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COR PULMONALE IN ASSOCIATION WITH RAYNAUD'S DISEASE REPORT OF A CASE WITH SYMPATHECTOMY*

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Raynaud's disease has generally been defined as an intermittent spasm of the digital arteries brought on usually by exposure to cold and by emotional stress. However, it is known that the disease is not always limited to the digital arteries; since cases have been reported in which intermittent blindness and transient hemiplegia have occurred; supposedly due to spasm of the retinal and cerebral vessels.

In 1941 Linnethal and Talkov[1] first reported three cases of Raynauds disease associated with diffuse fibrotic lung changes, rapidly developing respiratory difficulties and some degree of right sided heart failure. Unfortunately no post mortem examinations were obtained. In 1942 Linnethal[2] reported two additional cases with a post mortem examination in one case which revealed vascular changes in the lung and secondary associated pulmonary fibrosis.

Aside from these cases the literature is essentially devoid of reports of this syndrome. I would like to report an additional related case. Unfortunately, autopsy findings are again not obtainable; however, the course was so typical and the response to management so unusual that it is felt the situation warrants review.

The patient was a 46 year old white female who had first noted typical symptoms of Raynauds disease seven or eight years prior to her admission to the hospital. Despite routine management with priscoline, avoidance of cold, etc., her difficulty had become progressively severe until, at the time of her admission, gangrene had begun on several digits on both hands. Approximately two years after the onset of her peripheral vascular symptoms she had noted progressive dyspnea which had continued to increase in severity. One year prior to admission, ankle edema had developed and hepatomegaly had been described by her local physician. Her past history was essentially negative although it was interesting to note that previous examinations in earlier years, at various large centers, had revealed no signs of any cardiac pathology or lung disease and routine chest films and electrocardiograms has been normal.

The essential features, upon physical examination in 1956, showed dusky finger tips on both hands, with a markedly increased capillary filling time and areas of necrosis were noted over the terminal phalanges of several digits. The radial pulses were strong; a severe ankle edema was noted bilaterally and the liver was palpable three centimeters below the right costal margin. The cardiac examination was not remarkable, although the pulmonic second sound was greater than the aortic second sound and the left border of cardiac dullness was several centimeters outside the mid-clavicular line. A sinus tachycardia was present and no significant murmurs were noted. The routine laboratory work was normal. An electrocardiogram showed right

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ventricular hypertrophy without an associated block. The chest films also showed right ventricular enlargement with some slight increase in the vascular markings of the lung. Cardiac fluoroscopy failed to add any new findings and no signs of auricular enlargement could be determined. Routine pulmonary function tests were essentially normal.

The patient was rigorously treated with the usual measures for her peripheral circulatory findings as well as with digitilization, a low sodium regimen and diuretics for her right sided failure. Despite rigid hospital control little improvement was obtained. This necessitated a stellate ganglion block which produced a satisfactory ptosis, miosis and anhydrosis. There was considerable temporary relief from pain in the corresponding extremity and the capillary filling time was markedly reduced. Because of this result a bilateral upper dorsal sympathectomy was carried out with interruption of the rami, ganglia and chains from thoracic one, two, and three. The patient tolerated the operation poorly and developed a shock-like state near the end of the procedure. The patient survived but her situation was critical for several days. No obvious cause for the situation was noted at the time.

Following the sympathectomy her peripheral circulation was markedly improved and a sinus bradycardia replaced the previously noted sinus tachycardia. Inside of a week it became obvious that the signs of right sided failure were decreasing. Edema was much less marked and the liver edge began to recede under the costal margin. A chest film taken four weeks after surgery showed that "the heart is now appreciably smaller than previously noted prior to the operation". Diuretics, etc., were withdrawn but improvement continued despite any significant medication. When the patient was last seen at the hospital, all signs of right ventricular failure were lacking and the extremities were markedly improved with a continued decrease in capillary filling time and lack of duskiness and pain. Unfortunately, the patient died suddenly five months later in her home town during a minor surgical procedure and no autopsy was obtained.

This case is similar to Linnethal's in some respects. The patient started with typical Raynaud's disease which was followed shortly by progressive dyspnea. There were no other logical explanations for the symptoms and finds in either report. The rapid development of difficulty is not typical of any of the usual forms of pulmonary or cardiac disease. This patient differs somewhat from Linnethal's in that the x-rays were not especially suggestive of fibrosis and pulmonary function tests did not support pulmonary pathology.

There is a similarity between this situation and that presented by so called idiopathic primary pulmonary hypertension. In these cases only minimal vascular changes have been noted and it has been therefore postulated that the syndrome results from an increased pulmonary vasoconstriction similar to the essential hypertension in the systemic circulation. The primary objection to this concept has been the supposition that vasomotor control of the pulmonary arterioles is weak and irregular. The vascular lesions in these cases are only intimal thickenings suggestive of arterio-

losclerosis and are considered to be secondary to the pulmonary hypertension. A more severe syndrome had also been observed in which a necrotizing pulmonary arteritis had been seen at autopsy in association with right ventricular hypertrophy of unknown etiology. This situation has been considered analogous to the malignant hypertension of the systemic circulation. Sudden unexplained deaths and intolerance of minor surgical procedures have been reported as typical of these cases. The severe shock and the rapid death of the patient discussed would be compatible with these observations.

From the preceeding discussion I believe we have established some basis for the relief of symptomatology noted by the patient in this case. It would seem reasonable to suggest that further attempts at such surgery might be attempted in cases of primary pulmonary hypertension in an effort to eliminate the reduction in number and caliber of the pulmonary capillaries and arterioles.

In summary, an unusual form of cor pulmonale in assiciation with Raynauds disease has been reported and the dramatic and unexpected response to sympathectomy has been discussed. It would be hoped that a similar surgical approach will be evaluated in the future in the management of allied syndromes.

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