CASE REPORT

Congenital Subclavian Steal Syndrome Associated with Right Aortic Arch

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Since the initial reports by Contorni (1960) and Toole (1961) concerning the reversal of blood flow in the vertebral artery, several similar reports have been published (Reivich *et al.*, 1961; Parrott, 1964; Swales and Farrow, 1964; Steinberg and Halperin, 1963; Patel and Toole, 1965). This condition became known as the subclavian steal syndrome.

Almost all the reported cases describing the anatomical, clinical, pathophysiological, and angiographic findings were in adults with acquired lesions. A review of the published reports revealed that there were only 3 cases of congenital subclavian steal syndrome (Daves and Treger, 1964; Massumi, 1963; Stewart, Kincaid, and Edwards, 1964).

The purpose of this communication is to report one additional case of congenital subclavian steal syndrome, with right aortic arch.

Case Report

A 19-year-old white man was admitted to the cardiac service of Deborah Hospital because a right aortic arch was detected on a routine chest x-ray, in association with a difference in the amplitude of the brachial arterial pulses. He was born of a normal pregnancy and delivery, and his childhood as well as his adolescent life had been free of cardiovascular or neurological symptoms.

The past medical history was unremarkable. The family history did not show diabetes, hypertensive cardiovascular disease, or any other significant illnesses.

Physical examination showed a well-developed white man in no distress. His temperature was 37.2°C (99°F.) and his pulse rate was regular at 86/min. The right brachial and radial arterial pulses were normal. The left brachial and radial arterial pulses were of diminished amplitude, and their peaks appeared later than those of the right. The blood pressure on the right arm was 108/70 and on the left arm 80/60 mm.Hg.

The thyroid gland was not palpable and the neck veins were not distended. A systolic bruit was heard over the right and left supraclavicular arteries (Fig. 1). Examination of the chest revealed a quiet praecordium. The point of maximum impulse was not palpable. On auscultation the cardiac rhythm was regular with a rate of 86/min. No murmurs or abnormal sounds could be heard. The lungs were clear to percussion and auscultation. The abdomen was soft and nontender. No abnormal masses could be palpated. The femoral pulses were normal. Neurological examination did not show any abnormalities.

The electrocardiogram showed a normal sinus rhythm. The QRS axis was plus 45 degrees. The tracing was considered to be within normal limits.

Chest x-rays taken in the conventional positions with barium swallow revealed that the heart was normal in size; the pulmonary artery was slightly dilated; and the only outstanding feature was a right aortic arch (Fig. 2).

All the routine clinical and laboratory examinations were within normal limits.

A right heart catheterization disclosed no intracardiac or intervascular shunts, or any obstructive lesion of the right heart. A difference of 32 mm.Hg between the right and left brachial arterial systolic pressures was noted (Fig. 3). Retrograde aortography confirmed a right aortic arch. In addition, the proximal left subclavian artery was not visualized (Fig. 4a). The distal subclavian artery was later filled via the left vertebral artery (Fig. 4b).

Surgical intervention to correct the abnormality was not recommended at this time because of the absence of symptoms.

Discussion

The subclavian steal syndrome may result from three known causes. It may be due to: (1) arteriosclerotic changes in the arteries arising from the aortic arch, causing partial or complete obstruction; (2) surgery, such as the subclavian-pulmonary

FIG. 1.—Phonocardiogram recorded over the right and left infraclavicular areas. Note ejection click and systolic ejection murmur.

artery anastomosis (Blalock-Taussig operation) or a complication following correction of coarctation of the aorta and, (3) congenital reversal of flow through the vertebral-subclavian arterial system due to vascular anomalies. The latter is extremely rare. Daves and Treger (1964) and Stewart *et al.* (1964) reported 2 cases of congenital reversal of flow in the vertebral-subclavian system. This occurred in infants who had associated aortic arch anomalies. Massumi (1963) reported another case which, in addition, showed the right aortic arch ascending to the cervical region. None of these cases was known to have symptoms which could be attributed to the congenital subclavian steal. The patient in this report was asymptomatic. In addition to symptoms, if present, two important clues should make one strongly suspect the diagnosis of congenital subclavian steal syndrome. These are the difference in the amplitude of the arterial pulses and the blood pressures in the two arms and the *presence of a right aortic arch or arch anomalies.*

Once the possibility of the existence of the syndrome is entertained, a retrograde aortography aimed at delineating the aortic arch and its main branches should be performed for verification. Retrograde blood flow through the ipsilateral vertebral artery into the distal subclavian artery must be shown before the final diagnosis is established.

Summary

Congenital subclavian steal syndrome is extremely rare, only 3 cases having been reported previously.

One additional case is reported, also showing a right aortic arch, in which symptoms were absent.

Two important features that should raise the index of suspicion regarding the occurrence of this entity are the differences in the amplitude of the arterial pulses and blood pressures in the upper extremities and the presence of a right aortic arch or arch anomalies. A diagnosis can best be established by retrograde aortography demonstrating

FIG. 2.—Chest x-ray in the postero-anterior projection, demonstrating a normal cardiac silhouette and right-sided aortic arch.

FIG. 3.—Simultaneous right and left brachial arterial pressures obtained through an intra-arterial needle, demonstrating the difference in systolic pressures.

FIG. 4.—(A) Retrograde aortogram. Immediately after the injection, observe the right-sided aortic arch, innominate artery, and left common carotid artery. The left subclavian artery is not yet opacified. (B) Three and one-half seconds after the injection, the left subclavian artery fills distal to the origin of the left vertebral artery.

retrograde flow through the ipsilateral vertebral artery which in turn supplies the distal subclavian artery.

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STATEMENT BY THE EXECUTIVE COMMITTEE OF THE INTERNATIONAL SOCIETY OF CARDIOLOGY

Since the time of Hippocrates, the medical profession has preserved the ethics governing the extent of the information conveyed to the public at large and the way in which it is conveyed. With the availability today of modern media for mass communication there is a greater need to ensure that such information is spread in a responsible manner.

We deplore the fact that in recent times medical and surgical experiments have become matters of public entertainment and even sensationalism. Such a trend can only bring discredit to the profession as a whole and indirectly misrepresent to the public, who are not in a position to judge the implications of such developments, the dangers and limitations inseparable from such procedures in their initial phase.

The Council of the International Society of Cardiology feels that a lead must be given by responsible members of the profession. One method of avoiding extremes of anxiety or misplaced hope is to suggest strongly that no new procedures, either medical or surgical, are released to the lay press before being published in the reputable medical journals after full scientific evaluation.

The International Society of Cardiology, through its Council, feels strongly that the profession as a whole should support this view in the interests firstly of the patient and secondly of their standing and dignity. The world at the moment is in a scientific turmoil and many people are looking increasingly to doctors for moral leadership and calm judgment on the value of radical and sometimes irreversible surgical procedures, and on methods of patient investigation which may not be strictly necessary for diagnosis or therapy.

In regard to cardiac transplantation, the Executive Committee of the International Society of Cardiology entirely endorse the statement of the Board of Medicine of the National Academy of Sciences in the United States (release of 28 February 1968), which says in part: "The surgical team should have had extensive laboratory experience in cardiac transplantation. . . ." "Investigators skilled in immunology . . . should be readily available as collaborators. . . ." "Rigid safeguards should be developed with respect to the selection of prospective donors and the selection of prospective recipients. An independent group of expert, mature physicians . . . should agree and record their unanimous opinion as to the donor's acceptability on the basis of the evidence of crucial and irreversible bodily damage and imminent death." "... the Board strongly urges that institutions . . . without specific capabilities to conduct the whole range of scientific observations involved in the total study, resist the temptation to approve the performance of the surgical procedure until there has been an opportunity for the total situation to be clarified by intensive and closely integrated study." The Committee further finds itself in complete agreement with the Conseil National Francais de l'Ordre des Médecins (release of 3 May 1968) who have declared that in future, to avoid the diffusion of erroneous information, announcements of such experiments should be the subject of an official bulletin, which respects medical ethics and avoids distress to relatives and the creation of an emotional public reaction.

Geneva, 9 May 1968

Kempson Maddox—President