Case report

The Cobb syndrome

Association with hereditary cutaneous hemangiomas

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The Cobb syndrome is defined as a spinal cord arteriovenous malformation (AVM) associated with a cutaneous hemangioma (flame nevus) of the same metamere. The flame nevus is of significance in that it is suggestive of the presence of spinal cord AVM and indicates its location. The hereditary nature of this condition has been newly recognized.

Case report

A girl, 6 years and 5 months of age, was examined at the Cleveland Clinic in August 1973 because of a birthmark involving the left buttock. Birth and development had been normal, and she had been otherwise healthy and well. The left buttock was reported to have recently increased in size and the child's chief concern was related to her friends' comments about her asymmetry from the rear view. She also reported discomfort whenever she happened to fall in a sitting position.

Results of the physical examination were generally normal except for the region of the buttocks. The left buttock was larger than the right. A flame angioma covered most of the surface of the left buttock (*Fig. 1*). Deep cavernous elements were not identified in this angioma, but the increased size and warmth of the buttock were suggestive of a more extensive vascular anomaly than was apparent on the surface. Additional small flame angiomas were noted on both

hands and wrists, left anterior thigh, and left foot and ankle. A prominent pulsation was felt and a loud bruit was heard in the region of the coccyx. The rectal examination was negative. The left lower extremity was not overgrown and there were no abnormalities of strength, sensation, or reflex action. The heart was not enlarged and the blood pressure was normal. The diagnosis of a spinal cord AVM was made and the child was admitted to the hospital.

Chest roentgenograms were normal. A barium enema done to rule out an anterior meningocele was normal. Relevant laboratory values, including a platelet count, were within normal limits. Roentgenograms of the spine demonstrated spondylolysis along with a Grade I spondylolisthesis at the L-5 to S-1 level. Widening of the interpedicular distance from L-2 to L-4 was present as was flattening of the medial aspects of pedicles of the vertebral bodies at L-3, L-4, and L-5. The posterior surfaces of the vertebral bodies were scalloped in the lumbar region.

An arteriogram demonstrated a spectacular AVM of the spinal cord (*Fig. 2*). Subsequent selective catheterization revealed a major arterial supply through the artery of Adamkiewicz with large feeding vessels at T-9, T-11, L-1, and L-2. A small AVM was demonstrated in the area of the bifurcation of the aorta.

At this time, the parents and siblings were examined. The father had flame angiomas in several areas including the left arm, right chest, right groin, and left thigh. The 10-month-old male sibling had a flame angioma of the leg with no evidence of an AVM. It was recognized that the flame angiomas were hereditary in this family.

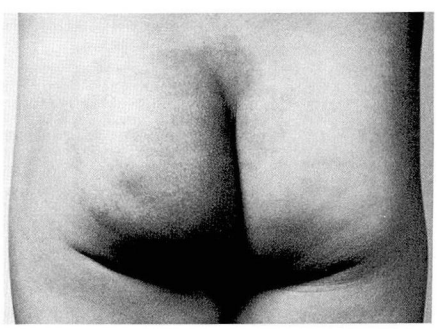

Fig. 1. Enlarged left buttock with large flame angioma.

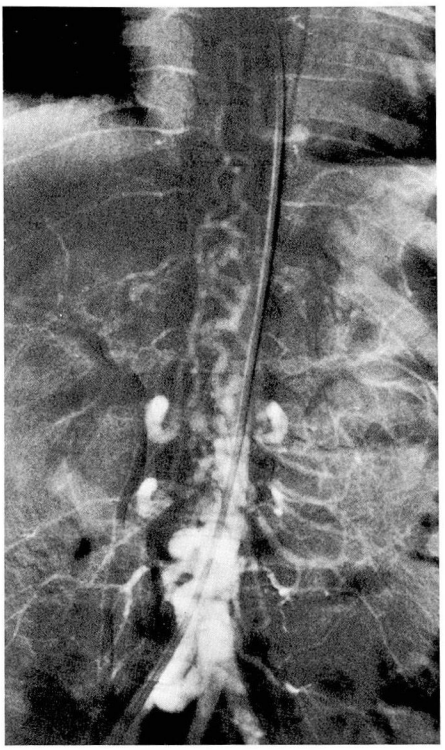

Fig. 2. High aortic injection demonstrates multiple large arteries feeding the huge arteriovenous malformation arising at T-9 and T-11 on the left, at L-1 and L-2 bilaterally and also between the aortic bifurcation.

This patient had previously been admitted to Akron Children's Hospital where an arteriogram had demonstrated the spinal cord AVM. This information was concealed from us by the parents until our own studies were completed. It was a case of examining the examiner.

Surgical intervention was not considered wise and the patient was discharged to be followed in the outpatient department. She remained well until May 1977, when she was readmitted with throbbing

pain in both upper thighs and lower abdomen. She had a slight limp and slight weakness of the left leg with no sensory or reflex abnormalities. Her symptoms subsided and she was again discharged for outpatient care.

Discussion

Cobb,[1] in 1915, first clearly described an angioma of the spinal cord associated with a nevus of the "port wine" variety of the same metamere. He referred to a previous case described by Berenbruch[2] in 1890 in which an angiolipoma of the back was connected with a large cavernous plexus of veins along the spinal column and by way of the intervertebral foramina with an angioma of the dura. There was also an extensive angioma of the cord. In Cobb's case, it was Harvey Cushing who recognized the significance of the angioma of the skin in predicting that the cord lesion would be an angioma. Cushing,[3] in 1906, had described three cases that we would now term the Sturge-Weber syndrome. Autopsy findings were described in one of these and operative findings in the second. Weber's[4] contribution to the syndrome which bears his name did not come along until 1922 and was only clarified when better roentgenograms of his patient were available in 1929.[5]

The Cobb syndrome has been thought to be uncommon and only 17 cases were accepted by Kissel and Dureux[6] in their extensive review in 1972. It is more likely, however, that the condition has been underdiagnosed. Doppman et al,[7] in 1969, carefully searched for segmentally related cutaneous hemangiomas in a series of 28 patients with spinal cord AVM and found six. This is a much higher prevalence than previously reported. Selective arteriography in each case demonstrated that the cutaneous and the spinal angiomas were supplied by posterior branches of the same intercostal artery. Additional arterial feeders were also present.

Although we recognized the hereditary nature of the cutaneous angiomas in our case in 1973, this association was first published by Kaplan et al[8] in 1976. Kissel and Dureux[6] specifically stated "The literature contains no familial cases of Cobb syndrome and no author mentions the presence of isolated skin naevus in the patients' relatives." Again, it would seem most likely that the hereditary nature of these angiomas when associated with the Cobb syndrome has not been recognized, because it has not been considered or investigated. The Cobb syndrome should take its place with other dominant hereditary syndromes of cutaneous angiomas associated with vascular malformations of the cord such as the von Hippel-Lindau and Rendu-Osler-Weber syndromes.

References

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