An Extraskeletal Myxoid Chondrosarcoma Diagnosed by Fine Needle Aspiration Biopsy

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Extraskeletal myxoid chondrosarcoma (EMCS) is a rare malignant soft tissue sarcoma described as a distinct clinicopathologic entity by Enzinger and Shiraki in 1972 (1). The tumor usually develops in deep parts of the proximal extremities and in middle aged adults. EMCS has a prolonged clinical course with a high rate of local recurrence (2). Only a few reports have been published on the fine needle aspiration biopsy (FNAB) of EMCS (3,4).

Case

A 43-year-old male patient was sent to the aspiration unit of our laboratory for a third recurrent mass in his right gluteal region that extended to the inguinal zone. The mass first appeared in the gluteal region about 3 years previously, with a second one appearing a year after that. Neither lesion had been examined histopathologically. The adjacent bone was not attached to the mass on MRI and CT scans. The gluteal mass was 9 cm and inguinal mass was 6 cm in diameter. Both lesions were aspirated by fine needles (20-22 gauges). Some of the slides were air dried and stained with May-Grünwald-Giemsa. Others were fixed in ethanol and stained with hematoxylin-eosin (HE) and Papanicolaou stains.

Microscopically the slides were generally hypercellular, and contained numerous tumor cells that were isolated or in small sheets and loose aggregates. The cells were embedded in an abundant, blush-pink granular myxoid matrix. The tumor cells in the smears were relatively uniform. The nuclei were hyperchromatic, round or ovoid, and showed mild nuclear atypia. They had variable amounts of eosinophilic cytoplasm (Figures 1-2).

The case was diagnosed cytologically as ‘malignant myxoid neoplasm’. Based on the clinical and radiological findings the tumor was reported as ‘could be extraskeletal myxoid chondrosarcoma’. The lesions were excised with extended surgical margins after the cytologic diagnosis. The diagnosis was confirmed histopathologically.

EMCS is a rare soft tissue tumor. This entity was first described in 1972 (1). The tumor has a prolonged clinical course with local recurrences and occasionally pulmonary metastases (2,5). Our patient had 3 different masses in about 3 years in the same location. After the last operation he was followed up for eighteen months, during which time he had no local recurrence or distant metastases. We are continuing to monitor his condition.

The usefulness of FNAB for the histologic subtyping of specific sarcomas is controversial (6). FNAB of soft tissue sarcomas can be classified into 6 categories: myxoid, spindle cell, pleomorphic, polygonal cell, round cell and miscellaneous (7). In myxoid neoplasms the smear background is strikingly myxoid. Cytologically, the tumors in the differential diagnosis of myxoid neoplasms include myxoid liposarcoma, myxoid malignant fibrous histiocytoma, EMCS, myxoma, metastatic mucinous adenocarcinomas, myxomatous degeneration of tumors, aggressive angiomyxoma and chordoma (2). Myxoid liposarcomas and myxoid malignant fibrous histiocytomas are the most important tumors in differential diagnosis. In myxoid liposarcomas lipoblasts are characterized by rounded contours and scant to moderate volumes of cytoplasm that is occupied by one or more sharply defined lipid vacuoles. Their nuclei vary in size and show
hyperchromasia. Another distinctive feature is the presence of branched delicate capillaries. Our smears had neither lipoblast-like cells nor branching arteries reminiscent of liposarcoma. Aspirates from malignant fibrous histiocytomas have abundant myxoid material in the smears with 3 morphologic cell types, small spindle shaped fibroblast-like cells, histiocytic cells and multinucleated giant cells. These giant cells are most characteristic of the myxoid malignant fibrous histiocytomas. These types of cells, especially giant cells, were not seen in our case. Aspirate smears from intramuscular myxomas are poorly cellular, and are characterized by voluminous matrix material with scattered spindled and stellate cells manifesting no atypia. Chondroblast-like cells that also had atypism were observed in our slides. Aggressive angiomyxoma is generally located in the perineal area. The smears of these tumors are usually hypocellular and cells are spindle shaped. The location of the tumor and the smear specifications were helpful in ruling out aggressive angiomyxoma. Metastatic mucinous adenocarcinomas tend to be more like epithelial groups and may have intracellular mucin. The cells in our case are configured smaller sheets of neoplastic cells that are not reminiscent of epithelial groups. There was no clinical history or evidence of mucinous carcinoma anywhere in the body nor only radiological or clinical aspects. The aspirates of chordomas are moderately to highly cellular and most are typical physaliphorous cells. They are generally located in the pelvic region and the retroperitoneum (2). None of these cells were present in our smears, and the location of the tumor was not generally in the expected area.

In conclusion FNAB can be helpful in the diagnosis of soft tissue sarcomas. In myxoid soft tissue neoplasms FNAB can successfully be used even in advanced differential diagnosis as an easily applicable method.

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