Primary Malignant Mesothelioma of the Pericardium
Report of a Case

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Primary malignant neoplasm of the pericardium is exceedingly rare. Its origin has evoked interest for many years and this has been demonstrated through many recent publications.

Many authors have denied its existence, and Willis1 has reached the conclusion that mesotheliomas of any serous membranes with few exceptions were not primary mesothelial tumors, but rather metastatic neoplasm. Robertson2 does not believe in mesothelioma as a distinct neoplastic condition.

Mesothelioma as primary tumor is being recognized by an increasing number of pathologists after the experimental demonstration of the properties of the mesothelioma cells in culture by Stout and Murray3 and confirmed later by Sano et al.,4 as well as the histochemical demonstration of hyaluronic acid in mesotheliomas,5,6 though a considerable amount of research is still required.

We have had the opportunity to observe a case of primary malignant mesothelioma of the pericardium, in a young man, extending through the parietal surface and causing obstruction of both venae cavae and resulting in a fatal course within a few weeks.

Case Report

A 22-year-old white man entered the Napoleão Laureano Hospital (Cancer Hospital), complaining of painless nodular mass in the right cervical region and chest pain of five months' duration, accompanied by non-productive cough. At the time of admission, he had no positive physical findings relative to the lungs or abdomen and he had no weight loss.

The blood pressure was 120/70 mm.Hg. Examination of the neck revealed a hard, fixed nontender lymph node mass, situated in the right cervical and supraclavicular region, 4 x 3 cm.

The skin was not adherent. The chest expanded well and the lungs were clear on physical examination. The axillary nodes were slightly enlarged, firm and the inguinal nodes were not enlarged. A biopsy of the cervical region was performed and the slides were reported as papillary carcinoma, probably metastatic.

Two weeks after admission, he noticed a small mass in the anterior chest wall, just below the manubrium, which increased in size slowly. A roentgenogram (Fig. 1), showed an area of rounded density in the mediastinum and frontal tomography disclosed at 8 to 12 cm. deep, a large anterior mediastinal mass, extending into both sides, but more evident on the right side of the aorta.

Four weeks after admission, he complained of dyspnea and occasional dry cough. At that time, he also developed swelling of the face, neck and venous neck distension with discrete cyanosis. Three days later, he complained of an increase in his dyspnea with orthopnea and paroxysmal nocturnal dyspnea besides edema of the abdominal and chest wall, scrotum, legs and feet. The liver was felt four fingerbreaths below the right costal margin and it was firm, smooth and not tender.

Figure 1: Posteroanterior view showing a mediastinal tumor.

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Despite active medical supportive therapy, the patient died one and one-half months after admission.

Necropsy Findings: At necropsy, there was an enlarged and thickened mass corresponding to the parietal pericardium covering the entire heart (Fig. 2) and more evident in the base of the heart. The entrance of superior vena cava into the right atrium was constricted so that the index finger could not be admitted. This tumor was firmly adherent around the entry of the inferior vena cava and in the posterior surface of the sternum which exhibited pathologic fracture with extensive osteolysis.

The tumor mass was pale in color and showed spongy white tissue with microcystic areas covered by a honey-like fluid. The central portions were hemorrhagic and necrotic and no calcifications were present.

The pleura was firmly adherent to the diaphragm and exhibited spongy grayish nodules, 1 to 2 cm. in diameter on its surface. This finding was limited to the right side. The left pleural cavity contained 1000 ml. and the right 1200 ml. of bloody viscous fluid.

The myocardium, endocardium and heart valves were normal. The left ventricular muscle measured 1.3 cm. in thickness and the right 0.4 cm. The mediastinal and hilar lymph nodes were involved and the cervical nodes were replaced by a friable mass with jelly-like appearance. The right lung showed fibrinous adhesions with the corresponding pleura. Both lungs exhibited moderate congestion and edema.

The liver weighed 1500 gm. and the cut surface showed the characteristic "nutmeg" appearance of chronic passive congestion.

The cranial contents were not examined.

No other organs showed the presence of either primary or metastatic tumor or other significant change.

The entire tumor measured 22 x 18 x 8.5 cm. At the base of the heart the neoplastic parietal pericardium was 4.2 cm. in thickness.

Microscopic Findings: The tumor was formed by marked proliferation of cells predominantly of the epithelioid type, which present a characteristic papillary and pseudoglandular pattern. The papillary processes were supported by cores of connective tissue and covered by columnar or cuboidal cells (Fig. 3). A detailed examination of these cells disclosed scanty cytoplasm, large vesicular nuclei with clumped chromatin, well-defined nuclear membrane and rather prominent nucleoli. In some areas, chiefly in the tubular or pseudoglandular pattern, the nuclear membrane assumed a crinkled appearance.

There were occasional atypical mitoses, which were more prominent among the cells arranged in cords. The parahilar and cervical lymph nodes were replaced partially by proliferation of cells arranged in cords and papillary processes, forming in some areas clefts and gland-like spaces (Fig. 4). Groups of epithelioid cells in lymph nodes were sometimes dispersed by very loose gelatinous stroma.

Amounts of fibrous tissue were seen in the tumor, associated with an infiltration of inflammatory cells, chiefly lymphocytes, monocytes and plasmocytes. Definite transition from epithelioid cells to fusiform cells in the tumor were not observed.

Microscopic examinations of sections taken from lungs, liver and other viscera showed no evidence of neoplasm.

**Figure 2:** Gross appearance of the heart showing a pericardial tumor surrounding the great vessels and constricting both vena cavae.
COMMENTS

For many years, the diagnosis of mesothelioma as a primary neoplastic condition of the serosa cells, has been the subject of considerable discussion and controversy. Some physicians have been enthusiastic regarding the conclusion that they were not primary mesothelial tumors, but rather neoplasm that had metastasized to the pleura, pericardium, or peritoneum. It would seem that this subject should be settled after the tissue culture studies by Stout and Murray confirmed later by Sano et al. However, confusion still persists regarding the diagnosis, though the demonstration of the primary tumor of serosa cells without evidence of tumor inside organs at necropsy would be a strong argument in favor of its primary origin. The clinical manifestations of the mesothelioma of the pericardium are chiefly dependent upon the encasing of the heart in a thick neoplastic hull that eventually is responsible for the patient's death. According to Dawe et al., the death in this patient is due to circulatory failure resulting from a loss of cardiac filling capacity or to acute fluid tamponade resulting from hemorrhage either within the pericardial space or within the tumor. The accumulation of serosanguineous pericardial fluid has been stressed by many authors. Thomas and Phytyon have pointed out that the clinical course in this case was marked by the rapid reaccumulation of massive amounts of bloody pericardial fluid.

Figure 3: Detail of papillary structure and cuboidal cells lining very little supportive stroma.

Figure 4: Pseudo-glandular area with well-defined cuboidal and columnar cells.
The majority of cases in the review of Dawe and associates showed diffuse tumors covering most of the heart, often obliterating the pericardial cavity and constricting the great vessels of the base. The superior venae cavae showed either occlusion with thrombus or constriction with reduction of its lumen. Thomas and Phythyon showed a tumor encasing the heart like a mantle and at the base of the heart, all the great vessels were surrounded by the neoplastic process, but did not seem to be compressed.

Dawe's case showed a diffuse mesothelioma replacing the entire surface of the parietal pericardium and constricting the superior venae cavae at the entrance into the right atrium. This case is quite similar to our present case which caused pronounced constriction of both venae cavae and showed clinically edema and venous distension of the neck, edema of the thoracic and abdominal wall, scrotum, feet and legs. This constriction was a factor responsible for the patient's death, added to the "tissue tamponade" of Dawe et al.

The presence of fibrous tissue in the present case showed close relation to the organization of hemorrhagic focus and inflammatory areas and we could not be certain of finding any relationship with the true neoplasm. However, there have been reports in the literature stressing the capacity of transformation of mesothelial cells into fibroblast and epithelial-like cells. This dual potentiality of mesothelial cells would account for the appearance of fibroblastic or epithelioid components of the tumor observed in the latter cases.

The histochemical demonstration of hyaluronic acid in mesotheliomas has been emphasized by many authors, as well as the demonstration of positive periodic acid-Schiff (PAS). These findings have been controversial and although they represent a great advance in the identification of mesothelial tumor cells, a considerable amount of research is still required.

References

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