PULMONARY ARTERIOVENOUS ANEURYSM*

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ALTHOUGH PULMONARY ATRERIOVENOUS ANEURYSM was recognized at autopsy by Churton[1] as early as 1897 and by Wilkens[2] in 1918, correlation of these lesions with a clinical syndrome leading to premortem diagnosis was apparently not made until 1939, by Smith and Horton.[3] The first reported surgical treatment, a successful total pneumonectomy, was carried out by Shenstone in 1942,[4] the patient having been previously studied and a diagnosis made by Hepburn and Dauphinee.[5] Since then over 50 cases have been recorded, many of them surgically treated, and it has become apparent that previous confusion of these lesions with congenital cyanotic heart disease, polycythemia vera and various benign lung tumors has delayed their recognition and appropriate surgical treatment.

An excellent and comprehensive survey of the literature, including some unreported case experiences, was published by Yater, Finnegan and Giffin in 1949.[6] It is the purpose of this paper to record our experience with an additional four patients, three of whom were treated surgically and each with a satisfactory outcome. The surgically excised lobes were prepared by vinylite-plastic bronchovascular casting technic,[7] and the results illustrated and summarized (Figs. 2, 4 and 6).

CLINICAL CONSIDERATIONS

The clinical syndrome in the fully developed case of pulmonary arteriovenous aneurysm is quite readily recognized. The essential points are cyanosis and clubbing in the presence of a normal heart, together with roentgenographic findings of a vascular tumefaction in the lung fields.

Cyanosis may however be entirely absent (*e.g.*, Cases 1 and 4). It usually appears in childhood, but sometimes rather abruptly in adult life. Dyspnea is slight at first but may be progressive. The "squatting" tendency observed in congenital cyanotic cardiacs has been described in one reported case.[45] Clubbing of the fingers and toes is nearly constant, if cyanosis is present, and usually noted to have about the same time of onset. It may be associated with arthralgic pains[8] and with a radiologic demonstration of pulmonary osteo-arthropathy. Bleeding from the nose is a common complaint (Cases 1 and 4), less commonly oozing from superficial cutaneous or visceral hemangiomas.[9] Hemoptysis may occur to a severe degree and may be lethal.[10]

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Symptoms of central nervous origin may be experienced, especially when the polycythemia is well developed. These include headache, vertigo, weakness, syncope, convulsions, paresis or paresthesia of the extremities, dysphagia and thickness of the speech.

A few patients have been aware of some discomfort in the chest, and an abnormal fremitus. Actual chest pain is not common unless a complicating adhesive pleurisy exists,[8] a not uncommon operative finding. General weakness, lassitude, and loss of weight have all been described; the latter has been quite marked in some cases (cf. Case 3).

The *physical findings* are principally those of cyanosis in variable degree and clubbing of the digits, but one or both of these may be absent (Case 1). In about half of the reported cases multiple "spidery" telangiectasia and discrete nodular hemangiomas of the face, neck, lips, mucous membranes or trunk are encountered, emphasizing the relationship to hereditary telangiectasia of the Rendu-Osler-Weber type (*e.g.,* Cases 2 and 4). Despite cyanosis and dyspnea, the heart is usually found to be normal in size, shape, and free of organic murmurs, except in cases with terminal decompensation or unrelated cardio-vascular disease (Cases 2 and 3). Blood pressure recordings are usually normal, as is the venous pressure by direct measurement, although veins of the retina, head, and neck may appear prominent. On auscultation over the site of the aneurysm there may sometimes be heard a continuous rough humming murmur accentuated in systole and in deep inspiration (Cases 3 and 4). In rare instances the murmur may be exaggerated by forced expiration.

In contradistinction to polycythemia vera, the spleen is normal in size or only suggestively enlarged.

LABORATORY FINDINGS

Typically there is an increase in erythrocyte count and hemoglobin content of the blood. In some cases these have been observed to increase over a period of observation. Values as high as 11.45 million red cells,[11] and 24.9 Gm. hemoglobin[12] have been recorded. In a small minority the red cell count and hemoglobin are normal or low; in the latter situation there will usually be a history of repeated epistaxis, hemoptysis, or unrelated causes of anemia. The hematocrit level tends to follow the polycythemia.

The total blood volume in the polycythemic cases is of course elevated, but the plasma volume shows little or no increase, in contradistinction to the findings with peripheral arteriovenous fistula.[3] Cardiac output is not usually increased.[13, 45]

The peripheral arterial oxygen saturation is usually decreased, even as low as 63 per cent,[13, 45] and decreasing further with exercise. The actual blood oxygen content may be relatively high due to the polycythemia.

The venous circulation time is normal, and biochemical studies of the blood plasma show no significant deviations from normal, unless a decrease in carbon-dioxide.

DIFFERENTIAL DIAGNOSIS

1. *Polycythemia rubra vera.* In this disease the age group is generally older; there is splenomegaly, with abnormalities of blood and bone-marrow morphology, with immature white cells and basophilic stippling of red cells. It is noteworthy that spherical homogeneous densities may appear in the lung fields during the course of this disease, as first demonstrated by Hirsch.[15] Such pulmonary lesions tend to be multiple, evanescent, and do not demonstrate vascular continuity on the hilar side.

2. *Congenital heart disease* is a possible mis-diagnosis. In pulmonary arteriovenous aneurysm the cyanosis is frequently of later development; the heart size and contours are normal, and murmurs are usually absent except for a bruit over the lesions themselves. The roentgen evidences of vascular tumors are absent in the congenital cardiac, unless proximal aneurysm or diffuse dilatation of the pulmonary arteries exists. These shadows are then hilar in type.

3. *Tuberculosis.* Pulmonary arterial aneurysms of a local type occur in tuberculous cavities, the so-called Rasmussen aneurysms[16] and very rarely dissecting aneurysms of the parenchyma.[17] These are hardly to be confused in the presence of active tuberculous symptoms and positive sputum.

4. *Benign Pulmonary Tumors.* These are not commonly associated with cyanosis and polycythemic changes, although clubbing may be marked. In this connection Arel and Saka[18] reported multiple mycotic tuberculous thrombosing aneurysms in a case with negative sputum, strongly simulating tumors of a discrete, and benign type.

ROENTGEN AIDS TO DIAGNOSIS

The roentgenographic features essential for the diagnosis of arteriovenous aneurysm are (1) a saccular, cirsoid or racemose cluster of sharply defined structures with homogenous density and (2) demonstration of enlarged tributary vascular channels. These features may be shown in standard radiographs, but usually are better demonstrated by laminagraphy, which also permits differentiation of the arterial and venous trunks, particularly when the lesion is situated in a lower lobe.

Changes in intrathoracic pressure as produced by the Valsalva and Müller tests are helpful in demonstrating the vascular nature of the lesion. More than half of the published articles describing the use of these tests report a positive response; that is, a reduction in size during the Valsalva maneuver. Two of the present series showed this (Figures 1B, 1C), the other two being inconclusive, since the appropriate radiographs were not made. The measured changes, when present, were more pronounced in the tributary channels than in the aneurysm proper. Pulsation during radioscopy is said to be an uncommon finding. No significant pulsations were noted in the present series.

Angiocardiography showed rapid filling of the malformation, usually one or two seconds after Diodrast reaches the right atrium. Prompt clearing of

the opaque medium from the lesion would be expected, but usually the aneurysm remained opacified after the pulmonary arteries had cleared.

Actual demonstration of successive filling of arteries and veins may be of value, and identification of anomalous origin of the tributary artery or insertion of the tributary vein is likely to be of practical interest to the surgeon. Small arteriovenous aneurysms, overlooked in the preliminary roentgenograms, may be intensified by Diodrast and thereby recognized. Since multiple pulmonary aneurysms occur in many cases, it is the radiologist's duty to recognize them, because the presence of more than one lesion may influence the surgeon's decisions on operability and on conservation of lung tissue. These considerations suggest a limited angiocardiographic technic, using large films and a standard stereo chest cassette changer, the two exposures being made at two and four seconds respectively. This would permit a more complete search of the lung fields than is possible with the newly developed rapid film advancer.

TREATMENT

The existence of an arteriovenous pulmonary aneurysm, even in the absence of symptoms, cyanosis or polycythemic changes, is sufficient indication for recommending surgical treatment, unless the patient has some forbidding unrelated pathology. This decision is based on the significant danger of serious or lethal hemoptysis[10] and fatal hemothorax.[2, 19] In the presence of marked cyanosis and polycythemia there are additional hazards of cardiac and cerebral anoxia, and thrombotic complications. One case of subacute bacterial endarteritis has been reported[13] and two cases of complicating brain abscess.[20, 21]

A therapeutic problem is posed by the tendency toward multiplicity in these lesions (*e.g.*, Case 4). In 27 of Yater's collected cases more than one aneurysm was recognized; others may possibly have escaped detection radiologically. If the lesions are multiple in a single lobe (Case 2) or contiguous lobes, the surgical problem is simpler. Bilateral multilobar lesions demand conservative local surgery, such as first practised by Janes.[22]

The surgical method of attack in the recorded cases has varied from simple ligature of the supplying artery or arteries, local dissection, and lobectomy to the extreme of total pneumonectomy. The enormous size of the lesions *"in vivo,"* and the remarkable dilatation of both afferent arteries and efferent veins may lead to a decision for radical surgery where lobectomy might suffice on more prolonged study and dissection.

Conservative or medical management has included repeated venesections, drugs such as phenylhydrazine, and artificial pneumothorax.[23] The latter has been proved to be of no therapeutic value, either in reducing size of the lesion or in ameliorating the polycythemia. It has inherent danger, as the lesions are superficially placed, thin-walled and adherent, because of the possibility of tearing and bleeding.

During operation upon the severely polycythemic cases, transfusion usually is undesirable, and supportive therapy consists of glucose solutions, saline, or plasma. In preparation for operation, as in congenital cyanotic cardiacs, it may be wise to practise phlebotomy, reserving the blood for possible postoperative transfusion. This procedure was used in our second case. Frequent postoperative checks on the erythrocyte count and hematocrit are desirable to control the therapy.

In the dissection of the involved lobe or lobes, caution must be observed to detect anomalous vascular patterns. In our second case, although the aneurysm lay in the lower lobe, an artery to the lateral segment of the middle lobe arose from the lower lobe vascular trunk and traversed the oblique fissure, leading to a decision for bilobectomy. In one reported case[23] there was an absence of the upper lobe vein, all venous return passing through a single posterior inferior vein.

Ligation or transection of the supplying artery without dissection of the aneurysm has been practised, but with doubtful results. On theoretic grounds this is an objectionable procedure, since it should be a stimulus for the development of bronchial collateral circulation, which might prevent the anticipated local thrombosis, producing a bronchial-pulmonary arterial shunt of some magnitude. Where this type of simple ligation has been practised, the lesion persisted by roentgen-ray.[24]

PATHOLOGIC CONSIDERATIONS

Our three lobectomy specimens were prepared by bronchovascular injections of colored vinylite plastic.[7] The three specimens and resultant casts had certain features in common and some minor differences. In all instances the aneurysmal sac approached the visceral pleura closely, and in Case 1 actually elevated this membrane in the fashion of a vesicle. Cases 1 and 3 showed single sacs; in Case 2 two separate aneurysmal loops were present in close relation to one another. In the immediate vicinity of the aneurysms the supplying vessels were larger than at a more proximal level, but even at their proximal origins they were of greater caliber and thinner-walled than normal.

Vessels of more than one bronchopulmonary segment were sometimes involved, and there was no regularity whatever in the number and arrangement of the constituent arteries and veins. In none of the specimens were communications observed proximal to the aneurysmal sac, as reported by Goldman.[25] Communications existed only in the sacs which showed multiple openings. The diameters of supplying arteries at point of entrance into the sacs varied from two to eight millimeters. The draining veins in each case were larger than the corresponding arteries. There was no evidence in the casts of a significant contribution from the bronchial arteries. In Case 3 where these vessels seemed slightly enlarged, they apparently functioned merely as vasa vasorum.

In Case 2 sizable peripheral venous branches draining presumably normal pulmonary parenchyma emptied directly into the aneurysmal sac (Fig. 4A).

During systole, at least, the pressure may have been higher in the proximal (sac) ends of these veins than in the pulmonary capillaries which they drained, thus producing a momentary stoppage or even a reversal of flow.

Evidence supporting the congenital origin of these lesions, or an extremely slow developmental rate, is gained from the observation that the contiguous bronchi and parenchyma were but little disturbed, being intimately interlaced between the vessels and about the aneurysmal sac. The sacs were analogous in their position to capillaries, as Hayward and Reid[26] pointed out. Yet they must be considered as more than dilated capillaries, since each aneurysmal sac was supplied and drained by more than a single artery and vein. Indeed some of the larger sacs were found to be entered directly by sizable peripheral venous branches, draining areas of capillaries which were in turn supplied by arterial branches not involved directly in the aneurysmal lesion.

DISCUSSION

These arteriovenous communications of the lung appear homologous with certain cavernous hemangiomas seen elsewhere in the body. As Reid[27] and Virchow[28] before him pointed out, they represent large channels joining arteries and vein. Albrecht[29] has referred to such cavernous hemangiomas as hamartomas, implying thereby that they represent more an error in development than a neoplasm.

The strikingly high incidence of cutaneous, mucosal and visceral angiomas in the reported cases, the frequent occurrence of telangiectasia or recurrent epistaxis in parents or siblings, the report by two authors of pulmonary arteriovenous aneurysm in brothers,[14, 25] all point toward the hereditary nature of these pulmonary lesions and their identification as a variant of Rendu-Osler-Weber disease (hereditary hemorrhagic telangiectasia).

Requiring explanation is the observation that a well-developed arteriovenous aneurysm may exist in the absence of cyanosis, clubbing, polycythemia, or other signs and symptoms (cf. Case 1). Likewise to be explained are the occasional case records in which cyanosis, clubbing, and polycythemia first appeared late in adolescence or in adult life. Sweet's case[12] of middle lobe arteriovenous neurysm was known to have had a normal hemogram two years before a surgical procedure. At the time of surgery, however, the red cell count had risen to over seven million, and the hemoglobin to 21.5 Gm. These observations pre-suppose either a hemodynamic block in a pre-existing arteriovenous communication or, what is more likely, a progressive increase in the size of the communications and in the degree of right-left shunt of venous blood with the passage of time.

SUMMARY

The clinical and diagnostic features of pulmonary arteriovenous aneurysm have been reviewed from the standpoint of the recorded cases, and from a

personal experience with four additional cases, three of which were treated by lobectomy. The pathologic anatomy of the lesions has been studied and illustrated by using the vinylite-plastic injection technic.

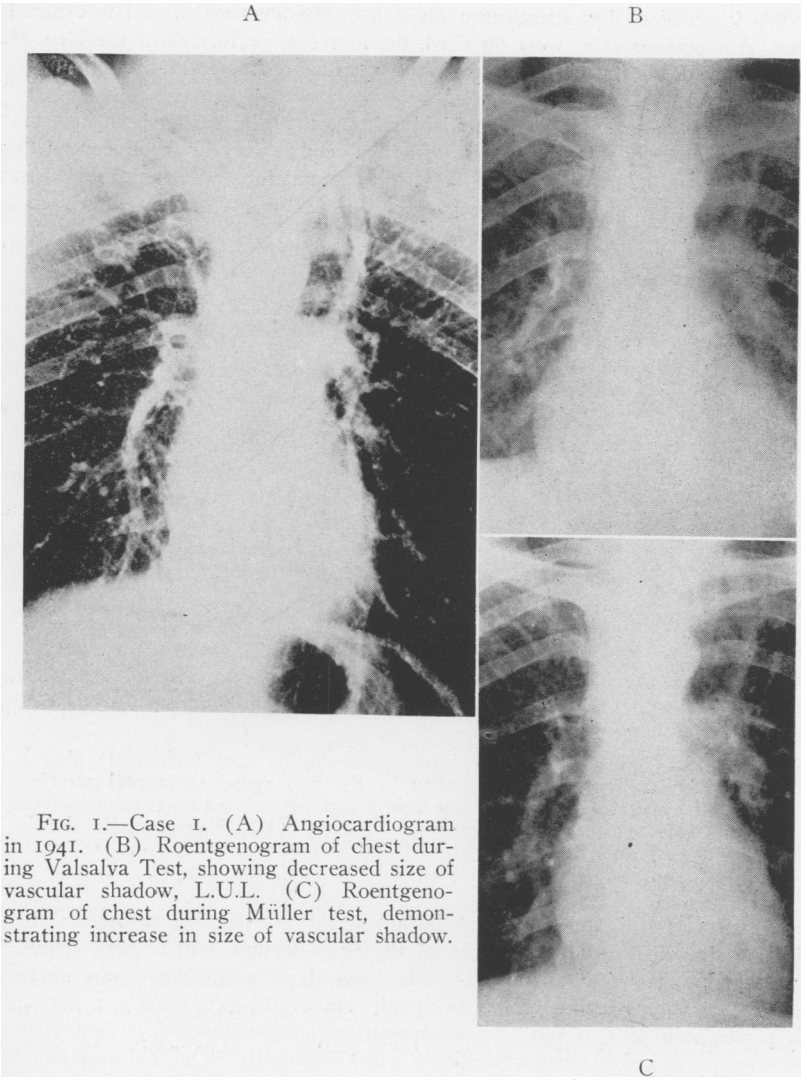

FIG. 1.—Case 1. (A) Angiocardiogram in 1941. (B) Roentgenogram of chest during Valsalva Test, showing decreased size of vascular shadow, L.U.L. (C) Roentgenogram of chest during Müller test, demonstrating increase in size of vascular shadow.

CASE REPORTS

Case 1.—(N. H. H., B20727) A. B., a 24-year-old white machinist was referred to the out-patient service on October 4, 1941, with a presumptive diagnosis of minimal pulmonary tuberculosis involving the left upper lobe. The lesion had been uncovered 3 weeks previously during routine roentgen ray examination for Army service. The patient was asymptomatic. He denied any pulmonary disease or tuberculosis contact. System review revealed frequent bouts of epistaxis as a child, occurring with lessened frequency as he grew older. Family history was not remarkable.

Upon physical examination his temperature was 99, pulse 64, respiratory rate 24, blood pressure 118/70, and weight 140 lbs. No positive findings were noted in the chest; there was an incidental finding of hypospadias. A tuberculin test was negative at 1.0 mg. O.T. Chest films indicated enlargement of the pulmonary vascular shadow in the left hilum extending as a tapering shadow into the apex. Laminagraphic studies confirmed this impression, demonstrating a division of the abnormal vessel into two large branches. Angiocardiograms were obtained, the first ever performed at the New Haven Hospital. They demonstrated conclusively the vascular character of the left upper lobe lesion (Fig. 1A).

The patient was not seen again until March 14, 1949, he having been inducted into the armed forces, serving for four years of active combat duty in the European theater. During this period he remained well. At this examination, an arterial blood sample revealed an oxygen content of 18.8 vols. per 100 cc., or 95 per cent saturation. The

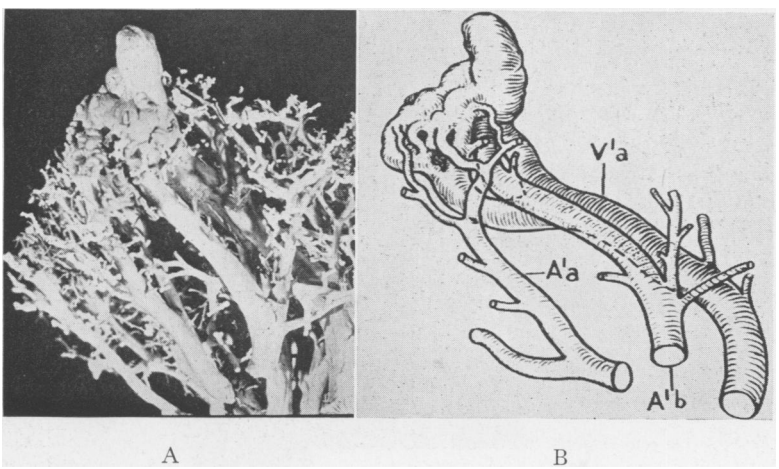

FIG. 2.—Case 1. (A) Bronchovascular vinylite cast (trimmed) photograph. The aneurysmal sac, supplied by the two apical segmental arteries (A_1a, A_1b) and drained by a single vein (V_1a), possessed an extremely thin wall and elevated the visceral pleura as a bleb. (B) Diagrammatic sketch of same.

family history was reviewed again, and did not reveal any bleeding tendencies. Physical examination showed a marked deviation of the nasal septum with a large spur. There were no telangiectases of the skin or visible mucosal surfaces. There was no cyanosis or clubbing; no pulmonary or cardiac murmurs. The erythrocyte count was 4.36 million, with a hemoglobin of 15.0 Gm. An electrocardiogram was normal. Chest roentgenograms were unchanged as compared with 8 years previously. The Valsalva and Müller signs were both positive (Figs. 1B and 1C). A diagnostic pneumothorax was performed, showing a separation of the upper lobe and its contained lesion from the parietal pleura.

On May 17, 1949, the patient was explored through the fifth intercostal space. A large pulsatile vascular tumefaction of the upper lobe was discovered, and a lobectomy carried out without drainage. Postoperatively the patient had a slight elevation of temperature for 3 days, and required 2 thoracenteses. On the fourth day after operation, nose blowing brought on a moderate epistaxis, which was controlled by packing. The patient was discharged from the hospital on the tenth day. When last seen 4 months after operation he was well and had gained 9 lbs. in weight.

Surgical Pathology No. 47360 (Figs. 2A, 2B). Beneath the pleura at the very apex of the lung was a thin-walled vesicle resembling an emphysematous bleb, but filled with blood. Examination of the lobar hilum showed both apical* arteries and vein to be greatly enlarged and thin-walled. In the cast made with vinylite plastic, the overall dimensions of the aneurysmal sac were 3 by 2½ by 2 cm. It received at least 3 small arterial branches from A_1a, a larger branch from A_1b, this having a diameter of 2 mm. at its point of entrance. Venous drainage was entirely into V1 by one large and several small channels. Delicate bronchial branches passed through the meshes of the interlacing arteries and veins, producing depressions on the surface of the aneurysmal sac, but the bronchi were themselves only slightly distorted.

Case 2.—(N. H. H. C35665) S. E. K., a 56-year-old housewife, was first admitted on December 20, 1949, with a life-long history of cyanosis and dyspnea. Three years previously she had a single episode of moderately severe hemoptysis. Chronic cough,

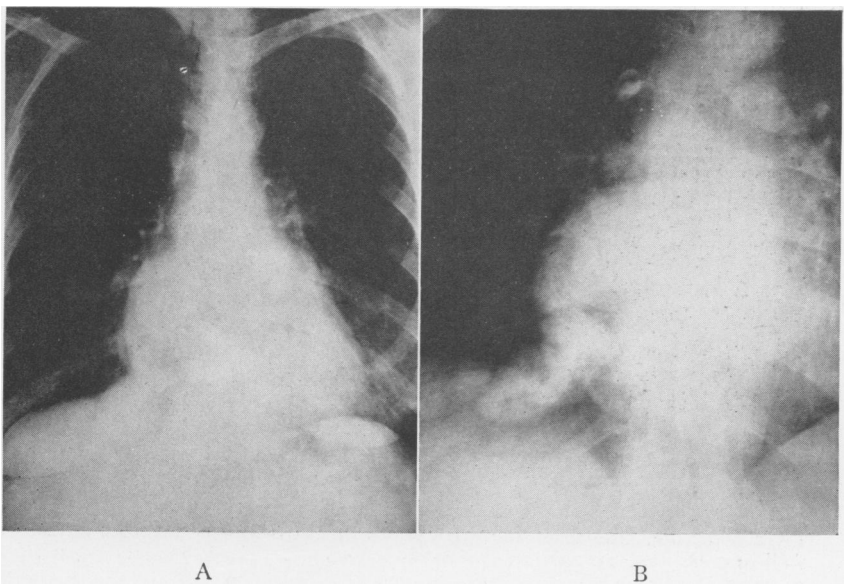

FIG. 3.—Case 2. (A) P. A. Roentgenogram. (B) Angiocardiogram.

sputum, and chest pain were denied. After contracting rheumatic fever at age 18, the patient had valvular disease and was known to have fibrillated for many years. Two and one-half years before this admission, the patient was treated conservatively at another hospital because of a saddle embolus. This left the patient with persistent severe pain and burning sensations in the right foot, with claudication of the right leg. The family history was not available in detail.

On physical examination the patient appeared somewhat obese, with obvious cyanosis of the lips, tongue, buccal mucosa, and nailbeds. Moderate clubbing of the fingers and toes was present. Her temperature was 98.6, pulse (apical) 80, respiratory rate 24, and blood pressure 130/80. The chest revealed no abnormal breath sounds or bruit. The heart was enlarged to the right with an apical systolic and diastolic murmur. The cardiac rhythm was irregular. The liver was palpable two finger-breadths below the costal

* For this and the following descriptions both the Jackson-Huber nomenclature and the Boyden numerical designations will be used.

margin. No femoral pulses were detectable bilaterally although there was a faint dorsal pedis pulsation in the left foot. The right leg and foot were cool, slightly edematous, and there was a rapid onset of pallor with elevation. Neurologic examination was negative. Roentgenograms of the chest (Fig. 3A) revealed left auricular and ventricular enlargement and several large lobulated densities in the posterior basilar portion of the right lower lobe with a bandlike density extending to the hilum. An angiocardiogram verified the vascular nature of the lesion (Fig. 3B).

The erythrocyte count was 6.29 million, the hemoglobin 18.8 Gm., the white count 6,750 with a normal smear and differential. The hematocrit was 53 mm. An arterial blood specimen contained 17.87 vols. per 100 of oxygen, or 78 per cent saturation.

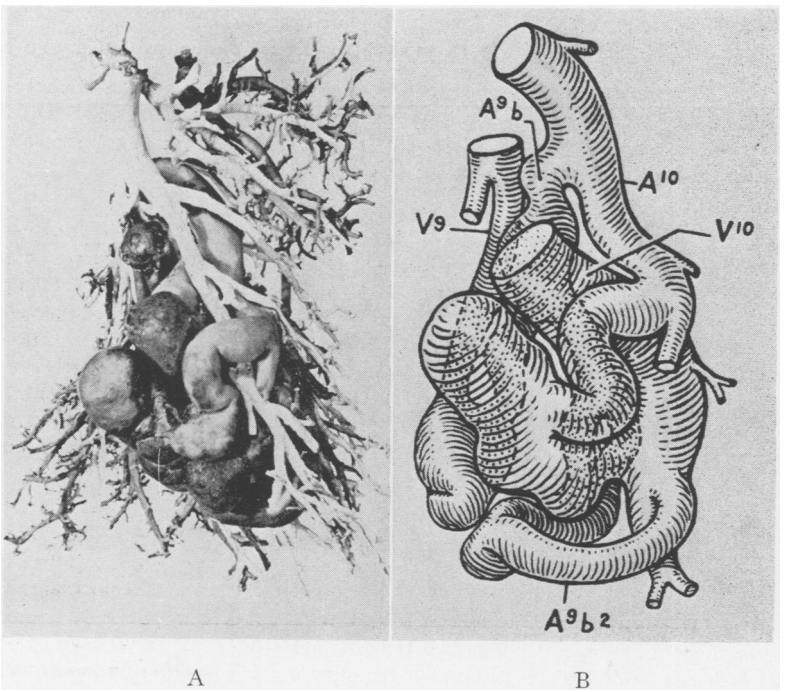

FIG. 4.—Case 2. (A) Postero-medial aspect of trimmed right lower lobe cast. Two separate vascular loops are shown. Situated beneath artery (A_9b_2) is a peripheral venous tributary, draining into the aneurysmal sac from capillaries of a parenchymal area which has a normal pulmonary arterial supply. (B) Diagrammatic sketch of same.

After breathing 100 per cent oxygen the content was 19.29 vols. per 100. On the night before operation, 250 cc. blood was removed by venipuncture, citrated, and administered during operation. On January 3, 1950, after resection of the right seventh rib, a large tense aneurysmal sac was found in the lower lobe. There was a distinct systolic thrill to palpation of the lesion. Because of an anomalous arterial branch passing from the lower lobe transfissurally to the middle lobe, a bilobectomy was performed. The main lower lobe vein measured about 3.5 cm. in diameter. Since ligation of this large vessel did not appear quite safe, it was followed distally, and two smaller but still very large branches ligated and transfixed. The pleural cavity was drained with a catheter, which was removed on the fourth postoperative day. The postoperative course was smooth and almost afebrile. The patient left the hospital on the fifteenth day after operation.

When seen on February 10, 1950, the patient had no cyanosis, and there was improvement in her exercise tolerance. The erythrocyte count was 3.54 million, and the hemoglobin 10.5 Gm. There was little if any subjective improvement in the claudication and foot pain.

Surgical Pathology. No. 49264 (Figs. 4A, 4B). The lesion involved the vessels of the lateral (B9) and posterior basal (B10) segments. The dimensions of the lesion were 9½ by 4½ by 3½ cm. Two major arteriovenous loops existed. The smaller aneurysmal sac occurred between a branch of A_9b and V_9. The larger sac was more complex and received 3 large arterial channels. Two of these were branches of A_9b. The third and largest, a branch of $A_{10}b$ measured 8 mm. in diameter at its entrance into the sac. Venous drainage was entirely into V_{10}, which measured 15 mm. at the point where it left the aneurysmal sac. A larger sac on the postero-medial aspect of the lobe had a diameter approximately one and a half times greater. Small peripheral branches, venous in type, entered the major sac in its most prominent aspect. These venous branches drained capillaries of the parenchyma supplied by arterial branches not otherwise involved in the lesion. The segmental bronchi ramified through the meshes of the vascular plexus and showed no ectasia or other significant change, although they curved somewhat around the vascular dilatations.

Case 3.—(N. H. H. C32336) H. N. C., a 54-year-old white housewife was first seen in the out-patient clinic on October 13, 1949, because of dizziness, flushing, sweating, and nervousness. These symptoms had been present for about 6 years, of increasing frequency and severity for 2 years. There had been no episodes of fainting, or convulsions, but occasional throbbing headaches were noted,and recent exertional dyspnea. There had been a steady weight loss of 20 lbs. in 2 years. The menopause occurred 6 years previously. For 15 years the patient was known to be hypertensive. Details of the family history were only vaguely remembered, and not considered reliable. The patient had had a radical mastectomy in a private clinic 15 months previously with a diagnosis of carcinoma, and no mention had been made of pathology in the chest at that time.

The physical examination revealed a small but well-developed and nourished female who did not appear ill, except for mild cyanosis of the lips and nailbeds. The pulse was 72, the temperature 99, respiratory rate 18, and blood pressure 250/130, after rest in bed 180/110. The retinal vessels were tortuous with no scarring or hemorrhages of the fundi. A left mastectomy scar was well healed with no evidence of local recurrence. There was slight dullness and diminished breath sounds over the right lower lobe posteriorly. A harsh bruit was heard in the auscultatory triangle synchronous with the peripheral pulse. The heart was enlarged to the left but without murmurs. There was minimal clubbing of the fingers. Neurologic examination was negative.

Roentgenograms of the chest (Fig. 5) revealed transverse enlargement of the cardiac shadow, moderate pulmonary emphysema, and a long lobulated density in the basilar portion of the right lower lobe, thought to represent an arteriovenous aneurysm. Angiograms were technically unsuccessful. Laboratory data were as follows: R.B.C. 6.55 million, Hgb. 15.9 Gm., Hematocrit 45, W.B.C. 5500. Urine was negative for albumin and sugar; the sediment showed occasional red and white cells but no casts. The N.P.N. was 30, total serum protein 6.6 Gm., chlorides 96.9 mEq. and CO_2 26.8. The arterial oxygen saturation at rest was 73.2 per cent and after breathing pure oxygen 94 per cent.

An electrocardiogram revealed slight left axis shift with abnormally slurred limb leads and an abnormally depressed RS-T segment in lead V-4. The T-waves were inverted in lead V-4 and diphasic in leads I and V-6. The venous circulation time (arm to tongue) was 22 seconds. The blood serology was negative.

On January 12, 1950, the patient was explored after resection of the right seventh rib. A tense pulsating tumor of the right lower lobe was readily palpated but not easily seen. A resection of the right lower lobe and a very small middle lobe was performed,

the latter necessitated by an air leak in its proximal bronchus. The pleural cavity was drained; the postoperative temperature ranged between 100 and 101 F. for 3 days, and there was left-sided chest pain for a similar period. The patient was discharged on the thirteenth day. On this date the arterial oxygen saturation was 96.8 per cent at rest, and 100 per cent after re-breathing oxygen. A postoperative red cell count was 4.4 million with a hemoglobin of 12 Gm., and a white count of 7300. The bruit previously heard on the posterior chest wall was no longer present.

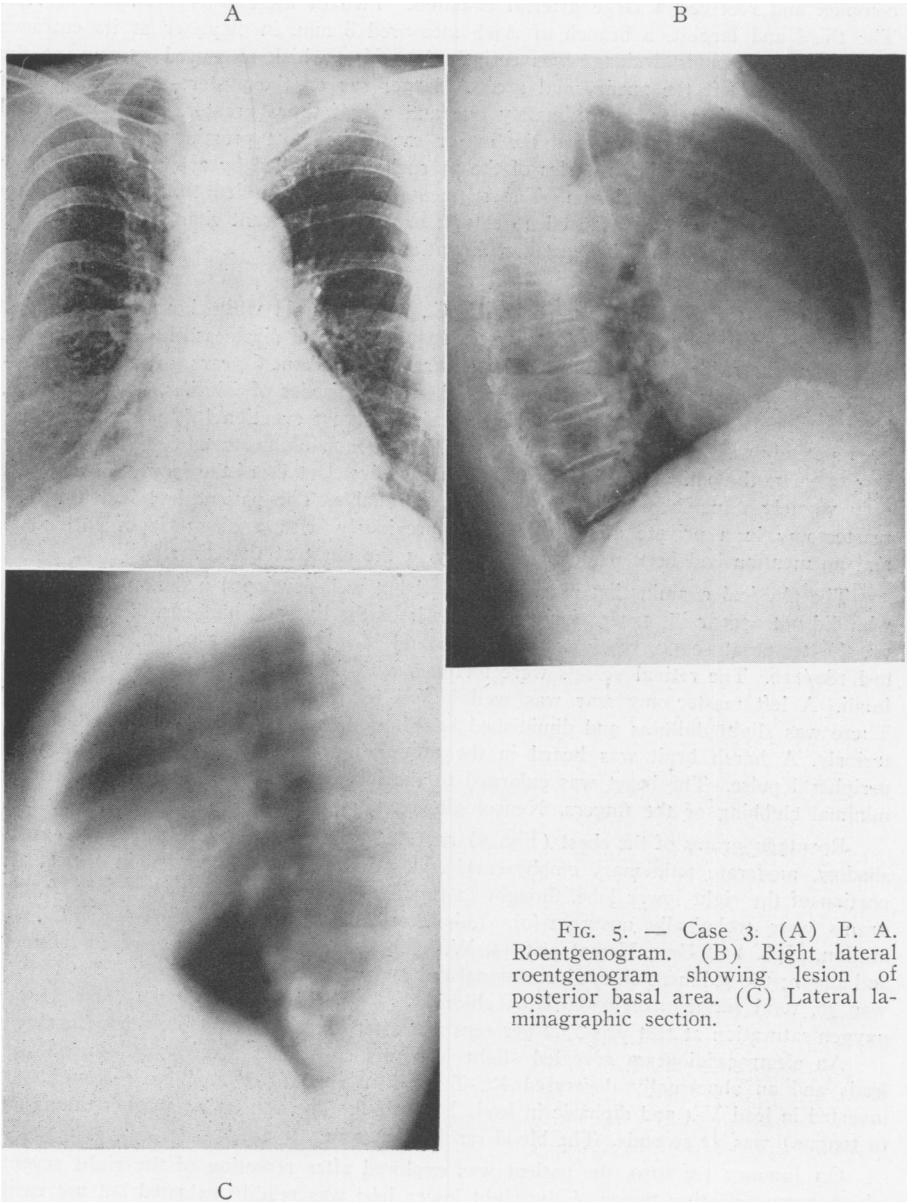

FIG. 5. — Case 3. (A) P. A. Roentgenogram. (B) Right lateral roentgenogram showing lesion of posterior basal area. (C) Lateral laminagraphic section.

Surgical Pathology No. 49333 (Fig. 6). A single aneurysmal sac measuring 3.5 by 2.5 by 2.5 cm. occupied the lateral basal segment. It was supplied by one large and several minute branches of A_9b, and was drained by two large and several small tributaries of V_9 and a small branch from V_{10}. Both A_9 and V_9 were greatly enlarged, the former measuring 8 mm. at the point of entrance into the aneurysmal sac, which was close to the diaphragmatic pleura. The associated vessels had a plexiform angiomatous arrange-

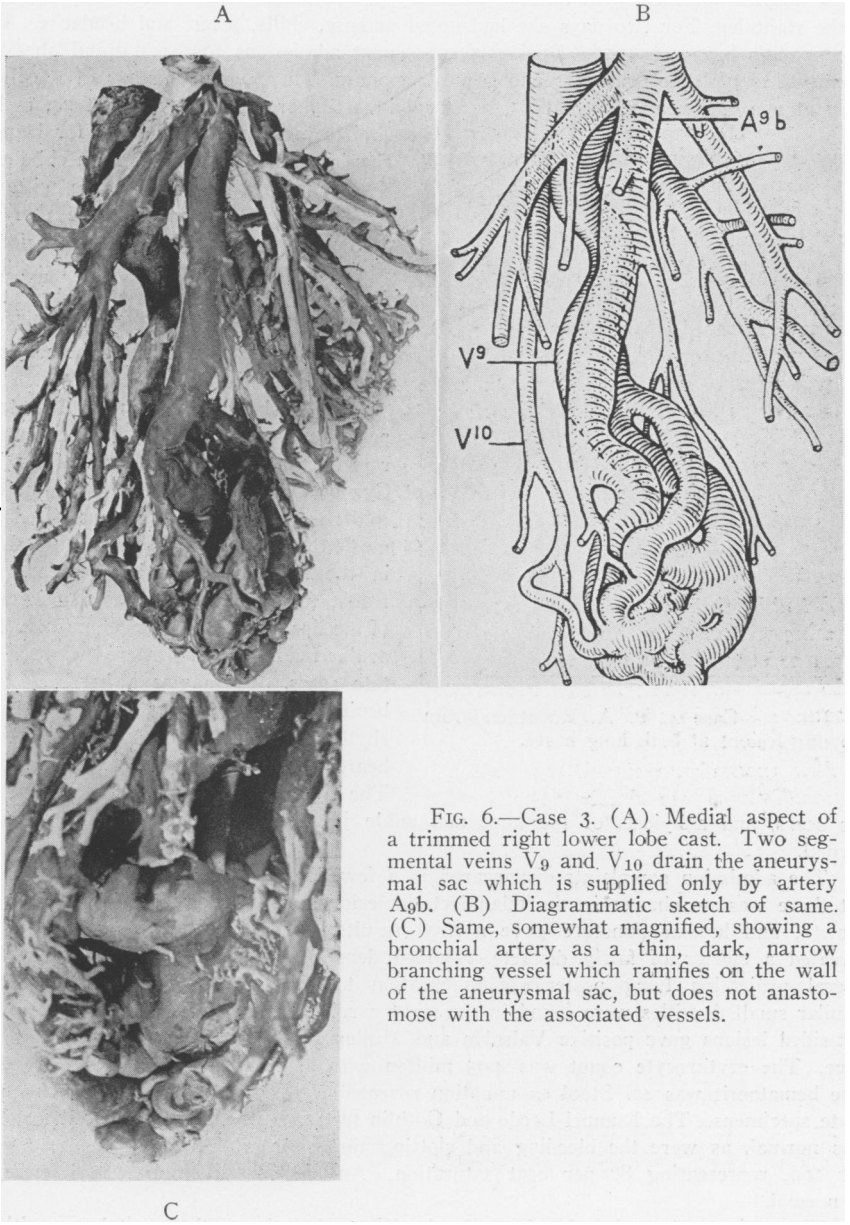

FIG. 6.—Case 3. (A) Medial aspect of a trimmed right lower lobe cast. Two segmental veins V_9 and V_{10} drain the aneurysmal sac which is supplied only by artery A_9b. (B) Diagrammatic sketch of same. (C) Same, somewhat magnified, showing a bronchial artery as a thin, dark, narrow branching vessel which ramifies on the wall of the aneurysmal sac, but does not anastomose with the associated vessels.

ment between the interstices of which the segmental bronchial branches penetrated. The bronchi were not abnormal in any way. A moderately enlarged bronchial artery followed the course of B_9 and was distributed upon the walls of the dilated vascular channels in the manner of a vas vasis, but without macroscopic communication with the arteriovenous complex (Fig. 6C).

Case 4.—(N. H. H. No. 48908) D. K., a 55-year-old white housewife was admitted to the hospital for the fourth time on February 5, 1949, because of chronic osteomyelitis of the right leg. For two days she had noted malaise, chills, fever, and headache. The osteomyelitis began in 1926. During 3 subsequent admissions to the hospital she was submitted to multiple drainages and sequestrectomies. There was a history of tonsillectomy at age 6, appendectomy at 15, right salpingo-oophorectomy for cyst at 24, and a similar procedure on the left for hemorrhagic cyst at 29. Dating from childhood the patient had frequent profuse epistaxes which seemed to be more severe after the artificial menopause. The epistaxes necessitated transfusions in 1924 and 1944. The family history revealed that two daughters had episodes of epistaxis from childhood and showed telangiectases of the conjunctivae, lips, and hands.

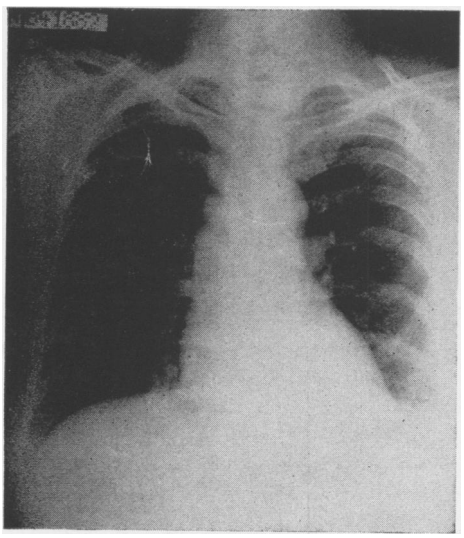

FIG. 7.—Case 4. P. A. Roentgenogram showing lesions at both lung bases.

Physical examination revealed a temperature of 99.2, pulse of 96, respiratory rate of 24, and blood pressure of 110/70. Cyanosis was not observed. Numerous spiderlike petechial areas were noted beneath the fingernails, in the conjunctivae, in the skin of the wrists, ankles, and trunk, together with small telangiectases of the lips, tongue and buccal mucosa. The ocular fundi showed scattered white punctate scars. There was a harsh systolic bruit at the left scapular angle. The heart rhythm was regular and no murmurs were heard. The fingers and toes were clubbed. The right leg was shortened with numerous scars, and ankylosis of the knee and ankle joints. Neurologic examination was normal.

The admission complaints disappeared in a few days and the skin petechiae faded, but there was no change in the telangiectatic lesions. Five blood cultures showed no growth, and lumbar puncture gave normal results. Chest roentgenograms (Fig. 7) revealed a cluster of fairly discrete, rounded densities noted particularly in the left lateral projection lying posteriorly in relation to the enlarged vascular markings. Similar small densities were found in the right cardiohepatic angle. On fluoroscopy the left-sided lesions gave positive Valsalva and Müller signs. Angiocardiography was not done. The erythrocyte count was 4.05 million with a hemoglobin value of 10.0 Gm. The hematocrit was 40. Stool examination revealed a positive guaiac test on two separate specimens. The Rumpel-Leede and Göthlin tests were negative. Red cell fragility was normal, as were the bleeding and clotting times. Arterial oxygen was 11.9 vols. per 100, representing 87 per cent saturation. An electrocardiogram was interpreted as normal.

The patient returned to her home in the West after the ninth hospital day without surgical treatment.

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DISCUSSION.—DR. WILLIAM E. ADAMS: My remarks will be confined chiefly to the presentation by Dr. Lindskog, who I think gave us an excellent report on this problem which we are seeing more frequently in recent years.

Although we have had experience with three of these patients, only one of the three had a large enough shunt to give rise to the characteristic blood picture changes and other physiologic findings. The one case was operated on first in 1943, at which