Cytological findings of an extragonadal yolk sac tumor presenting at an unusual site

ABSTRACT
Yolk sac tumor (YST) is a rare neoplasm that affects children and adolescents. Fine needle aspiration biopsy is an extremely useful procedure for the diagnosis of YST. Main objective is to describe the characteristic cytological features of fine needle aspirates of YST. We report a case of YST in a 4-year-old male child occurring at paravertebral region showing cytomorphological details.

Key words: Aspiration cytology; extragonadal; paravertebral area; yolk sac tumor

Introduction

Yolk sac tumor (YST) is a rare neoplasm, which affects children and adolescents. Majority of the YSTs occur in the gonads, but about 20% have been found in variety of extragonadal sites such as mediastinum, retroperitoneum, neck and presacral areas.[1] Teilum described this as a malignant germ cell neoplasm that showed differentiation towards extra embryonic yolk sac endoderm as an endodermal sinus tumor. The majority of ovarian tumor of this type should have a distinctive pattern with differentiation toward yolk sac structure.[2] Present case describes the cytological features of YST occurring in paravertebral region, which to the best of our knowledge has not been reported till now in the literature.

Case Report

A 4-year-old male child admitted with a mass in the left gluteal region of 2 months duration and paravertebral swelling since 15 days. The patient also had a history of urinary retention and bowel incontinence following incision and drainage on left gluteal region mass by a medical practitioner. General physical examination revealed paralysis of both the lower limbs. Ultrasonography abdomen showed enlarged liver with multiple well-defined mixed echogenic foci in both lobes of the liver, metastatic deposits, along with a large ill-defined (10 cm × 5 cm × 7 cm) hypoechoic mass in the deep pelvic region extending posteriorly into gluteal region. Genitals were normal. Serum alpha-fetoprotein level was elevated (1,020 IU/mL, normal <30 IU/mL). Fine needle aspiration (FNA) was performed from the gluteal and paravertebral masses using a 22-G needle. Air-dried smears were stained with May-Grünwald-Giemsa and Papanicolaou stain. Cytological findings—Smears were highly cellular. The cells were arranged in cohesive clusters of variable size and shape against a mucoid background. Some of the clusters had a distinct papillary configuration [Figure 1]. The tumor cells contained ovoid nuclei with irregular nuclear membranes, fine to coarse clumped chromatin and prominent nucleoli [Figure 2]. Majority of the cells showed abundant cytoplasm containing numerous irregularly shaped vacuoles. Some of the tumor cells revealed intra cytoplasmic inclusions of pink homogeneous material [Figure 3], which were periodic acid Schiff (PAS) positive. Single cells were infrequent. Background was mucoid and had scattered foamy histiocytes. Based on the cytological findings, a diagnosis of malignant germ cell tumor was made, but Schiller-Duval...
bodies were not identified. A wide excision biopsy was performed which showed the features of YST, exhibiting variable histological patterns (reticular, solid, pseudopapillary and polyvesicular vitelline) along with Schiller-Duval bodies. Patient was referred to the oncology department where chemotherapeutic agents (vincristine, bleomycin, and cisplatinum) were started and he responded well with decrease in size of the sacro-coccygeal mass.

Discussion

Malignant germ cell tumors account for about 3% of pediatric malignancies and among these tumors YST account for most of the cases. The tumor was not a generally recognized entity until the 1960s, when Teilum described the tumor in the testes of the young children and in the ovaries, and drew attention to their origin as these tumors are derived from totipotential cells that differentiate primarily into extraembryonic structures that resemble the endodermal sinus of the rat placenta. The Armed Forces Institute of Pathology Classification considered the lesion to be the infantile variant of embryonal carcinoma, and the lesion was given separate status among germ cell tumors in WHO classification in 1977.

Endodermal sinus tumor was reported to arise primarily from testis and ovaries, but some of the cases may occur in extragonadal sites such as the vulva, vagina, pineal region, broad ligament, prostate, cervix, mediastinum, sacro-coccygeal and retroperitoneum region, but by far the commonest site for extragonadal lesions is the sacro-coccygeal region. The cytological findings of the endodermal sinus tumor have been reported previously revealing clusters of cells with a glandular appearance, large pleomorphic cells, vacuolated cytoplasm and PAS positive, diastase resistant hyaline globules. Ultra structurally, the cell membranes were well defined with frequent desmosomes. The cytoplasm contained prominent rough endoplasmic reticulum, numerous mitochondria, free ribosomes, glycogen granules, some pinocytic vesicles and dense membrane bound inclusions with the appearance of the lysosomes. The nuclei had an irregular configuration with small clumps of heterochromatin in the periphery along with one or more nucleoli.

In our case, a 4-year-old male child presented with endodermal sinus tumor of extragonadal region, predominantly consisting of cohesive clusters of pleomorphic cells, arranged in papillary like configuration containing abundant intracytoplasmic pink vacuoles.

Endodermal sinus tumor has highly variable histological patterns (reticular, solid, pseudopapillary and polyvesicular vitelline) reflecting toward extra-embryonic yolk sac structures which are not appreciated in cytological samples. Since the tumor’s cytological spectrum is very
broad, difficult differential diagnosis of yolk sac tumor are poorly differentiated adenocarcinoma, dysgerminoma and embryonal carcinoma. The distinction between YST and Embryonal carcinoma on the basis of FNAC may be difficult because of the epithelial appearance of both the tumors. The tumor cells of embryonal carcinoma show pronounced nuclear pleomorphism and blurred nuclear membranes, cellular cohesion with groupings in ragged-edged clusters, sometimes attached to branching capillaries, is helpful in making the diagnosis whereas dysgerminoma discloses single cells and loose groups which contain cytoplasmic glycogen vacuoles and a large nucleus with presence of lymphocytes, plasma cells, epithelioid cells, furthermore, an amorphous background, called “tigroid background.” The presence of intracellular and extracellular hyaline globules that are PAS stain positive and diastase resistant and correspond to alpha-fetoprotein production. Alpha-fetoprotein positivity of the neoplastic cells, showing nuclear pleomorphism and vacuolated “bubbly” cytoplasm are the most consistent cytologic hallmarks of this neoplasm.[11]

Yolk sac tumor tends to recur locally and also have a high incidence of metastatic disease at the time of presentation. Our patient also showed metastasis in the liver at the time of presentation along with sacro-coccygeal swelling.

Complete excision should be attempted in a malignant lesion. The outlook is improving with the use of combination chemotherapy, and long-term responses are being noted. Adjuvant Vincristine, actinomycin-D and cyclophosphamide chemotherapy has been most extensively used in children with endodermal sinus tumors. The role of radiotherapy is less certain in YST of extragonadal origin. Metastatic lesions may require palliative treatment with local radiation. Patients with sacro-coccygeal tumors have a greater probability for survival than those with tumors at other sites. Early detection and therapy is important because it is highly aggressive tumor that shows good response to surgery and chemotherapy.[12]

Conclusion

Yolk sac tumors commonly occur during childhood and adolescence and arise in the sacro-coccygeal region, carry a poor prognosis if adequate chemotherapy is not given. YST can be diagnosed accurately by means of fine needle aspiration biopsy (FNAB). A rapid and specific diagnosis obtained by FNAB may be extremely beneficial since it may save considerable expenses and spare the patient a host of diagnostic procedures. FNAB diagnosis in these cases may also help in planning definitive therapy, which may include chemotherapy and radiotherapy, in addition to surgical resection.

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How to cite this article: Kataria SP, Misra K, Singh G, Kumar S. Cytological findings of an extragonadal yolk sac tumor presenting at an unusual site. J Cytol 2015;32:62-4.

Source of Support: Nil, Conflict of Interest: None declared.