COEXISTENCE OF CEREBRAL ANEURYSMS WITH INTRACRANIAL ARTERIOVENOUS MALFORMATIONS

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Intracranial arteriovenous malformations, described as early as 1845 by Luschka, arise as the result of an anomalous development of the brain vasculature in an early embryonic stage. These lesions are thought to represent the persistence of primitive arteriovenous communications (Kaplan *et al*, 1961). A section of the vascular network forms lacking the interposition of a capillary bed which normally breaks the arterial flow and pressure. In due course, a tangled coil of abnormal blood vessels develops, which, if sizable, is capable of shunting a significant volume of blood away from the cerebral circulation.

Arteriovenous malformations, also known as cerebral angiomas, angiomatous malformations and arteriovenous aneurysms, are liable to bleed and produce subarachnoid or intracerebral haemorrhage. This is the commonest mode of presentation encountered locally. When intracerebral haemorrhage occurs, brain shift may take place; this seldom poses a grave threat to the patient's life, but may occasionally lead to a fatal outcome (Tay *et al*, 1969).

The association of a cerebral aneurysm with an arteriovenous malformation is uncommon (Perret and Nishioka, 1966). If berry aneurysms which may be fortuitously present are not considered, the type of aneurysms coexisting with malformations are those closely related with the lesion, arising either from a feeding artery or a draining vein. The incidence of these aneurysms is even more infrequent. The rarity has prompted us to report 5 such cases seen in a local series of 50 arteriovenous malformations collected over a 7½-year period (Tay *et al*, 1971).

Case 1

A 50-year-old Chinese assistant manager was warded on 1.2.69 with a week's history of severe headache of sudden onset, associated with transient blurring of vision. He also suffered intense photophobia 3 days prior to admission.

He was found to have no other neurological deficit except for a pronounced neck rigidity and a positive Kernig's sign. Lumbar puncture showed an initial xanthochromic cerebrospinal fluid which cleared up subsequently.

Bilateral carotid angiogram revealed the presence of a small arteriovenous malformation (Figs. 1 and 2) deep to the left frontal operculum. It was supplied by branches of the left middle cerebral artery and drained mainly by the corresponding Sylvian vein. A small intracerebral haematoma was also seen at the site of the malformation, as indicated by the brain distortion and shift, and another associated feature was the occurrence of a saccular aneurysm.

The patient was managed conservatively and discharged with full clinical improvement. He rejected the suggestion of operative intervention.

Case 2

A 17-year-old Chinese school-girl, who had been previously well, was warded on 24.2.69 with a complaint of constant and intense headache of sudden onset. She was found to be drowsy, and apart from neck stiffness, had no focal CNS signs. Heavily blood-stained cerebrospinal fluid was obtained on lumbar puncture.

A right carotid angiogram demonstrated a medium-sized arteriovenous malformation located over the genu of the corpus callosum (Figs. 3 and 4). The lesion was supplied mainly by branches of the right anterior cerebral artery. A sizable aneurysm was seen arising from the hypertrophied callosomarginal artery. The contrast was very rapidly drained away by the inferior sagittal sinus and the right internal cerebral vein into the straight sinus. A small intracerebral haematoma was also present in relation with the malformation, as reflected by the slight deviation of the right anterior cerebral artery to the left. The left carotid angiogram showed minimal cross-feed of the lesion by way of the anterior communicating artery.

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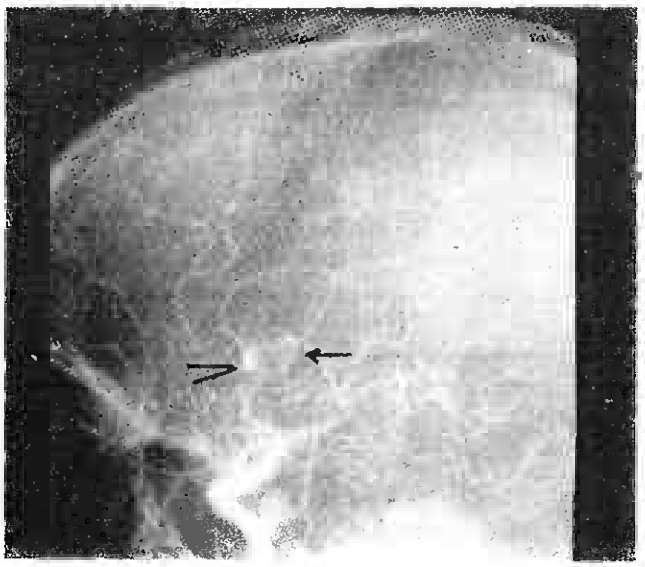

Fig. 1.

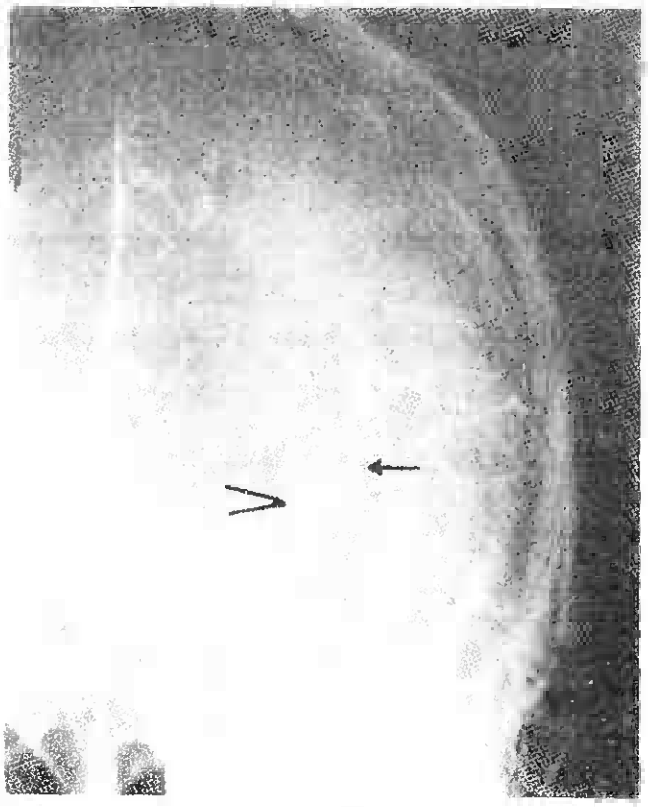

Fig. 2.

Figs. 1 and 2. Case 1: A small deep-seated arteriovenous malformation (⟵) associated with a saccular aneurysm (>).

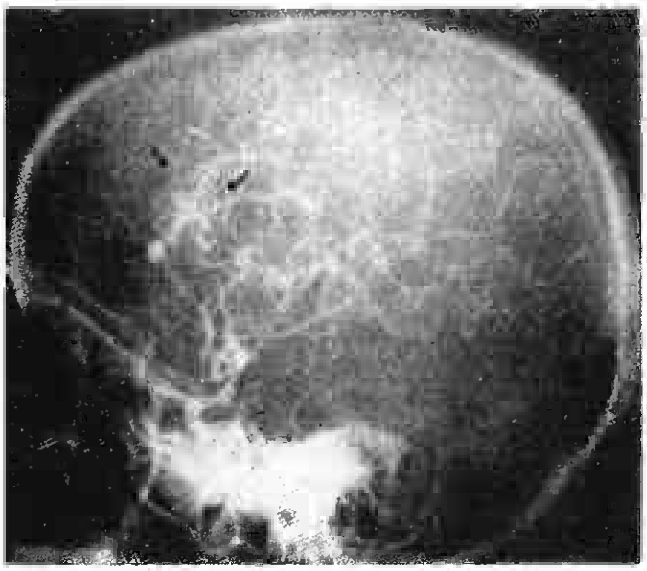

Fig. 3.

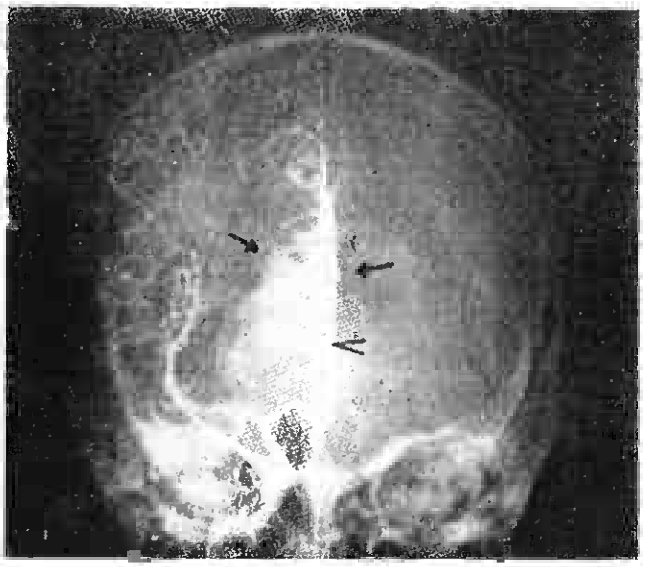

Fig. 4.

Figs. 3 and 4. Case 2: A medium-sized arteriovenous (⟵) in the right half of the frontal lobe. The aneurysm (>) related to it arose from the hypertrophied callosomarginal artery. The lesion was surgically excised.

The patient was operated in Bangkok where the lesion consisting of the arteriovenous malformation and the related aneurysm was excised. She made a good post-operative recovery and was now free of symptoms.

Case 3

A Chinese, aged 25, was a member of the Works Brigade when he was admitted to hospital on 24.8.65 with 2 attacks of generalised fits before lapsing into coma. He regained consciousness 6 days later. He had a similar attack at the age of 13.

In the ward, he showed signs of meningeal irritation. His left upper limb was smaller than his right and spastic. No other neurological deficit was elicited but the cerebrospinal fluid was uniformly blood-stained.

Bilateral carotid angiogram showed a large arteriovenous malformation on the convexity of the right frontal lobe. It was fed by hypertrophied branches of the right anterior and middle cerebral arteries. The left internal carotid also supplied the lesion via the anterior communicating artery. The drainage was effected by large superior cerebral veins emptying into the superior sagittal sinus. There was a saccular aneurysm at the periphery of the malformation fed by a large anomalous branch of the right cerebral artery. Two other aneurysms were suggested on the lateral radiographs but as these were not visualised on the anteroposterior view, they were, in all probability, kinks of dilated vessels.

The patient was discharged from hospital with improvement but defaulted from outpatient follow-up.

Case 4

A Chinese, aged 38, was a Shell employee from Brunei. He was transferred to Singapore after being hospitalised there for 2 months. Except for a history of a seizure 10 years, he had been well and free of fits until 3.4.65 when he suffered a Jacksonian attack involving the left face and arm. He was admitted to the Brunei hospital in coma and regained consciousness 2 days later.

He was found to have a left hemiparesis, left hemianaesthesia and left cerebellar deficit. Xanthochromic cerebrospinal fluid was noted on lumbar puncture. He was treated conservatively and made good progress. He was referred to Singapore for investigation of a possible berry aneurysm.

A right carotid angiogram demonstrated a medium-sized arteriovenous malformation (Figs. 5, 6 and 7) sited deep in the region of the trigone of the lateral ventricle. The points of supply were multiple and were derived principally from branches of the posterior cerebral artery. The drainage took place by way of the deep venous system, resulting in a marked fusiform dilatation of the great vein of Galen. A left carotid angiogram showed no cross-feed to this lesion but a vertebral angiogram revealed that the basilar artery contributed substantially to the malformation.

The neurosurgeon was consulted and he expressed the opinion that the operation was too hazardous to undertake in view of the numerous feeding vessels and the relative inaccessibility of the lesion.

In a letter from Brunei written 4 years after his last illness, the patient indicated that he was free of fits although he had occasional spells of giddiness. He had improved but his left limbs were still weak and clumsy and his hemianaesthesia remained poor.

Case 5

A 14-year-old Chinese school-girl was referred on 11.3.68 from a Government district clinic with a history of left-sided headache simulating migraine. She also had a birth-mark on the left forehead which had progressively increased in size in the last 2 years.

On examination, prominent dilated veins were noted on the left forehead and these were associated with a loud machinery bruit. No other significant physical signs were elicited. The cerebrospinal fluid was found to be normal.

A left carotid angiogram carried out under general anaesthesia revealed an extensive deep-seated arteriovenous malformation in the parietal lobe fed by branches of the left anterior and middle cerebral arteries. Drainage was directed mainly to the deep venous system, giving rise to an aneurysmal dilatation of the vein of Galen. Collateral venous return also took place through the left orbital plexus, accounting for the prominence of the superficial facial veins.

In view of the size of the lesion, operative exploration and correction were not considered. The patient was closely followed up and was relatively well except for an episode of epistaxis.

DISCUSSION

Five cases of intracranial arteriovenous malformations with coexisting aneurysms were presented. These aneurysms-developed from a major feeding artery or draining vein of the malformation and are thus distinguished from berry aneurysms which may or may not be anatomically related to the lesion. The diagnosis of only 1 case was

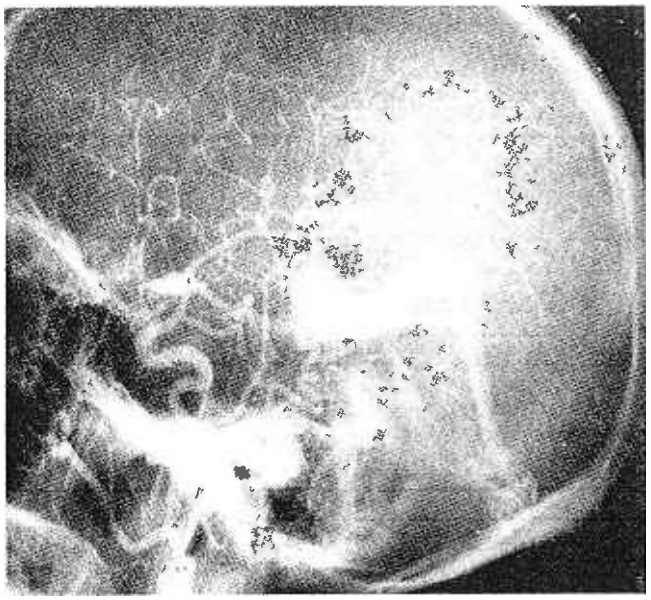

Fig. 5.

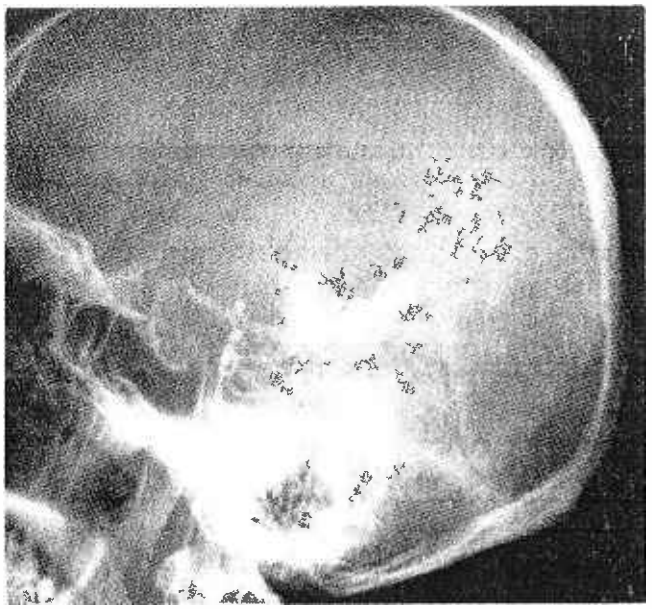

Fig. 7.

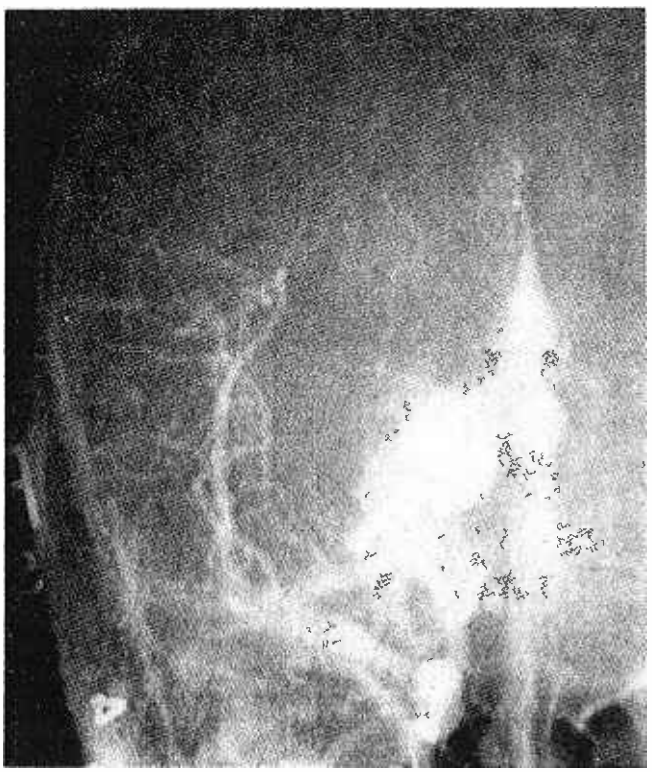

Fig. 6.

Figs. 5, 6 and 7. A medium-sized deep-seated arteriovenous malformation with secondary aneurysmal dilatation of the great vein of Galen, shown by right carotid angiogram (lateral and anteroposterior views) and vertebral angiogram.

ratified at craniotomy and the lesion excised. The other 4 lacked operative confirmation but had radiological appearance typical both of the malformation and the aneurysm.

There is unequivocal radiological proof of arteriovenous malformation. The most diagnostic feature is the rapid shunting of blood (or contrast) by the lesion. Early opacification of the draining veins is evident in all the cases. The increased workload has also given rise to hypertrophy of the feeding arteries and draining veins. The cluster of large abnormal blood vessels is another hallmark of the condition. An added feature is that, in the absence of haemorrhage, the lesion does not form a significant space-occupying intracranial mass.

The diagnosis of a coexisting arterial aneurysm has to be made with great caution. A possible pitfall is that in the presence of enlarged and tortuous vessels, a kink or "elbow" of such a vessel may erroneously be interpreted as evidence of an anuerysm. The diagnosis of an aneurysm is firmly established only when the localised pooling of contrast shows relatively consistent dimensions on at least 2 radiographic views and the possibility of artefacts caused by overlapping or tortuous blood vessels is excluded with certainty.

Russell and Rubinstein (1959) have adequately explained how this type of arterial aneurysms is formed. The normal lamination of the elastic and muscle fibres in the feeding arteries of the malformation becomes altered due to increased blood flow. The internal elastic lamella may be reduplicated, or interrupted and distorted. The media varies greatly in thickness in contiguous parts of the same vessel, and great thinning may lead to the formation of a large aneurysm. The pathogenesis therefore differs fundamentally from that of a berry aneurysm which arises as a result of developmental faults in the media at the points of arterial junction (Crawford, 1959).

Sugar (1951), one of the first to recognise this association, successfully resected 2 such saccular aneurysms which developed in relation to these malformations. The sacs were found on histological examination to be false aneurysms with no true vascular wall. These contained blood clot which was made up of multiple layers of various age, suggesting repeated occurrence of haemorrhage.

While the association of berry aneurysms with arteriovenous malformations has been widely recognised as shown in the review by Boyd-Wilson (1959), the coexistence of false arterial aneurysms with malformations seems to have escaped the attention of most commentators. Even in a comprehensive survey available in the "Report on the Cooperative Study of Intracranial Aneurysms and Subarachnoid Haemorrhage" (Perret and Nishioka, 1966), no distinction was made between berry and false aneurysms. Out of 502 cases of intracranial arteriovenous malformations investigated, 34 (6·8%) had coexisting aneurysms. Of these, 15 (3%) of the aneurysms were located on a major feeding artery to the angiomatous malformation while 19 (3·8%) were anatomically unrelated to the lesion. In this series, there were also 5 (1%) cases of aneurysms of the vein of Galen.

Aneurysms of the great vein of Galen is a well-known clinical entity. These may occur as an isolated lesion or as part of a complex arteriovenous malformation. Such malformations are invariably deep-seated so that a voluminous flow of blood is carried by the vein of Galen which undergoes aneurysmal dilatation. Unlike the solitary lesion which is frequently spherical, the aneurysm associated with a malformation is usually fusiform (Epstein, 1966).

The 2 cases presented showed few symptoms and signs commonly associated with this condition. The occurrence of the classical clinical picture is not invariable as evident in a review of cases by Thomson (1959) and is dependent on the size and site of the aneurysm producing obstruction to the flow of cerebrospinal fluid and compression of the mid-brain and brainstem.

Unlike the adverse prognosis of berry aneurysms, it is the general consensus of opinion that arteriovenous malformations are compatible with long life. Svien and McRae (1965) suggested that conservative management was appropriate for 80-85 per cent of such patients. They estimated that haemorrhage carried a 6 per cent mortality and the risk of dying from a subsequent haemorrhage after recovery from an initial bleed was also 6 per cent.

Where the malformation is complicated by the coexistence of a false arterial aneurysm, the prognosis is not so clearly defined. The assessment is made difficult by the fact that many workers failed to differentiate coexisting berry aneurysms from false aneurysms. In the case of aneurysms of the vein of Galen, the deep-seated malformation often precludes the possibility of adequate and successful surgical attack. The prognosis of such a case is determined to a large extent by the severity of pressure exerted on the brain and the degree of interference to the pathway of the cerebrospinal fluid.

SUMMARY

5 cases of intracranial arteriovenous malformations with coexisting aneurysms are presented.

These arose from a major feeding artery or draining vein of the malformation. It has been postulated that an arterial aneurysm of this type resulted from weakening of the elastic and muscular layers of the arterial wall due to increased blood flow. Similarly, aneurysmal dilatation occurred in a draining vein secondary to a voluminous flow of blood shunted by the angiomatous malformation. This study thus differs from most other works which are mainly concerned with the association of arteriovenous malformations with berry aneurysms which may or may not be anatomically related.

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