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Pleomorphic high-grade sarcoma of the heart mimicking cardiac myxoma

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DESCRIPTION

A 22-year-old male patient with no significant previous medical history presented with fatigue, dyspnoea, palpitations and signs of upper inflow tract congestion. Echocardiography (figure 1a) and CT revealed a mass of 4×6 cm (*) in the left atrium which prolapsed into the left ventricle during diastole and displayed radiological and ultrasonographic traits of an atrial myxoma (sharp borders, pedunculated appearance, mobility during cardiac cycle, localisation in left atrium, lack of detectable infiltration into cardiac/pericardiac structures). Histopathological examination of the surgically excised mass revealed a myxoid tissue structure low in cell count (figure 1b) with multi-layered perivascular cell agglomerates resembling myxoma

ring structures (figure 1b inset). However, the perivascular cells involved were strikingly large and other domains of the extirpated tumour corresponded to a pleomorphic spindle cell neoplasia (figure 1c) with elevated nuclear polymorphism, mitosis rate (arrows) and proliferation (MIB1-immunostaining, figure 1c inset) as well as immunohistochemical positivity for actin, vimentin and partially for keratin (not shown). As postoperative tumour staging ruled out the presence of a primary extracardiac neoplasia, a primary pleomorphic epithelioid high-grade sarcoma of the heart (G3) was diagnosed (reference pathological confirmation by Professor C Fletcher, Harvard Medical School). Further postoperative staging revealed multiple lytic bone lesions (figure 1d arrows), which were confirmed

Figure 1 (a) Echocardiography of the heart revealing a tumour mass in the left atrium which prolapses into the left ventricle during diastole (highlighted by *). (b) Histopathological image of extirpated tumour mass showing myxoid tumour areas with low cell count and multi-layered agglomerates of large, pleomorphic cells around blood vessels (inset). H&E-stain. Magnification: 20x (main image), 40x (inset). (c) Histopathological image of extirpated tumour mass exhibiting a pleomorphic spindle cell neoplasia with markedly elevated nuclear polymorphism, mitosis rate (arrows) and proliferation (MIB1 staining, inset). H&E-stain (main image) and MIB1 immunohistochemistry (inset). Magnification: 60x (main image), 40x (inset). (d) CT scan of the spine showing multiple lytic lesions (arrows) and histopathological identification of punch biopsy samples from bone lesions as metastases of a pleomorphic high-grade sarcoma (inset). H&E-stain (inset). Magnification: 20x (inset).

as sarcoma metastases (figure 1d inset). Malignant cardiac sarcomas show a tendency toward myxoid degeneration.^{1 2} Therefore, in case of preoperative doubt, particularly diligent clinical and histopathological scrutiny of suspicious cardiac tumours is advisable and should be augmented by further imaging examinations as well as tumour staging procedures.

Learning points

- Cases diagnosed as cardiac myxoma should be particularly diligently worked up in order to rule out the presence of cardiac sarcoma with myxoid degeneration.

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Competing interests None.

Patient consent Obtained.

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