Arteriovenous Fistula of the Lung and an Associated Hemangioma of the Vertebra: Case Report

JAMES W. NIXON, M.D., F.C.C.P. and JOHN F. PERRY, M.D.
San Antonio, Texas

The subject of arteriovenous fistulae is of interest not only because such lesions are of unusual occurrence, but also because these cases are amenable to surgical treatment, with satisfactory results being expected therefrom. It is likewise interesting that the description and the diagnosis of this disease have been recorded only since modern thoracic surgery has been able to offer relief.

In the earliest reports, Reading in 1932,11 and later Rodes in 1938,12 presented each a case. Both of these cases had shown manifestations of polycythemia, clubbing, and cyanosis though neither case had been diagnosed during life. At autopsy both cases had been shown to have a pulmonary hemangioma or arteriovenous fistula. The literature of the decade following the latter report is found to contain more numerous examples of the disease, manifested by the same group of symptoms. A most recent and most excellent review of the literature is given by Yater, et al.,13 in which 45 cases are presented.

Pulmonary arteriovenous fistula is a disease belonging to the class of hereditary hemorrhagic telangiectasia and it is possible to demonstrate cutaneous and mucous membrane lesions in 50 per cent of those afflicted.2

The etiology is not known beyond the fact that the lesion is congenital and that the tendency is apparently familial, since pulmonary lesions, as well as generalized telangiectasia have been found in siblings and parents of patients with pulmonary arteriovenous fistulae. The tendency is apparently transmitted as a simple dominant characteristic, with both sexes being affected.

The onset of symptoms may occur at ages ranging from birth to the third decade. The congenital character of the disease and the late onset of symptoms suggest that the lesions are progressive. Once the vascular channels of pulmonary hemangioma have become enlarged to the point of functioning as an arteriovenous fistula in the lesser circulation, a characteristic symptom complex appears: The patient, usually in the third or fourth decade of life, becomes cyanotic although the heart is normal in both size and
function. Dyspnea, hemoptysis, transient dizziness and cerebral manifestations of numbness and paresthesias may occur. There is clubbing of the digits, and the presence of telangiectasia may be found throughout the skin and mucous membranes. Both the blood-pressure and the pulse remain normal and no enlargement of the spleen occurs. Over the lung fields there may be an audible bruit.

Laboratory examinations reveal the presence of polycythemia, with increased red blood cells, hemoglobin, and hematocrit. The white blood count remains normal. Arterial oxygen saturation is reduced. This reduction, it has been found, is inversely proportional to the degree of polycythemia present, which condition in turn reflects the degree of arteriovenous shunting that is occurring in the pulmonary arteriovenous fistula.9

X-ray examination of the chest demonstrates one or more circumscribed lobulated densities in the lung field which may pulsate fluoroscopically. Laminograms and angiography may demonstrate tortuous vessels leading from the hilus to the area of increased density. The heart shadow is normal in size. Coupled with the clinical picture the x-ray findings may be considered practically diagnostic.

The prognosis in untreated cases and in cases unsuitable for radical treatment is poor. The major threats of thrombosis resulting from increasing density of the blood, or of pulmonary hemorrhage by rupture of the lesion into a bronchus, are the chief indications for surgery; but symptoms secondary to polycythemia must be included in the criteria.

In the differential diagnosis of this disease, the following possible conditions must be considered: (1) polycythemia vera, (2) secondary polycythemia, which may be due to (a) high altitude, (b) poisoning from heavy metals or amaline dyes, (c) pulmonary diseases such as Ayerza's Disease or emphysema, both of which prevent adequate oxygenation; and (d) cardiac anomalies with a right-to-left shunt.9

In the roentgen diagnosis of the lesion, cysts, adenomas, metastatic lesions, tuberculoma, and aneurysm of pulmonary artery branches must be ruled out. Pulmonary densities must not be confused with those that are secondary to infarcts in cases of polycythemia vera.

A peripheral arteriovenous shunt increases all elements of blood volume proportionally and likewise increases cardiac output with an accompanying dilatation of the heart. A pulmonary shunt has little effect on either cardiac output or cardiac dynamics.8 The increase in red cell mass is secondary to decreased oxygenation
FIGURE 1: A-P projection of chest showing anterior-venous aneurysm in left lower lung field. The enlarged pulmonary hilar vessels are evident.

FIGURE 2: Lateral view of cervical spine showing herniation of third cervical vertebra.

FIGURE 3: X-ray film of barium filled surgical specimen showing extent of arterio-venous aneurysm.
of the blood in the pulmonary circuit. The blood-pressure remains normal, the heart does not enlarge.

Moyer and Ackerman, in reviewing the pathology of several pulmonary arteriovenous fistulae, found these lesions to consist of distended afferent arteries and distended efferent veins. The intercommunicating capillaries which normally function between the arteries and the veins were absent and in their place was found either large vascular trunks furnishing direct communication or else a mass of distended vessels. Degenerative changes had occurred in the vessel walls due to arterial pressures having been transmitted directly to the abnormal vessels and veins.

The treatment of pulmonary arteriovenous fistulae is surgical. Hepburn and Dauphine in 1942 reported the first cure by pneumonectomy. Yater, et al., series includes 26 operated cases, two of which died post-operatively. The choice of pneumonectomy, single or multiple lobectomy, or segmental resection will depend upon the number, extent, and distribution of the lesions.

The following case report is presented because the relative infrequency of this lesion warrants description of individual cases and because we believe that this specific example presents certain features which have not hitherto been described.

**Case Report**

M. H., a 26 year old Latin-American male was admitted to the Santa Rosa Clinical Service November 8, 1949, with a chief complaint of pain in the neck and dyspnea on exertion. The neck pain had developed seven months prior to his admission, in the right posterior auricular region, and had increased in severity somewhat with radiation to the right posterior triangle of the neck. The pain was severe enough that he had stopped working at its onset. There was no history of trauma. The second complaint of dyspnea, coupled with easy fatigability, had been present since age eight. At this time a school physician had noted that the boy's fingernails were blue and enlarged. A diagnosis of "heart trouble" had been made at this time and he was advised to refrain from strenuous activity. He completed the sixth grade in school and had since worked in a cleaning and pressing shop until onset of his neck pain in April 1949. He had been known in his community as the "Blue Boy." Past history was negative except for one attack of pneumonia during childhood. Family history was significant in that a paternal aunt had had a similar affliction.

*Physical Examination* demonstrated a tall, thin male of about stated age, cyanotic, with no respiratory distress. Cyanosis was particularly noted in the nail beds, lips, tongue, and face. There was marked tenderness in the right post auricular area and tenderness of the musculature of the neck below the right mastoid. Discrete, shotty, lymph nodes were palpable bilaterally, with one larger node on the right. There was increased pain on turning the head, referred to the right post auricular area. The teeth were carious with marked pyorrhea. The tongue was dark red in color. A deformity of left third, fourth, and fifth ribs consisting
of slight elevation was present. The heart was not enlarged and heart sounds were normal. Expansion was equal bilaterally and the breath sounds were normal. Over the left posterior chest there was heard a bruit, unrelated to the cardiac sounds. The remainder of the physical examination was negative except for cyanosis and marked clubbing of the digits. Blood-pressure was normal.

**Laboratory Examinations:** RBC 9,280,000; HBG 137 per cent, 21 gms.; WBC 7,500 with normal differential; MCV 72 cu. microns; MCH 23 micrograms; MCHC 31 per cent; Hematocrit 66 per cent. Circulation Time (3 cc. Decholin) 34 Sec. Vital Capacity 62 per cent of normal or 2.9 litres. Urine negative. Stool negative for occult blood. Serum acid phosphatase 5 units.

X-ray examination of the chest demonstrated a smooth, rounded, soft tissue mass six cm. in diameter in the left lower lobe with many markedly dilated vessels extending from this mass to the hilus. No definite pulsation of the mass was seen but there was some pulsation of the vessels extending from the hilus. The heart was slightly enlarged, predominantly left ventricular. A roentgen diagnosis of pulmonary arteriovenous aneurysm was made. X-ray films of the cervical spine demonstrated loss of normal lordosis and demineralization with irregular mottled osteoporosis of C3 suspicious of neoplastic destruction.

Electrocardiograms were reported basically normal with a single nodal extra systole.

In view of the elevated acid phosphatase, and of the evidence of destruction of the third cervical vertebra, diagnosis of neoplastic destruction of third cervical vertebra and the presence of an arteriovenous fistula of the lung was made.

The patient received ten x-ray treatments of 165 R each to the cervical spine with marked improvement. He was discharged November 26, 1949, to be followed in the cardiac clinic in order that the course of the cervical

**Figure 4:** Hand showing marked pulmonary osteoarthropathy.

**Figure 5:** Surgical specimen: The arteriovenous fistula is easily seen.
lesion might be determined following the x-ray therapy. He was re-admitted on January 5, 1950, with interval note that he had gained six pounds weight and that the pain in his neck was much improved. Repeat examination of cervical spine revealed no essential change. I. V. Pyelograms and x-ray of remainder of spine was negative. It was felt that since no primary site for neoplasm had been demonstrated, and that since the lesion of C 7 was apparently stable, that this probably presented a hemangioma of the vertebrae. Accordingly, the patient was seen by the senior author in consultation, and exploration of the left chest was decided upon. On January 19, 1950, operation was scheduled but was abandoned because bleeding from the gums was so marked following intubation for anesthesia that it was felt any surgery would be precarious.

After removal of several loose teeth, the patient was successfully anesthetized on January 28, 1950. The left chest was opened by subperiosteal resection of the sixth rib. Upon entering the chest the left lung was found to be densely adherent to the parietal pleura over a greater part of its surface. After freeing the lung it was found that the left pulmonary artery and left inferior pulmonary vein were dilated to three or four times normal size. A dark red globular mass occupied the greater part of the lower left lung. Over this mass a thrill was palpable which disappeared when the pulmonary artery was clamped, then reappeared with release of the artery. A total pneumonectomy was carried out with individual ligation of the great vessels with heavy braided silk and by closure of the bronchus with medium cotton. The hilus was replerualized, and the chest was closed without drainage. Within a few seconds, following ligation of the pulmonary artery, the patient lost his cyanotic hue, became pink and has remained so.

During his post-operative course he was visited by many of his friends who came to see if the rumor were really true that he was normally pink and no longer the "Blue Boy." His post-operative course was satisfactory, without incident, and he was discharged on February 10, 1950. He was re-admitted March 4, 1950 with interval history that following his discharge from the hospital he had developed a cough which was particularly worse during eating and as a result of which he at poorly, lost weight and became malnourished. Fluoscopy with barium swallow failed to reveal any pathology. He was re-admitted because of cough and progressive weakness from malnutrition. On the afternoon of admission he developed 100.4 degrees F. temperature. The right chest was clear at this time, but by the following day the right chest was full of rales, with temperature of 102 degrees F., pulse 100 to 140, and with x-ray film revealing mottled congestion of right lung field. Cyanosis and dyspnea again developed. On this occasion the diagnosis of broncho-pneumonia was made. The patient responded to digitalls, antibiotics, and oxygen, and was discharged on April 8, 1950, recovered. His subsequent progress has been satisfactory.

Pathological Report: This lung weighs 450 gms. which is within the normal weight range. Externally the lung is pink in color and even in contour. Over the lateral surface of the superior lobe there are a few moderately dense adhesions. Otherwise the outer surface of the lung is smooth. On the medial aspect over the central portion of the lower lobe there is a swelling approximately 5 cm. in diameter and raised approximately 3 cm. above the surrounding lung. This represents the area over the aneurysm.
On cut section through the junction of the inferior pulmonary artery and vein a direct communication between the artery and vein is demonstrated. This was demonstrated before cutting into the lung by injection of a paraffin solution into the artery. The artery and vein each are approximately 3.0 cm. in diameter and the communication between the artery and vein is approximately 2.0 cm. in diameter. Diagnosis: Arteriovenous aneurysm between the inferior pulmonary artery and the inferior pulmonary vein.

Discussion

The explanation of the extreme degree of cyanosis which exists in these cases is that the resistance offered by the arteriovenous aneurysm is less than that offered by the capillaries of the normal lung. This results in a greater blood flow through the arteriovenous aneurysm than through the normal pulmonary capillary bed, as a result of which a larger volume of blood from the right ventricle passes through the pulmonary circuit and into the systemic circulation without being oxygenated.

Hemangioma of vertebra is an uncommon clinical finding although these lesions have been demonstrated in 10 per cent of routine autopsies by Schmorl (quoted by Ghormley and Adson\(^5\)). These are usually asymptomatic and not demonstrable roentgenologically. A classification of these lesions by Foster and Heublein\(^4\) is given below:

1) Asymptomatic discovered incidentally.
2) Symptomatic with localized radicular or spinal pain without objective signs of damage to nerve roots or spinal cord.
3) Hemangioma with subjective and objective signs or myelopathy.
4) Hemangioma which has undergone pathologic fracture and loss of normal contour of vertebral body with or without myelopathy.

Women are affected more often than men.\(^1\) The commonest site for symptomatic lesions is the middle dorsal vertebrae. Bucy and Capp\(^1\) reported no cases of cervical vertebral involvement being found.

Pathologically the intratrabeular areas are filled with dilated cavernous spaces instead of normal myelogenous tissue. These spaces are lined with a single layer of endothelial cells which rest on a thin network of connective tissue. Some trabeculae are seen lined with osteoblasts in process of new bone formation. Elsewhere erosion of bony spicules is evident.

The x-ray appearance of these lesions is characteristic.\(^7,3,1,5\) The irregular absorption of bony trabeculae of the vertebral body and the thickening of the remaining trabeculae cause the appearance of coarse vertical and parallel striation in a field of decreased
density. The abnormal trabeculations may extend into the vertebral arches and laminae.

In the differential diagnosis of vertebral hemangioma, metastatic lesions of vertebral bodies are considered. In the presence of involvement of the spinal cord, intraspinal tumors are to be differentiated. Usually metastases produce symptoms more rapidly progressive than those produced by hemangioma. The primary malignant site can usually be demonstrated.

Most authors agree4,7,3,1 that if the diagnosis can be made with any degree of certainty, deep x-ray therapy is the choice of treatment. Regression of symptoms occurs following roentgen therapy, but the appearance of the vertebral body does not change.

SUMMARY

In reviewing the cases of pulmonary arteriovenous fistulae that have been reported we have been unable to find a record of any such combination of lesions as that here described. Apparently the vertebral hemangioma represents but another manifestation of a generalized telangiectatic tendency in this patient. The response of the lesion to x-ray therapy, without demonstrable change in the appearance of the vertebral body, and the absence of any primary site from which a vertebral metastasis might arise, support the diagnosis of cervical vertebral hemangioma.

RESUMEN

Revisando la literatura sobre fistulas pulmonares arteriovenosas no hemos encontrado referido caso alguno de una combinación como la descrita aquí.

Aparentemente el hemangioma vertebral representa otra manifestación de la tendencia telangiectásica en este enfermo.

La respuesta de la lesión a la terapia sin cambio demostrable en la apariencia del cuerpo vertebral y la ausencia de cualquiera ubicación primaria de la cual pudiera haber emergido la metастasis vertebral, sostiene el diagnóstico de hemangioma de una vértebra cervical.

RESUME

En passant en revue les cas d’anévrismes artério-veineux, les auteurs n’ont pu retrouvé l’association qu’ils décrivent ici. Apparemment, l’hémangiome vertébral ne représente qu’une autre manifestation de la tendance généralisée à la télangiectasie. La réponse favorable à la radiothérapie sans modification de l’aspect de la vertèbre, et l’absence de foyer primitif susceptible d’être à l’origine d’une métastase vertébrale, justifient le diagnostic d’hématangiome vertébral.
REFERENCES


