Congenital Arteriovenous Fistulas of the Thoracic Wall

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Arteriovenous fistulas of the thoracic wall may produce murmurs and thrills which suggest an intrathoracic vascular abnormality. Congenital arteriovenous fistulas in the precordial region, the site of the fistula in the case here reported, may be confused with anomalies of the heart and great vessels. The differential diagnosis of blood vessel tumors of the chest wall is discussed. Occasionally arteriovenous fistulas involve both the thoracic wall and intrathoracic viscera.

Congenital arteriovenous communications within the structures of the thoracic wall are uncommon. Such lesions are of interest to the cardiologist, however, since occasionally they may be confused with intrathoracic vascular abnormalities. Moreover, the pathologic process may involve both the chest wall and the intrathoracic viscera, either as a single or as separate lesions.

If a congenital arteriovenous fistula in the precordial region causes a continuous murmur and thrill, the clinical features may simulate those of a cardiac lesion. A patent ductus arteriosus had been the original diagnosis in a case which we are reporting, but a few years later, when a mass was palpable in the chest wall, it was realized that the lesion was extracardiac.

Case Report

R. M., a woman, was 30 years old on admission to Lenox Hill Hospital in January 1947. Twelve years previously, when she was examined while attending college, a murmur was heard in the precordial region and a patent ductus arteriosus was diagnosed. Five years later, the patient palpated a soft mass underlying the inner portion of the left breast and became conscious of occasional throbbing in that region. The area was slightly tender on pressure. A diagnosis of aneurysm of an intercostal or internal mammary artery was made by Dr. Ernst Boas. Operation was recommended but was not carried out at that time.

The prominence of the inner portion of the left breast increased very slightly during the next seven years. The mass, which was quite inconspicuous, became more evident when the patient was excited. There had never been any cardiac symptoms. There was no history of trauma.

Physical examination revealed a well developed and well nourished woman in apparent good health. There was no cyanosis or clubbing of the fingers. The breasts were symmetrical but there was a slight rounded prominence in the medial portion of the left breast. The overlying skin showed no discoloration. Palpation revealed a continuous thrill which was diminished by firm pressure at the level of the third intercostal space just to the left of the sternum. On auscultation there was heard a loud, continuous, rough murmur with systolic accentuation, which was transmitted out to the left. There were no other abnormal findings over the rest of the precordium. The blood pressure was 130/80. The remainder of the physical examination, including fluoroscopy, revealed no abnormality. There was no cardiac enlargement. Roentgenogram in lateral projection showed questionable evidence of phleboliths in the region of the lesion. A preoperative diagnosis of congenital arteriovenous fistula of the internal mammary vessels was made. Operation was performed on January 18, 1947.

An incision was made in the region of the second costal cartilage which was resected. The internal mammary vessels were exposed at this site and found to be several times larger than normal. The artery and vein were ligated and divided with resultant diminution in the thrill and pulsation in the upper mesial portion of the vascular mass. Dissection was then carried downward and other branches from the internal mammary vessels entering from the medial aspect were divided. One large vessel perforated the intercostal muscle just lateral to the sternum and was ligated superficial to the intercostal muscle. The skin incision was extended and the breast, which was uninvolved, was reflected downwards and laterally. The lateral portion of the pectoralis major muscle was thus exposed. Two
large pulsating vessels could be felt entering this muscle in the axillary region. These vessels were abnormally large branches of the acromiothoracic and lateral thoracic arteries. After ligation of these vessels, the pulsations ceased. The pectoralis major, which contained the mass of blood vessels, was then excised except for the uninvolved clavicular and axillary portions. At operation the gross findings were interpreted as those of a cavernous hemangioma of the pectoralis major muscle. The breast was replaced and sutured in position. The postoperative course was uneventful. The patient was well when seen two years after operation and there were no signs of recurrence.

Pathologic Findings. On gross serial section of the operative specimen a number of thick-walled dilated blood vessels were found scattered through the central portion of the muscle. A microscopic section cut longitudinally paralleling the fibers of the muscle showed a group of arteries and veins. The arteries were conventional in the arrangement of muscle and elastic tissue save for the occasional thickening of the subintimal tissues and interruption of the internal elastic lamina by fibrosis. The veins were for the most part thick walled and fibrous—similar in structure to varicose veins. There was one unusual feature: although the muscularis was not hypertrophic and did not have the appearance of arterial muscle, there was in places an internal elastic lamina which had some resemblance to the arterial type. This was found at one part of a long tortuous vein and was absent at the other. The two parts were joined by a very narrow communication. The appearance suggested a fistulous communication (fig. 1). In another area, two thick-walled veins were shown with a marked proliferation of capillaries, arterioles, and venules between them (fig. 2). The whole picture suggested the formation of fistulous communications between arteries and veins with some tendency for the elastic tissue of one vein to approximate the appearance of the internal elastic lamina of an artery. There had also occurred a tumor-like proliferation of capillaries, venules, and arterioles—a phenomenon which has been noted in other cases of congenital arteriovenous fistulas.

Discussion

At the time of operation a diagnosis of cavernous hemangioma of the pectoralis major muscle was made. On microscopic examination of the lesion this diagnosis was proved to be incorrect. A cavernous hemangioma is a vascular tumor composed of blood vessels of the order of capillaries enlarged into cavernous spaces filled with blood. No such formations were seen in our specimen. The possibility of cirrhotic aneurysm and venous racemose aneurysm may both be excluded as well. The former is composed exclusively of a congeries of vessels of the order of arteries and the latter exclusively of veins. In our case a congeries of large vessels was present but both arteries and veins were represented with probable fistulous communications between them. In addition there was a proliferative conglomeration of small vessels including capillaries, arterioles, and venules.

An arteriovenous fistula may occur in various structures of the chest wall. Often more than one tissue is involved. If the vascular mass lies within a skeletal muscle, differentiation between cavernous hemangioma of muscle and arteriovenous fistula may not be apparent on gross inspection.

Hemangiomas of skeletal muscle are relatively uncommon and muscles of the trunk are rarely involved. Shallow and his associates collected 335 cases of hemangioma of striated muscle from the literature. In this group there were only 63 cases in which the muscles of the trunk were involved and 39 of these hemangiomas occurred in the chest wall. Hemangiomas are most common in the muscles on the posterior aspect of the thorax. Only 6 cases of hemangioma of the pectoralis major muscle have been reported. In 2 of these the adjacent muscles and skin were involved.

Acquired arteriovenous fistulas, chiefly as the result of a penetrating wound, are far more common than congenital lesions. Occasionally it is not possible to differentiate between those of congenital and those of traumatic origin, especially in cases of nonpenetrating injuries. An arteriovenous communication is rarely established through erosion of the blood vessels.

Fig. 1.—(Upper illustration) Low-power photomicrograph made through the pectoralis major muscle, fibers of which are shown above and below. Between them are the large vessels; the rounded ones are arteries cut in cross section and the longitudinal ones are thick-walled veins. Near the right-hand end a large vein communicates by an exceedingly narrow lumen with a larger vessel which is venous in aspect but has an incompletely developed internal elastic lamina.

Fig. 2.—(Lower illustration) Low-power photomicrograph showing two thick-walled veins and between them a massive circumscribed proliferation of capillaries, venules, and arterioles.
by an inflammatory or neoplastic process. The infrequency of traumatic arteriovenous fistula of the chest wall as compared with traumatic intrathoracic arteriovenous communications is indicated by Schumacker's statistics. Of 354 traumatic arteriovenous fistulas and arterial aneurysms only a single one involved a vessel of the chest wall; this was an arteriovenous fistula of the internal mammary artery. Neoplasms involving the chest wall, especially the sternum, may occasionally manifest considerable pulsation and thus be confused with aneurysms and arteriovenous fistulas. Metastatic carcinoma of the thyroid gland and hypernephroma are the neoplasms most likely to present such a clinical picture. Marked vascularitiy of the tumor mass usually accounts for the pulsation. When a tumor or inflammatory process causes destruction of the sternum, a pulsation may be transmitted from the large underlying vascular structures in the mediastinum.

Since intrathoracic aneurysms with projection into or through the chest wall are more common than pulsating vascular masses arising in the thoracic parietes, aneurysm of the aorta or of other large vessels is usually to be considered first in the diagnosis. Fluoroscopy and angiocardiography may be of great aid in the differential diagnosis.

Arteriovenous communications in the deeper layers of the chest wall may cause notching of the lower borders of the ribs such as is usually associated with coarctation of the aorta. Similar rib notching resulting from the collateral circulation has been observed in a case of tetralogy of Fallot without coarctation of the aorta.

Arteriovenous fistulas of the thoracic wall may be confused with intrapulmonary arteriovenous fistulas. Most patients with the latter lesion also have small hemangiomata in the skin and mucous membranes. The characteristic features of a pulmonary arteriovenous fistula are cyanosis and polycythemia associated with roentgen-ray evidence of one or more lesions in the lungs. Cyanosis and polycythemia are occasionally absent. A continuous murmur may be heard over the area of the lesion in the lung. Angiocardiography establishes the diagnosis. Surgical removal of the involved portion of the lung cures the pulmonary lesion.

Clagett and Burchell reported a case of pulmonary arteriovenous fistula of the right middle lobe associated with marked enlargement of the internal mammary artery and veins. Owing to the large quantity of blood which entered the lung from the vessels in the thoracic wall, the lobe became more and more congested as the hilar structures were ligated during operation. Separation of the lobe from the chest wall was accompanied by severe bleeding. One of us has observed a patient with arteriovenous fistulas in the chest wall on one side and apparently an arteriovenous fistula in the lung on the other side causing hemoptysis. This patient also had notching of the ribs on the side of the arteriovenous communication in the chest wall.

**SUMMARY**

Congenital arteriovenous fistulas in the thoracic wall may simulate intrathoracic vascular lesions. A case of arteriovenous fistula in the precordial region which originally presented physical signs suggestive of patent ductus arteriosus is reported. The lesion was successfully removed by operation.

**REFERENCES**


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