Rhabdomyosarcoma of the Diaphragm

A Case Report

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Although secondary tumors of the diaphragm are fairly common, primary tumors in that location are rare. Binney (1) was able to find only four cases in the literature of the fifty-year period prior to 1931. Of these four, one was a benign fibromyoma, one a fibromyosarcoma, one a "round-cell sarcoma," and one a lipoma. Two of the four were found at operation and two at autopsy.

In 1932 Burvill-Holmes and Brody (2) reported the autopsy findings in a case of primary angiofibroma of the diaphragm which was considered benign; the patient also had pulmonary tuberculosis. Kirshbaum (3) reported two cases in 1935, one a rhabdomyosarcoma and the other a leiomyosarcoma. He also found one additional case in the literature, a myoblastic sarcoma reported by Müller.

We have been able to find only one example reported since Kirshbaum’s paper appeared. This was a lipoma of the pleural aspect of the diaphragm, which Söderlund (4) was able to diagnose by means of x-ray and thoracoscopy. The tumor was successfully removed and the patient was relieved of the pain in the side which caused her to seek medical advice.

It would appear, then, that there are in all 9 cases of primary tumor of the diaphragm recorded, of which 5 were malignant. Most of the reported cases occurred in females in the fourth decade of life (3). The following report adds another case to the literature, the patient being apparently the youngest on record.

Case Report

J. A., a fourteen-year-old Negro boy, was admitted to the Roper Hospital on Oct. 16, 1936, complaining of pain in the chest, at the right costal border and superior to the liver. The pain was not made worse by coughing or deep breathing. About two months before admission the patient had suffered a severe blow to the right side of the chest. Shortly after this a swelling appeared in the right lower anterior portion of the chest wall, which was moderately painful and tender. A dry cough and shortness of breath developed later, and shortly before admission to the hospital the boy began to raise a small amount of mucoid sputum. No history of hemoptysis could be obtained. There had been some fever for several weeks before admission, but no chills. Loss of weight and strength had occurred. The only previous illness had been whooping cough.

On examination it was found that the right side of the chest failed to move with respiration, although the movements of the left side were normal. There was a firm swelling over the lateral chest wall from the fifth rib downward in the anterior axillary line. This gave the impression of a swelling rather than of a distinct mass, and was intimately attached to the ribs. In the right upper quadrant of the abdomen was a firm resistant area, not defi-
nately continuous with the mass above, and itself not clearly defined. Although the lower border of this abdominal area of resistance was not sharp, it was thought to be the liver. It extended slightly below the umbilicus on the right, but not quite so low on the left. Overlying the third right rib at its costochondral junction was a nodular swelling 3 cm. in diameter, only slightly painful, firm and not fluctuant. This mass was distinct, and appeared not to be a part of the swelling below it. The right chest was dull to percussion throughout, and the breath and voice sounds were completely absent. Over the left chest the percussion note was hyperresonant and the breath sounds were normal. The heart was displaced to the left, but its action was regular and no murmurs were heard. Several firm, globular masses were felt in the right axilla, apparently lymph nodes, and another node was felt above the right clavicle.

On admission, the hemoglobin was 81 per cent, leukocytes 9,500 per cu. mm., polymorphonuclears 75 per cent and lymphocytes 25 per cent. Several urine examinations were completely negative, as were Kolmer and Kline tests on the blood.

The first x-ray examination of the chest showed the presence of an effusion on the right. Thoracentesis was done several times; the fluid was slightly bloody on a number of occasions, and at other times was straw-colored. In a pleural fluid specimen that contained little blood the specific gravity was 1.012, and lymphocytes predominated (85 per cent) in the stained smears, with 15 per cent eosinophils. No tumor cells were observed. A Gram-positive, encapsulated bacillus was grown from the pleural fluid, but was thought to be a contamination. Guinea-pig inoculation and culture of the fluid revealed no evidence of tuberculosis. Later, as much fluid was removed from the chest as possible, air was injected, and further chest films were made in several positions. One of these showed a large, roughly circular shadow of increased density continuous with the antero-lateral chest wall at the level of the fourth and fifth ribs near their costochondral junction, projecting into the thoracic cage (Fig. 1). The diaphragm was hazy and irregular in contour. The x-ray diagnosis was endothelioma of the pleura or chondrosarcoma.

Several weeks after admission an aspiration biopsy was made from the mass about the fourth and fifth ribs. The smears prepared from the aspirated material, as well as sections of blood clot obtained, showed numerous large polyhedral cells in a granular, eosinophilic background. A diagnosis of malignant tumor was made, but the tumor could not be classified.

Fluid reaccumulated rapidly in the right chest, so that aspiration had to be done every four or five days, with removal of about 1000 c.c. of fluid each time. In spite of thoracente-
sis, dyspnea became progressively more distressing and cyanosis developed. The temperature was irregularly elevated, fluctuating from 98° to 101°. The hemoglobin gradually fell to 50 per cent and the red cell count to 3.5 millions. The patient became weaker and more emaciated, and death occurred Dec. 2, 1936, almost eight weeks after hospital admission, and about four months after the injury to the chest wall and onset of symptoms. Among the diagnoses made during life were chondrosarcoma of the ribs, hypernephroma, and medullary carcinoma of the adrenal gland.

Autopsy was done a few hours after death. The right side of the chest wall was unduly prominent, and there was a row of nodular swellings intimately attached to the ribs on the right, curving anteriorly and upward from the posterior axillary line at about the tenth rib to the costochondral junction of the third right rib. No masses were palpable to the left of the mid-line. Several nodular masses were felt in the right axilla, and one in the right supraclavicular fossa.

On midsection there was a small amount of free bloody fluid in the abdomen. Tumor tissue infiltrated subcutaneously over the lower portion of the right side of the chest. The liver was not enlarged but was pushed downward by a mass in the diaphragm, and its free border extended 10 cm. below the right costal margin. The right pleural cavity contained about 1000 c.c. of bloody fluid and much air; the left contained about 50 c.c. of slightly blood-tinged fluid. In the pericardial sac was a slight excess of clear fluid.

The heart was of average size and appeared normal. The thyroid and larynx were normal. There were some enlarged lymph nodes in the mesentery, especially about the right iliac fossa and in the pelvis, but the intestinal tract proper was normal. The liver weighed 1150 gm. Just beneath the diaphragmatic surface of the right lobe of the liver, was a small, cylindrical mass of tumor tissue surrounding an hepatic vessel, but there was no apparent invasion of the vessel and no evidence of thrombosis. Except for this, the liver was free of tumor. The spleen was normal except for a small tumor nodule in its capsule at the hilus. The kidneys, ureters, bladder, prostate and testes were normal. The pancreas was surrounded by enlarged lymph nodes but itself appeared unchanged. The suprarenals were likewise surrounded by lymph nodes replaced by tumor, but were themselves normal.

The tumor was confined almost exclusively to the right side of the body. It extended from the anterior, posterior, and lateral attachments of the right lobe of the diaphragm to slightly beyond the mid-line, enveloping the diaphragmatic orifice of the inferior vena cava as a solid ring. The anterior aspect of the orifice of the aorta was composed of tumor, and the orifice for the esophagus was surrounded by flat nodules of tumor tissue in the diaphragm, but not by a definite mass. The tumor did not extend farther to the left than this, except for some small flat nodules on the abdominal surface of the diaphragm. On the right the mass was very firm; it had a white, whorled appearance on section, and was 5 to 7 cm. in thickness. It was rigid and inelastic. The normal dome-like curvature of the right diaphragm was lost, and the diaphragmatic mass gradually sloped forward from the crura to the attachment to the chest wall anteriorly. This depression of the right lobe of the diaphragm, together with moderate shift of the mediastinum to the left, resulted in a great increase in the size of the right pleural cavity. The mass at the right crus of the diaphragm was just as thick and definite as in the body of the diaphragm proper. The cisterna chyli was filled with tumor tissue and surrounded by numerous large, firm lymph nodes, evidently replaced by tumor. Similar nodes were noted about the hilum of the liver.

Arising from the upper surface of the flat mass representing the right lobe of the diaphragm, and continuous with it, was a mass of almost equal thickness, extending upward in the mediastinum as far as the arch of the aorta. In this mediastinal mass many matted nodules were to be made out, apparently the structural remains of lymph nodes replaced and enlarged by tumor tissue. Similar tumor tissue extended upward along the right parietal pleura, penetrated between the ribs on the right, and lay in the subcutaneous tissues over the right side of the thorax. This pleural growth extended to the apex of the pleural cavity and was then continuous downward with the mediastinal growth. In the upper mediastinum the tumor was mainly posterior to the heart, surrounding and compressing the azygos vein, although in no place was its lumen invaded. The right lung was completely collapsed against the mediastinum and was only 4 cm. thick at its widest portion. The tumor growth was
heaviest over the mediastinal surface of the lung, but also extended over its lateral aspect and into the interlobar fissures. The growth in the lung was confined to the ramifications of the visceral pleura, and did not extend into the lung proper, although it occupied many of the lymph nodes at the hilum. On the left, the lung was partially collapsed by fluid, but was in the main air-bearing. There were nodules of tumor tissue in a few of the hilar nodes on the left, but these were not so conspicuous as on the right, and again there were no parenchymal nodules in the lung tissue proper. The mediastinal vessels, trachea, and esophagus showed no invasion of their walls, and were not greatly compressed from without.

The blood vessels in the region of the diaphragm and liver were dissected as well as possible, and showed no gross evidence of vascular invasion. The bones of the head and extremities were not examined carefully for metastatic lesions, but none was palpable externally, and the pelvis and vertebral bodies showed no evidence of tumor when exposed through the usual incision. Even the ribs and costal cartilages on the right, which were surrounded by tumor tissue in many places, showed no sign of actual invasion.

Fig. 2. Section of the Diaphragmatic Surface of the Liver, Showing an Area of Small-cell Growth

Hematoxylin and eosin: reduced from a photomicrograph × 240.

The impression was gained that the tumor began in the right leaf of the diaphragm, extending downward along the right crus of the diaphragm to involve the cisterna chyli. From this point, retrograde lymphatic extension to the retroperitoneal lymph nodes and to the pelvic and mesenteric lymph nodes took place. At the same time the tumor extended throughout the lymphatics of the right lobe of the diaphragm, along the lymphatics into the triangular ligament of the liver, and for a short distance into the liver proper; along the lymphatics of the anterior and posterior mediastinum; via the lymphatics of the peripheral portion of the diaphragm to reach the attachments of the diaphragm to the ribs; by the pleural and intercostal lymphatics to spread throughout the parietal pleura and between the ribs and costal cartilages into the subcutaneous tissues. From the subcutaneous tissues of the right side of the chest wall extension occurred to the right axillary lymph nodes and to the right supraclavicular node.

Microscopic sections of the tumor in the diaphragm showed anaplastic cells, varying greatly in size and shape. Most of the cells were small and polyhedral, the nuclei large and hyperchromatic, with the chromatin so dense that the nucleoli could scarcely be made out
as such (Fig. 2). The cytoplasm was quite eosinophilic and somewhat granular, and frequently extended backward from one end of the cell as a tail. Between the cells were the remnants of the voluntary muscle fibers of the diaphragm, and in addition a prominent eosinophilic, fibrillar, stroma-like material whose fibrils could frequently be traced to tumor cells. More conspicuous and more unusual, although less numerous than the smaller cells, were the multinucleated giant cells. Some of these were round or ovoid, with large nuclei
about the periphery of a densely eosinophilic cytoplasm. A more remarkable appearance, and probably a more characteristic one, was that of the large "tadpole cells" (Fig. 3). In the rounded heads of these cells were clustered many large nuclei, while projecting backward from the head was a tail-like cytoplasm, fibrillar and eosinophilic. Cross-striations could not be demonstrated in these cells or in any others. Another cell that was thought to be rather characteristic—at least we have never encountered such a cell in any other type of tumor—was an exceedingly large oblong one, roughly resembling the tadpole cell, but having a much broader tail, with nuclei gathered about the outer surface of the tail also (Fig. 4). The cytoplasm of this cell had the appearance of sarcoplasm, although there were no apparent striations; its appearance slightly suggested a muscle bud. The tadpole cell and the larger oblong cell were fairly common, although the latter was encountered only once or twice in each slide. These cells lay intermingled with other types of tumor cells, and their shape seemed to bear no relation to external pressure or to mechanical distortion of any sort.

The tumor had a similar appearance in all the metastases; in fact, the giant cells seemed more numerous in sections of pleura and lymph nodes than in the diaphragm. The growth in lymph nodes and pleura had the same fibrillar stroma as that seen in the primary tumor in the diaphragm.

Definite vascular invasion could not be made out in any section, and it seemed that the tumor spread entirely by the lymphatic route rather than by the blood stream.

A final diagnosis of rhabdomyosarcoma of the diaphragm was made, and confirmed by consultation with Dr. Fred Stewart of the Memorial Hospital, New York.

SUMMARY

A case of malignant tumor of the diaphragm (rhabdomyosarcoma) in a fourteen-year-old Negro boy is presented, with autopsy findings. The tumor appeared to metastasize entirely by the lymphatic route.

This is apparently the tenth case of primary tumor of the diaphragm on record, the sixth case of malignant tumor primary in the diaphragm, and the second case in which the parent tissue was voluntary muscle.

REFERENCES