Coarctation of the Aorta with Congenital Hemangioma of the Face and Neck and Aneurysm or Dilatation of a Subclavian or Innominate Artery

A New Syndrome?

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Congenital cutaneous hemangioma associated with congenital heart disease is extremely rare. We report four infants with congenital cavernous hemangioma of the face and neck and coarctation of the aorta. Three also had congenital aneurysm of a subclavian or innominate artery. One patient also had mild congenital valvular aortic stenosis. The unusual combination of lesions may represent a new syndrome.

The presence of cardiac anomalies with cutaneous hemangioma is rare, and so is the association of peripheral vascular anomalies with coarctation of the aorta. The files of 68 patients with coarctation of the aorta studied in the Heart Institute, the Sheba Medical Center, between 1971 and 1980, were reviewed for the presence of congenital hemangioma and other vascular anomalies. Clinical history, physical examination, electrocardiogram, thoracic roentgenogram, cardiac catheterization and angiocardiography were available in all cases.

RESULTS

In four patients, congenital hemangioma of the face and neck was found. Three had congenital aneurysm of a subclavian or innominate artery (Fig 1), one with associated mild congenital valvular aortic stenosis. The remaining patient had mild mitral insufficiency. Clinical data are presented in Table 1.

DISCUSSION

Congenital hemangiomas are common dermatologic disorders in infants, in whom they may be found as a single lesion or as multiple hemangiomas.1,2,4 Primary congenital heart disorders have not been reported in association with hemangiomas in infancy. None of the 2,400 children who underwent cardiac catheterization in our laboratory, except the four reported here, presented with congenital facial hemangioma. Cutaneous and visceral hemangiomas may, however, be associated with cardiac hypertrophy and dilation secondary to large

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Figure 1, Case 1. Angiocardiography in the anteroposterior projection with injection of contrast material into the right ventricle. In the levophase, coarctation of the aorta (arrow) is demonstrated in the usual position, with a large aneurysm of the subclavian artery. In the left side of the neck, an abnormal vascular plexus is filled. (A) aneurysm; (P) plexus.
arteriovenous shunts.\textsuperscript{1,5,6} Thus, the presence of cutaneous hemangiomas with signs of heart disease should raise the suspicion of secondary high-output heart disease.

Our four cases represent a new entity in the differential diagnosis of hemangiomas of the face and neck with signs of heart disease, i.e., coarctation of the aorta with aneurysm or dilatation of a subclavian or innominate artery and with or without valvular aortic stenosis. Palpation of the femoral pulses can easily differentiate between these lesions and a high cardiac output state.

CVA, rupture of Berry aneurysm, and subarachnoid hemorrhage are well known in coarctation of the aorta. Other associated peripheral vascular anomalies were not reported. Coarctation of the aorta was defined by Edwards and associates\textsuperscript{6} as a localized narrow, constrictive lesion which presents histologically as an infolding of the aorta media into the lumen.\textsuperscript{5,6} As such, coarctation of the aorta is considered to be a localized lesion, and not part of a generalized vascular disorder, although other congenital cardiac defects may be associated with it.\textsuperscript{3}

The associated lesion in our cases suggests that coarctation of the aorta may, in some instances, be part of a widespread vascular lesion. The possible relationship of cutaneous hemangioma to cerebrovascular anomalies in patients with coarctation of the aorta is yet to be determined. The marked similarity between our patients, in view of the rarity of congenital cardiac lesions in patients with congenital cutaneous hemangioma, may suggest that they are cases of a new syndrome. A large scale study is needed to determine the statistical significance of this phenomenon.

Another possible explanation for the unusual association is that the coarctation resulted from severe ishmal hypoplasia, due to increased flow through the hemangiomas.

A case which may support the latter theory was reported in 1969.\textsuperscript{7} In a four-day-old infant with congestive heart failure, the femoral pulses were not palpable. Angiocardiography demonstrated what seemed to be a filling defect in the aortic isthmus with slow filling of the descending aorta. Autopsy revealed no congenital heart disease. The abnormal findings were due to a cerebral arteriovenous fistula which caused decreased flow to the descending aorta.

\textbf{REFERENCES}

1 Cooper AG, Bolande RP. Multiple hemangiomas in an infant with cardiac hypertrophy. Pediatrics 1965; 35:27
6 Edwards JE, Christensen NA, Clagett OT, McDonald JE. Pathologic considerations in coarctation of aorta. Proc Mayo Clinic 1948; 23:334