CLINICAL CONFERENCE

Tumor of the Heart

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DR. CASSELS: In April, 1954, a 19-month-old white female was admitted to the hospital. Her family history was irrelevant. She had a normal birth and had not been ill until about a week before admission. At that time, while sitting at the table eating, she suddenly made a noise, threw up her hands and became unconscious, and would have fallen if she had not been caught. She regained consciousness, and promptly again lost consciousness. On the way to a neighborhood hospital she regained and lost consciousness twice. During the first 24 hours of illness she had eight episodes of unconsciousness. In each of these she was limp and flaccid. At no time was there any rigidity or movement nor did she vomit or lose sphincter control. At that hospital it was noted that the eyes were swollen somewhat and the abdomen was protuberant. Urinalysis revealed 2+ albumin. No other abnormalities were found.

On admission to Bobs Roberts Hospital there was only a trace of albumin in the urine without other abnormalities. The eyes were swollen, the abdomen greatly distended, and the liver was very large. There was no evidence of cardiac disease and the blood pressure was normal in all extremities.

During the first month or two of hospitalization she was quite distressed. She had difficulty in breathing, urinated poorly, and developed marked generalized edema. She had astonishing dyspnea without orthopnea and, curiously, she sought peculiar positions of comfort in the oxygen tent. One of the peculiar positions of comfort was lying on the left side, turned at about a 45-degree angle toward a face-down position. She resumed this position repeatedly when moved for examination or nursing care and resisted the usual sitting position assumed for comfort by a patient with respiratory distress.

In the hospital she remained in the oxygen tent where she had a number of difficult episodes: A pneumothorax following a thoracentesis; following administration of intravenous fluids thrombosis occurred with an associated thrombophlebitis, the blood platelets dropped markedly, and she developed thrombocytopenic purpura.

At this time the diagnosis remained somewhat in doubt. It is proper to say that the people who saw her had a variety of suggestions. The original idea, that this was a nephrotic episode, was quickly dismissed. Also, an idea that she had nonspecific myocarditis was rather promptly dismissed. She responded badly to treatment with cortisone, becoming grossly edematous without any relief of apparent cardiac distress. Because I had recently seen a myxoedematous infant with somewhat similar signs and symptoms, thyroid was administered, in spite of the fact that the protein-bound iodine was normal, without benefit.

Although by August she was much improved, vascular and venous markings, which had appeared over upper thorax, neck and head, became more prominent. The edema in the lower extremities and fluid in the chest and abdomen diminished. There was increasing evidence of obstruction to the superior vena cava (Fig. 1). However, we could not understand the edema in the legs and the presence of ascites and a large liver, if there was an obstruction of the superior vena cava.

In October she seemed well enough to undertake studies to investigate an obstruction to the superior vena cava. It was thought that cardiac catheterization could only show obstruction, but that an angiocardiogram might reveal some details of size and position of an obstruction, and whether it was extrinsic or intrinsic.

During the 6 months of her illness electrocardiograms showed no gross abnormalities, although the amplitude of all complexes in-

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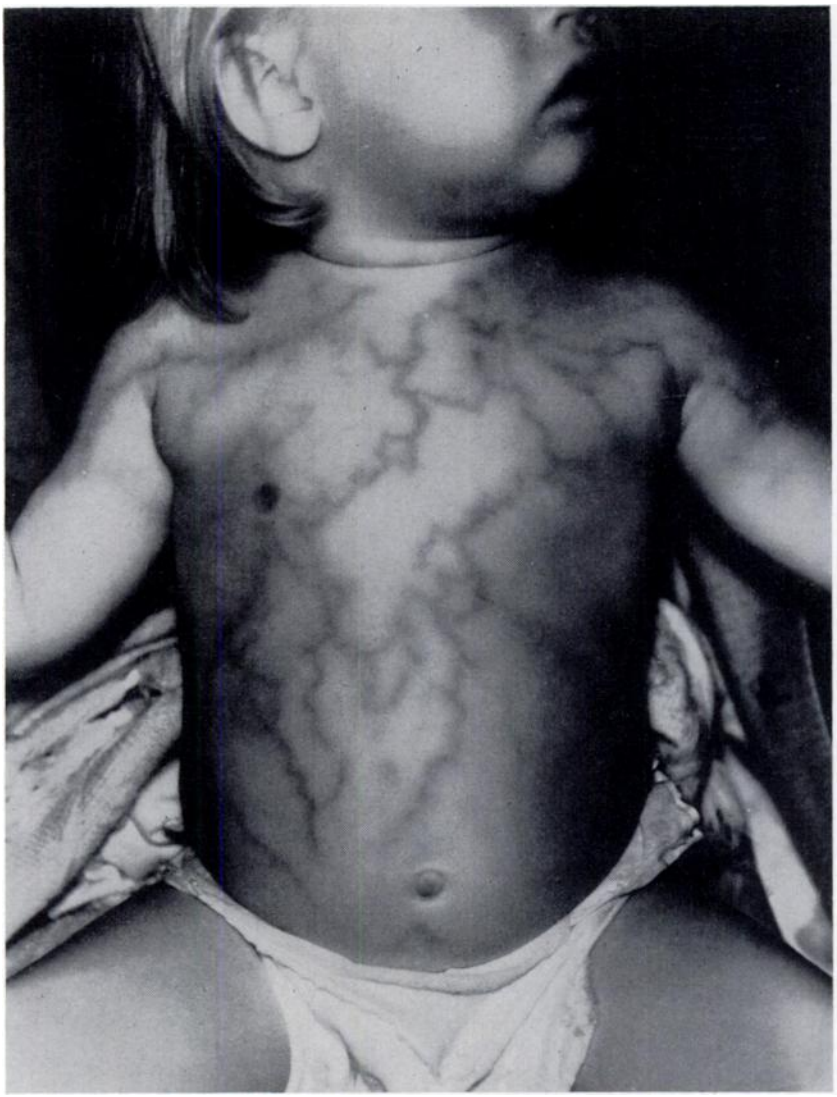

FIG. 1. Infrared photograph showing the collateral venous circulation over the upper part of the body.

creased as she improved. At no time were there ST changes compatible with myocarditis, nor was there ever any frank arrythmia.

The size and shape of the heart changed markedly, possibly related to pericardial fluid, although this was not demonstrated. The pulmonary markings also varied as though there were variations in pulmonary blood flow (Figs. 2, 3, and 4).

The angiocardiogram (Fig. 5) showed filling of the right heart through the distended collateral veins, and apparently the azygos vein was an important adjunct. Figure 6 reveals the basis of the venous obstruction, a large defect in filling which almost completely occupied the right atrium. There was a rim of opaque material around it, and not much flow into the pulmonary artery.

The presumptive diagnosis was an intracardiac tumor in the right atrium. It would be assumed, on the basis of incidence, that the mass would be a huge thrombus or a myxoma. The duration of the disease of this patient and the slow progression of symptoms and the fact that the circulatory status improved suggested that the tumor was not malignant. It was hoped that the mass was pedunculated and could be removed surgically.

The thoracic surgeons opened the chest

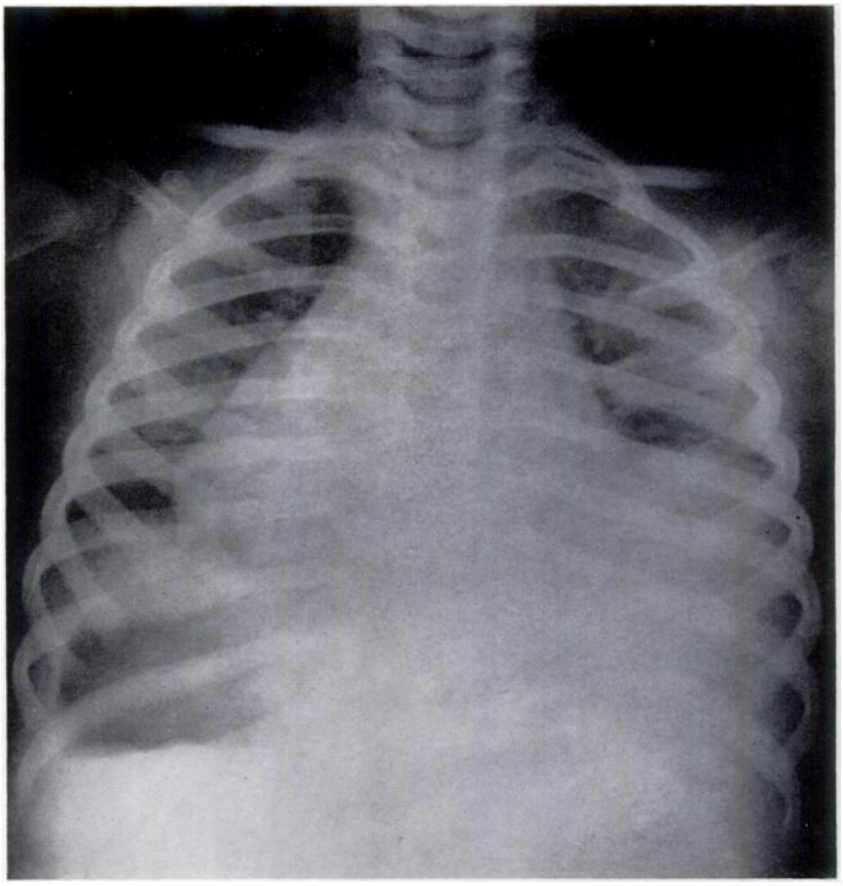

FIG. 2. Cardiac enlargement and hydrothorax seen on admission. Posterior-anterior view.

and as the heart was explored it was quite obvious that the right atrium, including the atrial appendage was filled with a huge firm mass widely attached to the atrial wall. A small nodule of tumor had grown through the wall and this was removed for examination. The patient recovered from the operation uneventfully.

Dr. Eleanor Humphreys, of the department of Pathology, identified the tumor as a mesothelioma. On the European continent this particular tumor of the heart is referred to as a benign coelothelioma of Tarawa's node, a term which describes the origin of the tumor from the tissue of the primitive coelome and indicates that it is usually found in the region of Tarawa's node.

Mahaim has suggested that the inclusion of epicardial tissue of the coelome in the substance of the heart occurs at the time of invagination of the auricular canal into the ventricle of the primitive cardiac tube.

On the basis of the great diversity of the cellular structure of the tumor, it was thought that it might be quite radiosensitive and therapy with x-rays was undertaken. After the first few hundred roentgen units, there was a marked improvement in the status of the circulation. Peripheral venous congestion diminished and respiratory distress completely disappeared. This early improvement was not maintained, and she again developed increased respiratory distress and great distension of the veins over different parts of the body. The respiratory distress was uncontrollable and she expired in March, 1955.

At necropsy the size and position of the tumor offered an explanation of most of the signs and symptoms observed. The superior vena cava was partially obstructed. The tumor mass, in addition, partially obstructed the inferior vena cava, thus explaining the

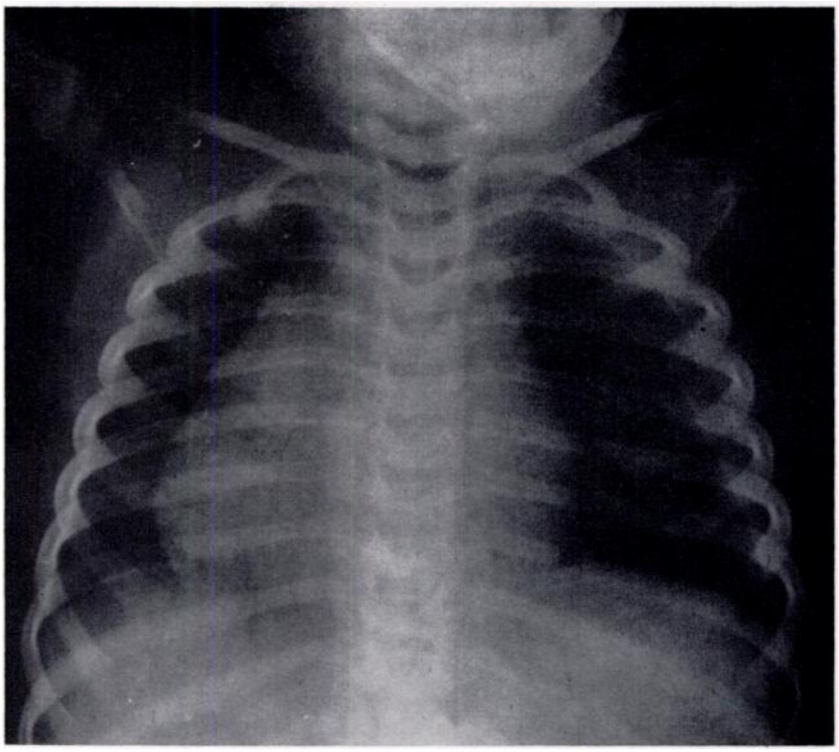

FIG. 3. Ten days after admission. The lung fields are abnormally clear.

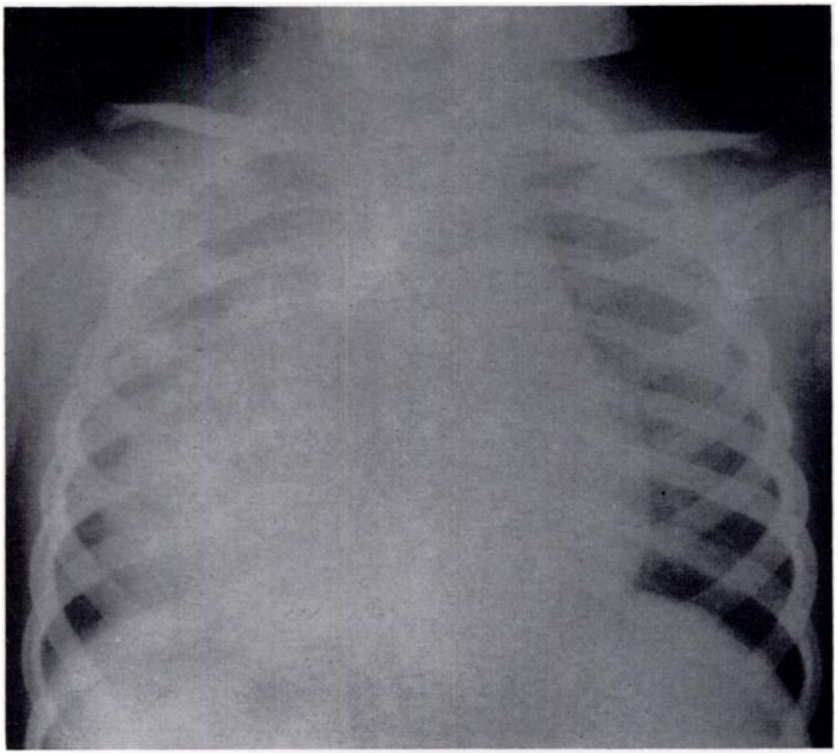

FIG. 4. Posterior-anterior view of chest in November, 1954, prior to operation. The heart is again large.

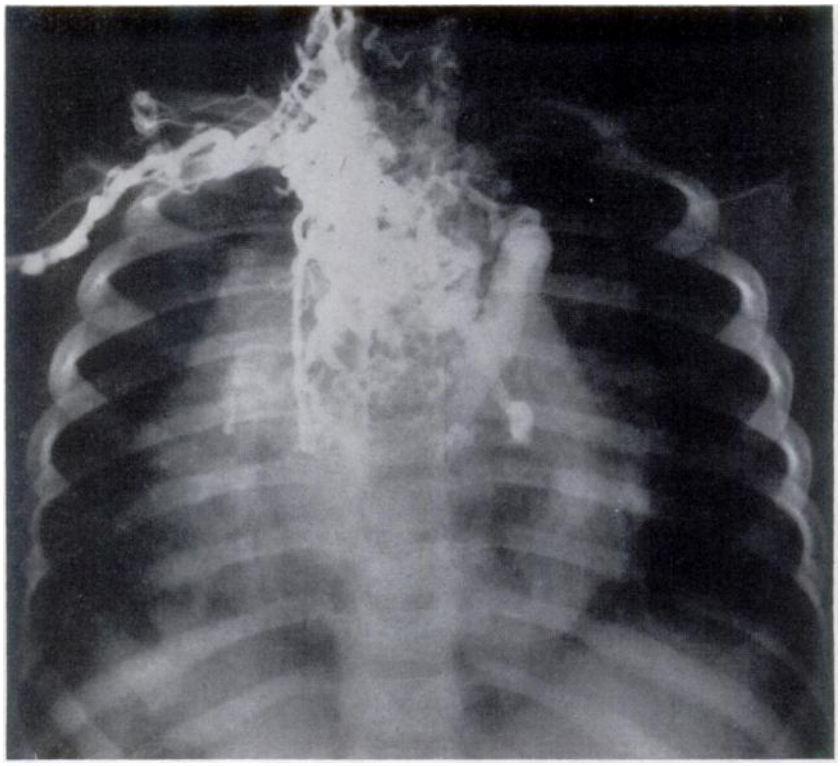

FIG. 5. Angiocardiogram, posterior-anterior view, showing the distended collateral veins and the large azygous vein. The right atrium is partially opacified, and a non-opacifying mass is seen.

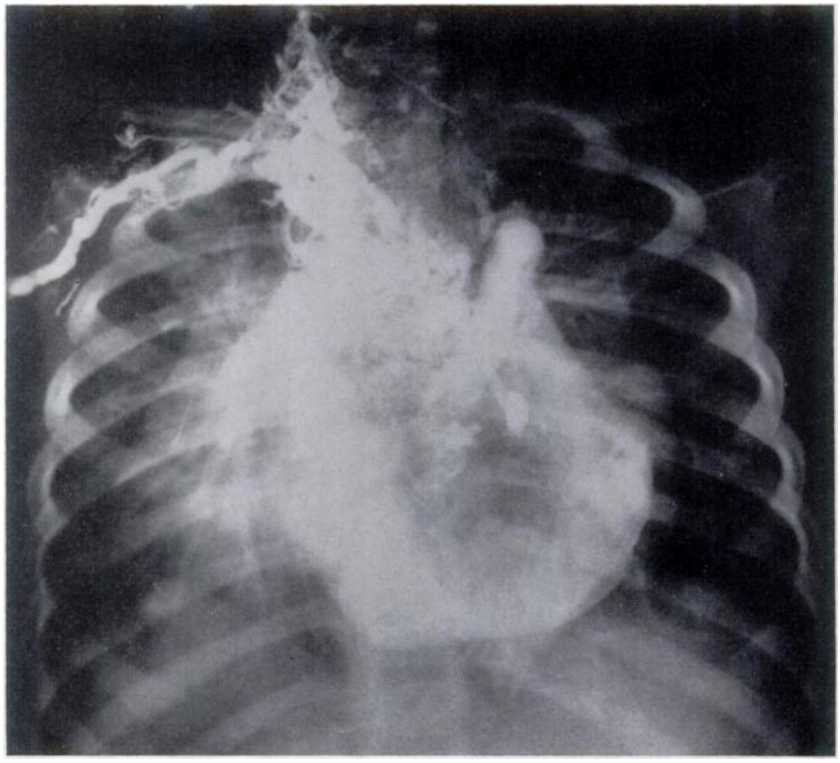

FIG. 6. The huge mass fills the distended right atrium. A small amount of contrast medium has entered the right ventricle and the pulmonary artery.

large liver, ascites and edema of the legs. In addition, the position of the mass was such that it could partly occlude the tricuspid valve. The partial face-down position, which at one time seemed to give comfort, would appear to have lessened obstruction to venous inflow from the inferior vena cava. It is postulated that the early episodes of loss of consciousness were related to intermittent diminution of venous flow, either into or out of the right atrium.

While tumors of the right atrium have been reported not infrequently, this instance of mesothelioma is the seventh example of this type of intracardiac tumor, coelothelioma of Tarawa's node.

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