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Cytologic-Pathologic Correlations

Imprint cytology of primary cardiac sarcomas: a report of 3 cases

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Abstract Primary cardiac sarcomas are rare instances and only occasionally documented in the cytologic literature. Usually, the diagnosis of these rare lesions can be made at echocardiography, aspiration biopsy cytology, cardiac biopsy, and open cardiac surgery (intraoperative diagnosis). In this study, cytologic configurations and immunohistochemistry for 3 primary cardiac sarcomas (rhabdomyosarcoma, angiosarcoma, and malignant fibrous histiocytoma) were revealed. In rhabdomyosarcoma (right ventricle), the tumor cells exhibited an anisocytotic spindle-shaped nuclei with hyperchromasia and an obscure cytoplasmic margin. Vimentin and myosin were positive throughout the cytoplasm for the tumor cells. In angiosarcoma (right atrium), small clusters of anisocytotic spindle-shaped tumor cells appeared as vascular-like structures and hemosiderin-laden macrophages in many erythrocyte-rich backgrounds. Nuclei showed round to oval shape with hyperchromasia and prominent large nucleoli. Cytoplasm was obscure and elongated. Factor VIII related antigen and CD34 were strongly positive throughout the cytoplasm for the tumor cells. In malignant fibrous histiocytoma (right ventricle), the tumor cells exhibited oval to spindle-shaped and elongated nuclei and coarse granular chromatins with hyperchromasia. The nuclear margin was thin. A few small round nucleoli appeared. Elongated obscure and foamy cytoplasm was stained pale blue. Vimentin and α_1-antitrypsin were positive throughout the cytoplasm for the tumor cells. This study elucidated the cellular characteristics and immunohistochemistry for cardiac sarcomas using imprint smears as an aid to cytopathologic diagnosis.

Keywords: Cardiac sarcomas; Imprint cytology; Immunohistochemistry; Rhabdomyosarcoma; Angiosarcoma; Malignant fibrous histiocytoma

1. Introduction

Primary cardiac sarcoma is a rare tumor accounting for a mere 0.05% of all tumors [1,2] and has a poor prognosis with a median survival of appropriate 12 to 16 months [3]. At present, cardiac tumors are fairly easy to diagnose clinically antemortem via echocardiography (ECG), computed tomography (CT), positron emission tomography, and magnetic resonance imaging [4,5]. However, reports on its pathologic features are rarely encountered in the literatures [6,7] and consisted of only a few illustrations with brief comments in a few cytologic articles [8,9]. In recent years, immunocytochemistry has been used to corroborate cytomorphology, and this approach has greatly enhanced the diagnostic accuracy of the lesions. In this article, we present cytologic and immunohistochemistry features of a cytologic material from 3 cases of primary cardiac sarcoma and discuss the differential diagnosis.

2. Materials and methods

Three primary cardiac sarcomas were used in this study: rhabdomyosarcoma (case 1), angiosarcoma (case 2), and

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malignant fibrous histiocytoma (case 3). Tissues obtained from surgery and autopsy were smeared on a few slide glasses from one portion of tumor mass and fixed with 95% ethanol preventing air drying, and then stained using Papanicolaou and immunohistochemical stain. Giemsa stain was also performed. Excised samples for the histopathologic examination were fixed with 10% formalin. Paraffin-embedded tissue sections of the tumors were stained with hematoxylin and eosin (H&E) stain and compared with cytology. Immunohistochemical staining was performed using the Envision/horseradish peroxidase (HRP) system (DAKO, Glostrup, Denmark), with monoclonal and polyclonal antibodies obtained commercially, epithelial membrane antigen (EMA), CD68, α_1-smooth muscle actin, α_1-antitrypsin, desmin, vimentin, keratin, CD34, myosin, lysozyme, and factor VIII related antigen (DAKO). Envision/HRP system of tissue sections and imprint smears was described as follows: (1) treat with proteinase K (DAKO) for 5 minutes, (2) rinse in tap water for a few minutes, (3) add 3% hydrogen peroxidase for 5 minutes, (4) rinse in tap water for a few minutes and wash in 0.01 mol/L phosphate-buffered saline (PBS), pH 7.4, for 1 minute, (5) add drops of antibodies for 30 minutes, (6) wash with 3 changes of PBS for 5 minutes each, (7) add drops of polymer reagent for 20 minutes, (8) wash with 3 changes of PBS for 5 minutes each, (9) add drops of diaminobenzidene (DAB) for 5 minutes, (10) rinse with tap water for 5 minutes, and (11) counter stain with hematoxylin for 1 minute and mount with resin. All incubations were done in a humid chamber at room temperature. Negative controls were incubated with nonimmune rabbit serum in the first steps of the indirect procedure. The presence of brown reaction products in the cytoplasm was indicative of antigen positive.

3. Reports of case

3.1. Case 1

A 66-year old man was admitted to the National Cardiovascular Center, Osaka, Japan, complaining of heart palpitations, dyspnea, and weight loss. Chest CT and ECG examination revealed an abnormal shadow from the right ventricle to the pulmonary artery that was judged to be a tumor. During operation, the pulmonary artery was found to be completely obstructed by the tumor mass. Unfortunately, 1 year later, the patient had died, and restricted autopsy was performed on the heart and lungs. Autopsy findings revealed that the primary tumor having ivory color (10 × 5 × 5 cm) was situated at the right ventricle outflow with obstruction and was growing into the right ventricle space with massive hemorrhagic necrosis and extended to the antiseptal wall. The tumor had metastasized to both lungs (2 cm in diameter), the diaphragm and liver with tumor nodules.

3.1.1. Histologic findings

The diagnosis was a pleomorphic type of rhabdomyosarcoma; the tumor cells consisted of eosinophilic and pleomorphic spindle-shaped tumor cells with massive necrosis and hemorrhage. Multinucleated giant cells with several nuclei in abundant cytoplasm and strap-shaped cells were seen with hyperchromasia (Fig. 1A). Negligible mitotic structures and cross-striations were found with phospho-

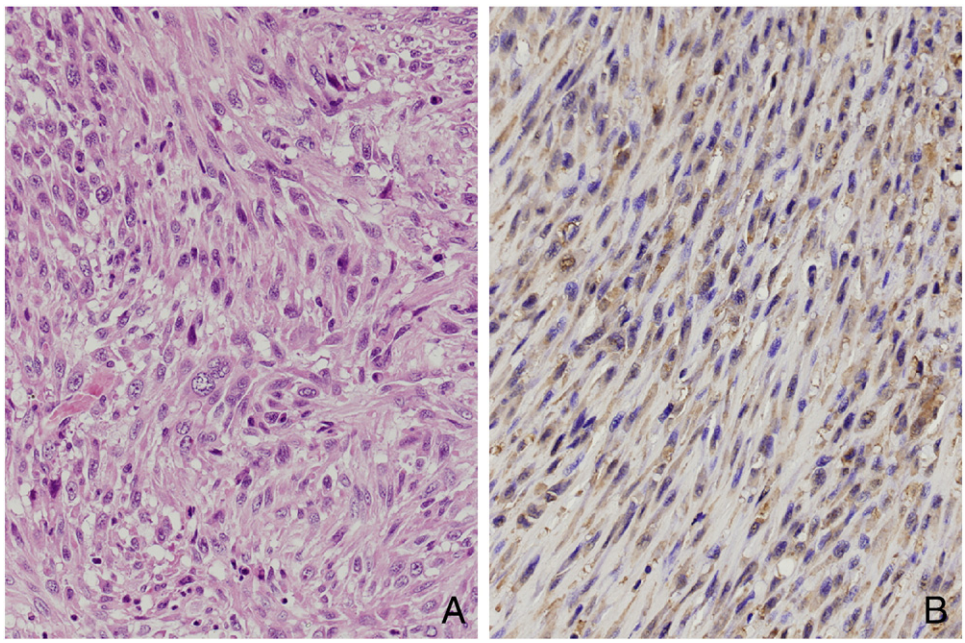

Fig. 1. Histopathology of rhabdomyosarcoma. (A) Pleomorphic spindle-shaped and multinucleated tumor cells with eosinophilic cytoplasm (H&E). (B) Myosin is positive throughout the cytoplasm for the tumor cells (immunohistochemistry).