The differential diagnosis of abdominal pain associated with neurological disturbances includes temporal lobe epilepsy, diabetic ketosis, and rarities such as acute porphyria, lead poisoning and ‘cerebral’ malaria; all of these may be excluded by the clinical picture and laboratory investigations.

Peripheral neuropathy
Because nerves receive several anastomosing nutrient arteries from adjacent trunks, ischaemic neuropathy can be produced by partial or complete occlusion of a main artery or by spasm of collateral arteries supplying the nerve. The clinical picture is usually dominated by sudden onset of severe limb pain which does not follow peripheral nerve distribution and may be referred distally. Numbness, tingling and coldness of the limbs follow or may rarely be (as in Case 4), the presenting feature. Muscle weakness, which is sometimes one of the initial complaints (Case 4), may be masked by immobilization of a painful extremity. However, the sudden onset of symptoms with areflexia and peripheral sensory impairment aids diagnosis by suggesting a vascular aetiology.

Ischaemic necrosis of the spinal cord
Necrosis of the spinal cord due to involvement of intercostal and lumbar arteries is uncommon; there are only nine such cases in the literature and in one of these there was necrosis not only of the cord, but also of the vertebral bodies (Hill & Vasquez, 1962).
Dissecting aneurysm of the aorta

Clinically spinal cord necrosis is characterized by a flaccid paraplegia and sphincter disturbances, with segmental sensory loss exhibiting an upper level on the trunk. If the peripheral arterial pulses are normal difficulty may arise in distinguishing aortic dissection from other acute transverse segmental sensory loss exhibiting an upper level on a flaccid paraplegia and sphincter disturbances, the trunk. If give pain, e.g. spontaneous abscess or tumour.

In spontaneous haematomyelia, the commonest site of haemorrhage is the cervical cord. The resultant pain in the upper limbs is rapidly followed by muscular weakness with diminution or loss of tendon reflexes; below the level of the cervical lesion there is usually a spastic paraplegia. Such a picture is most unusual with dissecting aneurysm of the aorta but dorsal or lumbar haematomyelia may closely mimic ischaemic necrosis of the spinal cord due to aortic dissection.

Myelitis develops less rapidly than aortic dissection and if part of an acute encephalomyelitis, cerebral symptoms may be prominent and are a helpful diagnostic feature. Herpes zoster myelitis is distinguished by its characteristic skin eruption.

Anterior spinal artery thrombosis may develop in the cervical lesion producing a quadriplegia, but more commonly the upper level of paralysis and sensory loss is the D.10 dermatome, when the condition is indistinguishable from intercostal artery occlusion due to dissecting aneurysm of the aorta.

Pathological fractures of the vertebrae can be differentiated from aortic dissection by the clinical picture and radiological investigations.

Examination of the CSF, which is normal in aortic dissection, may show increased protein and an excess of cells in transverse myelitis or a raised protein with or without xanthochromia in spontaneous haematomyelia. The CSF in spinal cord compression due to abscess may show a raised protein with an excess of cells. With spinal tumour not only is the protein raised, but xanthochromia is commonly found; cells, however, are usually absent from the CSF although an increase in mononuclears is rarely found.

Treatment

The recovery of patients with neurological complications of aortic dissection depends upon speed of diagnosis and promptness of treatment.

Immediate efforts should be made to prevent extension of the dissection and this is most effectively brought about by treatment with drugs and bed rest (Palmer & Wheat, 1967). The rationale for drug therapy places the main emphasis on reducing myocardial contractility and pulsatile waves in the large vessels rather than upon blood pressure control. In the acute stage, treatment with trimetaphan, reserpine and guanethidine was initiated and at a later stage propranolol and hydrochlorothiazide therapy was instituted in some subjects. A number of long-
term survivors have been reported following drug therapy and Fig. 1(a) and (b) is a transfemoral aortogram of one such patient who has survived 18 months following a fairly extensive aortic dissection.

If, however, in the acute stage of aortic dissection there is failure of the blood pressure to respond to therapy, surgery is advised. Immediate surgery is also indicated if there is severe aortic incompetence, failure to control progression of the dissection or involvement of innominate, carotid or spinal arteries with their resultant neurological complications.

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References

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