Primary liposarcoma of the heart

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Summary: A rare form of primary heart tumour, a liposarcoma, is reported. A discussion of diagnosis and modalities of treatment of primary heart sarcomas is presented.

Introduction

Primary tumours of the heart are rare, occurring in one large autopsy study in fewer than 0.017 per 1000 cases (Strauss & Merlis, 1945). In approximately 75% of cases the tumours are benign, being either myxomas in adults or rhabdomyomas in children (Heath, 1968). The remaining 25% are malignant, virtually all of which are sarcomas. In the following report a rare type of primary heart sarcoma is presented together with a discussion of the various means of detection and modalities of treatment of such tumours.

Case report

A 60 year old woman with a history of increasing palpitations and dyspnoea on exertion, of a few months duration was admitted to the hospital with pulmonary oedema. Following treatment, a soft diastolic murmur was heard along the left sternal border. Chest X-ray and electrocardiogram were consistent with a slightly enlarged left atrium. An echocardiogram and angiogram performed to rule out mitral stenosis revealed a solid mass fixed to the posterior wall of the left atrium.

In January 1978 the patient underwent surgery. A 6 cm, gelatinous, unencapsulated, broad-based mass was excised from the postero-lateral wall of the left atrium. Pathological examination revealed a pleomorphic tumour with ample mucoid ground substance staining positively with alcian green, and numerous blood vessels. Cells of different size and shape with atypical hyperchromatic nuclei and mitoses, as well as cytoplasmic vacuoles, were seen (Figures 1, 2). Sudan staining for fat was positive in the cytoplasm of many cells, and based on the heterogeneity of the tumour

cells, a diagnosis of a pleomorphic liposarcoma was made. A work-up to exclude metastases or other possible primary tumour sources – including tomograms of the mediastinum, a liver-spleen scan, and abdominal computed tomographic (CT) scan – was negative.

Two months following surgery a pleural effusion and a mass in the right lower lobe of the lung appeared. A thoracotomy one month later revealed a 15 × 20 cm mass attached to the right diaphragm histologically similar to the previously excised mass in the heart. Four weeks following excision of the mass, radiation therapy was delivered to the mediastinum and right diaphragmatic regions to a dose of 3800 cGy using a cobalt 60 unit; this was followed by a boost of 1500 cGy to a reduced field. Four months later bone metastases appeared after which chemotherapy consisting of cyclophosphamide, vincristine, doxorubicin, and DTIC (CYVADIC) was started. Following the

Figure 1 Liposarcoma of the heart revealing atypical mitoses, pleomorphic nuclei and vacuolization. H & E × 400.

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fourth course of this treatment, supraventricular rhythm disturbances developed. Cardiotoxicity was suspected and on echocardiography, enlargement of the left and right ventricles was noted. Doxorubicin was therefore stopped after a total dose of 280 mg and in its place actinomycin was substituted. No objective or subjective response was noted under this treatment and in January 1980, 24-months after diagnosis, the patient died.

Post-mortem examination showed extensive skeletal, lung and liver metastases. The heart was diffusely dilated and showed severe fibrous pericarditis, a mural thrombus in the left atrium with a nest of tumour cells and non-bacterial thrombotic endocarditis of the mitral valve.

Discussion

In order of decreasing frequency the histological types of primary malignant heart sarcomas are angio-, rhabdomyo-, fibro-, and lymphosarcomas (McAllister & Fenoglio, 1978). Least common are liposarcomas with only three cases prior to the present one being reported in the literature (McAllister & Fenoglio, 1978; Fletcher, 1963; Blake *et al.*, 1972). Only in one of these cases was a diagnosis made ante-mortem.

Depending on their size and location within the heart, malignant tumours may present with symptoms of right- or left-sided heart failure, pericarditis with or without tamponade, or rhythm disturbances (Silverman, 1980). On chest X-ray the cardiac contour may be normal or it may display chamber enlargement mimicking obstructive valvular heart disease. Whereas none of these signs or symptoms are pathognomonic of malignancy, the use of two-dimensional echocardiography has made a high degree of diagnostic accuracy possible (Feigenbaum, 1981). Intramural, sessile or pedunculated masses can be

localized, and echolucencies within the masses, characteristic of vascular channels of haemangiomas and angiosarcomas, visualized as well. Other cardiac imaging techniques including computerized tomography and angiography have also proved helpful (Weyne *et al.*, 1985).

The prognosis of primary heart sarcomas is poor. There is usually a rapidly progressive course, with death occurring within weeks to months of diagnosis, either due to widespread infiltration of the myocardium and/or distant metastases (Colucci & Braunwald, 1984). Owing to the scarcity of cases of primary heart sarcomas, no controlled study of treatment approaches exists. Yet there is no evidence to suggest that they behave differently from aggressive sarcomas arising in other organs. The role of surgery is mostly in diagnosis and palliation, the tumours being generally infiltrative and difficult to excise in their entirety. Rare reports of prolonged survival following curative attempts, however, do exist (Grabelman *et al.*, 1979). Radiotherapy likewise serves a palliative role, at times sterilizing the tumour bed (Sagerman *et al.*, 1964). Recently, reports of improved survival in patients with sarcomas of the extremities who received doxorubicin-containing chemotherapy as part of their primary treatment have appeared (Eilber *et al.*, 1984). The implications for cardiac sarcomas are unclear due to the scant experience with chemotherapy in this disease. It should be noted, however, that an increased potential for cardiotoxicity due to the interaction of radiation and doxorubicin on the heart does exist and may hinder the use of doxorubicin-containing regimens as seen in the present case (Rosen *et al.*, 1974). In such cases the careful clinical monitoring of the patient, including repeated electrocardiograms, echocardiograms, and ejection fractions as calculated by cardiac scanning, is warranted. Endomyocardial biopsies may also be considered in the framework of controlled studies (Friedman *et al.*, 1978).

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