International Journal of Pathology; 2004; 2(2):108-110

Case Report

Monophasic Synovial Sarcoma Diagnosed on FNAC

Ambreen Moatasim and Anwar Ul Haque

Department of Pathology, Pakistan Institute of Medical Sciences, Islamabad.

Fine Needle Aspiration Cytology (FNAC) has found great grounds in various areas to provide accurate, rapid, painless and inexpensive mode of rendering early diagnosis. Its marriage with imaging technologies had broadened and widened its horizons which are now virtually limitless. Bone and Soft tissue tumors are quite suitable for diagnosis by FNAC. Not only common but rare tumors can be easily diagnosed by FNAC. Synovial sarcoma is a well defined clinical and morphological entity. It is so named because of its resemblance to developing synovial tissue under light microscope.

Synovial sarcoma comprises 8-10% of all sarcomas and most commonly affects adults in the third-to-fifth decades of life. The malignancy most commonly involves the extremities, especially the lower extremities around the knee joints.[1] The FNAC plays key role in diagnosis of soft tissue and bone tumors. The FNAC smears are generally hypercellular in malignant tumors. These show crisp, artifact free cellular details on cytology smears. In collaboration with X-Ray findings, history and physical observations, FNAC can provide valuable diagnosis quickly and without much pain and cost.

Here we describe a case of rather uncommon monophasic Synovial sarcoma, which we diagnosed on FNAC and later confirmed on surgical pathology.

Case Report

A 52 year old male presented with a large painful swelling on the anterolateral aspect of proximal right leg approximately 12 cm in diameter. The swelling was progressively increasing in size and was associated with off and on fever. On examination it was a large non specific, somewhat lobulated swelling. It was tender, mobile, fluctuant with no restriction of knee movements. His base line investigations were within normal limits. Chest X-ray, CT chest and ECG were normal. MRI of the right leg suggested a sarcomatous lesion. Bone scan revealed increased uptake. Fine needle aspiration cytology and subsequently, a core needle biopsy was performed.

Cytologic Findings: Atypical spindle shaped plump cells were present scattered as well as in cohesive clusters. These atypical cells had pleomorphic, hyperchromatic nuclei with irregular chromatin clearing and nuclear indentations. The cytoplasm was eosinophilic and relatively ample.

Histopathological Findings: The microscopic examination of the core needle biopsy specimen revealed a malignant mesenchymal neoplasm in which the malignant cells were arranged in solid compact sheets. These cells were spindle shaped with pleomorphic, hyperchromatic nuclei having prominent nucleoli and irregular chromatin clearing. The cytoplasm was plump and relatively ample. The N/C ratio was nonetheless higher than reactive Synovial cells. The picture conformed with the cytopathological findings. There were 3 mitoses per high power field. On the basis of these features, a diagnosis of Monophasic Synovial Sarcoma was made.

Discussion

Synovial sarcoma arises from pluripotential mesenchymal cells near joint surfaces, tendons, tendon sheaths, juxta-articular membranes, and fascial aponeuroses. Synovial sarcoma is the fourth most commonly occurring sarcoma, accounting for 8-10% of all sarcomas. Approximately 800 new cases of synovial sarcoma are diagnosed per year in the United States. Overall, survival rates are 23.5-64% at 5 years and 11.2-34% at 10 years. For synovial sarcoma of the head and neck region, the prognosis is better than that of sarcoma involving the extremities, with 5-year survival rates of 47-82%.[1]

A slight male predilection exists in synovial

sarcoma. The male-to-female ratio is 3:2. Synovial sarcoma can occur in patients with a wide age range,

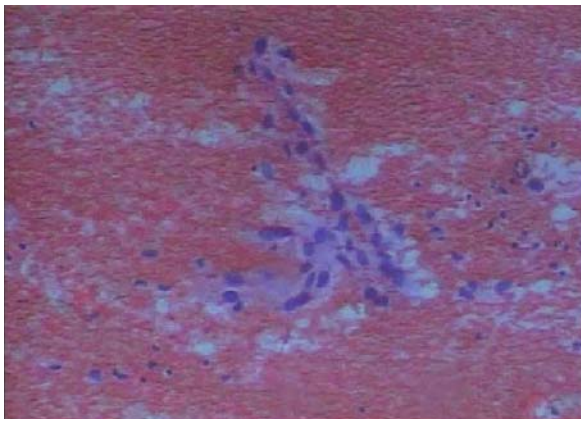

Fig. 1: FNAC of the lesion showing cohesive clusters as well as scattered atypical spindly cells (H&E X 100)

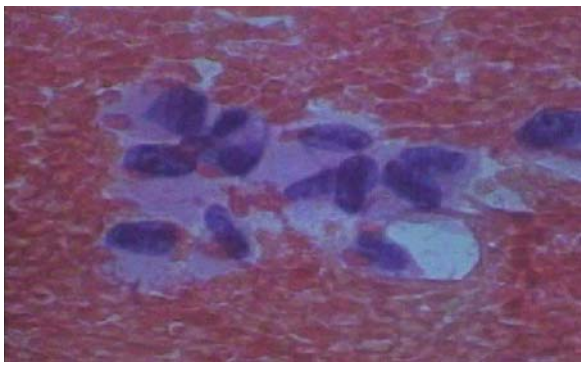

Fig. 2: Morphological details: Atypical cells are plump and spindly having ovoid, pleomorphic, hyperchromatic nuclei with irregular chromatin clearing. Cytoplasm is abundant and pale. (H&E X 400)

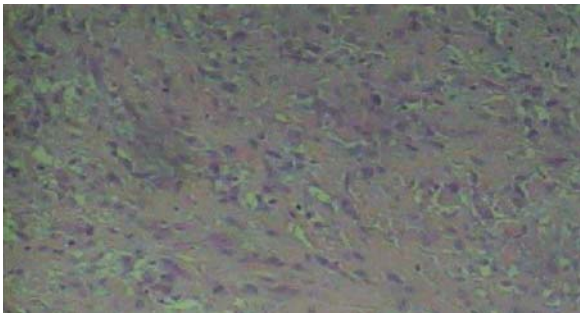

Fig. 3: Histopathology showing hypercellular areas composed of spindly atypical cells. (H&E X 100)

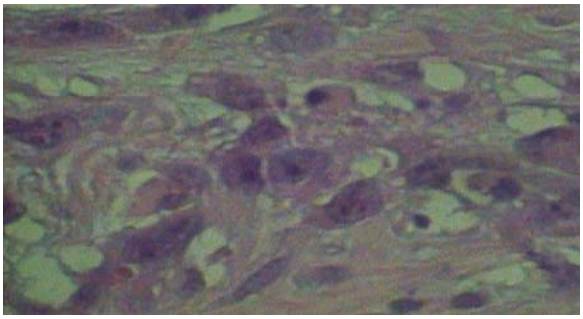

Fig. 4: Higher magnification showing cellular details more clearly (H&E X 100).

but it is most common in patients in the third-to-fifth decades of life. In one series of 121 cases, 83.6% of tumors occurred in patients aged 10-50 years, with a median age of 31.3 years.

Most synovial sarcomas are found within 5 cm of a joint. Despite the misnomer, only 10% of cases are intra-articular. The malignancy most commonly involves the extremities, especially the lower extremities around the knees. Tumors that occur in the upper extremity tend to affect the distal extremity rather than the elbow or shoulder. Less common sites of involvement include the retroperitoneum, mediastinum, and head and neck regions. The most common site in the head and neck is the hypopharynx. The clinical features of synovial sarcoma are nonspecific. Most commonly, patients notice a slowly enlarging, deep-seated mass, which is painful in slightly more than one half of patients. In head and neck involvement, patients complain of symptoms such as dyspnea, dysphagia, hoarseness, and headache.

Specific cytogenetic abnormalities have been identified. More than 90% of patients have a t(X;18) translocation mutation, which is not associated with other sarcomas. The translocation involves the SYT gene on chromosome 18 (at 18q11) and the SSX1 or

SSX2 gene on the X chromosome (at Xp11). These genes appear to be transcription regulators, whose functions occur primarily through protein-protein interactions. Subtypes of these translocations have been shown to correlate with distinct histologic subtypes.

Gross specimens are usually well-demarcated, pink, fleshy masses with a heterogeneous appearance. The specimens may display solid, hemorrhagic, or cystic components on sectioning. Calcification foci occasionally are noted. Heavy calcification tends to indicate less aggressive lesions and offers a more favorable prognosis. Microscopically, two forms are recognized: biphasic and monophasic; biphasic form is composed of both epithelial-cell and spindle-cell components, whereas the monophasic form can be either the epithelial-cell or the spindle-cell type. In spindle cell form there is predominant spindle cell component (monophasic synovioma) which contains cords of spindle cells which may resemble fibrosarcoma. The differential diagnosis includes malignant hemangiopericytoma, fibrosarcoma and spindle-cell squamous-cell carcinoma. The epithelioid form may be confused with adenocarcinoma. It reveals a biphasic pattern: intermixed areas of "glandular" synovial like cells & spindle shaped fibrous cells; this glandular area will stain PAS positive. The synovial cells have an acinar, ductal, or longitudinally arranged tall columnar cells around acellular "slits" containing mucin. The fibrous component is arranged in the herringbone pattern of fibrosarcoma. Immuno-histochemically, there is strong reactivity for keratin in the epithelial areas and often in the spindle cells as well. CEA, epithelial membrane antigen, vimentin and occasionally S-100 protein are also expressed by this tumor.[2]

X-ray reveals hazy, soft tissue density with discrete intrinsic calcifications in 30% of cases; periosteal reaction or even bone erosion or invasion. There is marked radioisotope uptake on bone scan.[3]

Open biopsy is often difficult to perform, and the procedure is associated with an increased prevalence of complications such as anesthetic complications, unnecessary amputations, poor wound healing and breakdown, fracture, bleeding, and infection. Image-guided biopsy is now commonly performed for the initial histologic diagnosis of soft-tissue tumors because of its comparative ease, safety, and cost-effectiveness of this less-invasive procedure. Core biopsy can be easily performed by using a 14-gauge Tru-Cut needle or similar device. Fine-needle aspirations are performed with smaller 20- to 22-gauge needles. FNAC is now being used more and more as a diagnostic procedure, alternate to excision biopsy.[4] FNAC has distinct advantages over open biopsy. It is a safe, simple, highly economical and relatively painless procedure which can be performed in office, at bed side and in out patient department, as it does not require anesthesia and operation theatre facilities. It provides reasonably accurate pre-op diagnosis in vast majority of cases.[5] At most centers, it has achieved undisputed status of a definite diagnostic tool forming the basis on which therapeutic protocol is selected.[6] Some authors report that fine-needle aspiration is adequate for the diagnosis of suspected metastatic disease, in which case the accuracy is high. Some authors advocate the use of both techniques in the percutaneous biopsy of soft-tissue tumors, stating that the information obtained is complementary, and that many lesions are diagnosed by means of one and not the other. Accuracy rates for the diagnosis of lesions biopsies by both fine-needle aspiration and core biopsy are generally higher than that of either technique alone.

Tumors are treated aggressively with limb-sparing therapy when possible. The recommended treatment is wide resection with negative margins, which often include surrounding muscle groups or total amputation. Resection is commonly followed by localized irradiation. Multi-drug adjuvant chemotherapy is currently recommended for systemic control of the cancer. Fine needle aspiration cytology is not associated with needle tract deposits of malignant cells. FNAC diagnosis in monophasic synovial sarcomas thus provide a preoperative diagnosis with ample time to the patient and surgeon to decide about most appropriate management in individual cases.

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