

# Cytodiagnosis of extragonadal pure yolk sac tumor of sacrococcygeal region

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## ABSTRACT

Yolk sac tumors (YST) are rare in extragonadal location. Cytologic diagnosis of YST in such instances may be a challenge to the pathologist. We studied Fine needle Aspiration (FNA) material from a sacrococcygeal mass in a 18-month-old female child presenting with a progressively increasing sacrococcygeal mass. Cytologic features include large pleomorphic balls of tumor cells with high nuclear to cytoplasmic ratios, papillary or micro-glandular pattern of growth, cytoplasmic and intra-nuclear vacuoles, and prominent nucleoli. Per Iodic Acid Schiff (PAS) positive intra-cytoplasmic as well as extracellular hyaline globules were also seen. The subsequent biopsy was consistent with the FNA diagnosis, and immuno-histochemically, the tumor cells were positive for Alpha Feto Protein (AFP) supporting the diagnosis.

**Key words:** Endodermal sinus tumor, extragonadal, fine needle aspiration, sacrococcygeal, yolk sac tumor

## INTRODUCTION

Yolk sac tumor (YST) is a malignant germ cell tumor that usually arises in the gonads. Their occurrence in extragonadal locations are rare.<sup>[1]</sup> We report FNA findings in a rare case of sacrococcygeal YST in a 18-month-old female child and review the pertinent literature.

## CASE REPORT

An 18-month-old female child presented with a progressively enlarging sacral swelling since 2 weeks. Parents of the child also complained of decreased urination since 4 days. Clinical examination revealed a diffuse boggy swelling measuring 4 × 5 cm over the sacrococcygeal area. CECT revealed a heterogeneously enhancing necrotic mass in the sacrococcygeal region with invasion of lower rectum, anal canal, and spinal canal space with destruction of the coccyx. In addition, there were multiple secondaries in the lung. Pre-operative serum AFP levels were also raised (181,500 units).

FNA was performed from the mass and smears were stained with Giemsa stain. Microscopic examination revealed numerous, large, tightly cohesive branching papillary fragments comprising of moderate to severely pleomorphic tumor cells having ovoid to elongated irregular enlarged nuclei, reticulogranular chromatin, inconspicuous nucleoli, moderately abundant finely vacuolated basophilic cytoplasm, indistinct cell borders [Figure 1], and frequent tendency towards acini formation. Few clusters revealed pink hyaline basement membrane-like material in between the tumor cells. Numerous mitotic figures, occasional tumor giant cells, and PAS-positive intra-cytoplasmic as well as extracellular hyaline globules were also seen. There were increased amount of PAS-positive globules in the vicinity of these hyaline bands [Figure 2]. On the basis of the cytomorphologic features, a diagnosis of YST was rendered. The subsequent biopsy was consistent with the FNA diagnosis and immuno-histochemically the tumor cells were positive for AFP supporting the diagnosis.

## DISCUSSION

Malignant extragonadal germ cell tumors are rare in infants and children. They occur in the body's midline and may have internal and external manifestations.<sup>[2]</sup> These uncommon neoplasm accounts for approximately 3% of childhood tumors that may arise in a variety of anatomic sites.<sup>[3]</sup>

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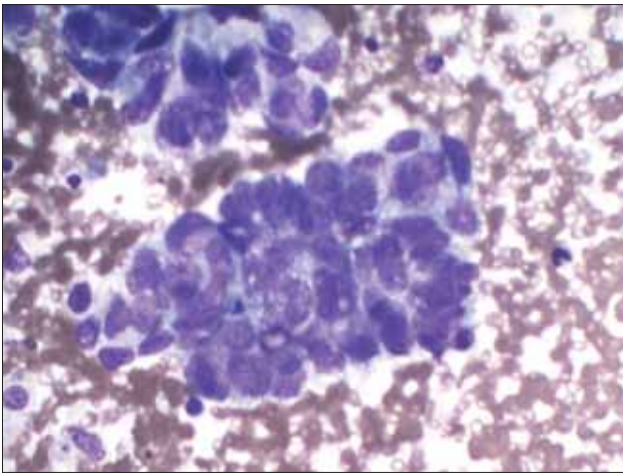
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DOI:

10.4103/2278-0513.125814

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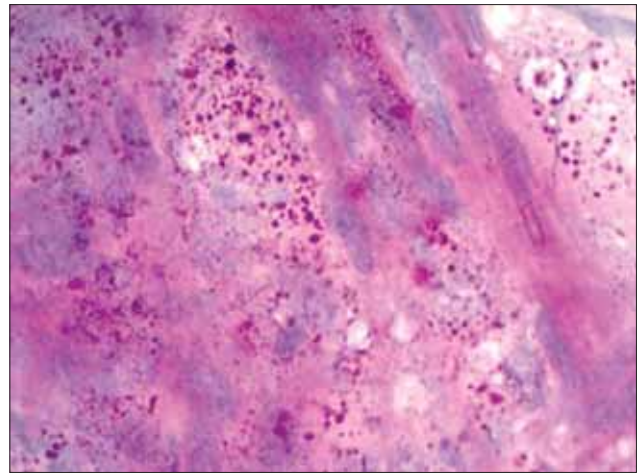


**Figure 1:** Large papillary clusters of pleomorphic tumor cells with high N/C ratios, micro-glandular pattern of growth, cytoplasmic and intra-nuclear vacuoles, and prominent nucleoli (Giemsa stain,  $\times 40$ )

Incidence of such extragonadal germ cell tumors in children is less than 1/million/year.<sup>[4]</sup> Sacrococcygeal YST can be external or internal, i.e., pre-sacral and intra-pelvic or intra-abdominal extension. Tumors which are external have a lower malignant potential than pre-sacral tumors that are always malignant.<sup>[5]</sup> For infants with a external sacrococcygeal mass, the differential diagnosis include meningomyelocele, chordoma, neurogenic tumors, lipoma, vestigial tail, and hemangioma.<sup>[5]</sup>

Sacrococcygeal YST of infants and children reflect the transformation of primordial cells that failed to migrate to their pre-destined location. These primordial germ cells gives rise to an undifferentiated germ cell line, which can undergo differentiation into embryonic or extrembryonic cells of yolksac, chorion, and allantoic cells.<sup>[3,6]</sup> Malignant transformation of these cells gives rise to tumors that reflect their embryonic features. Tumors of extra-embryonic cells have trophoblastic features as in choriocarcinoma or characteristics of yolk sac, endoderm, etc.

Afroz *et al.*<sup>[7]</sup> analyzed the cytomorphic features of YST of childhood. Cytologic examination showed a combination of morphologic patterns in their study. Characteristically, tumor cells were arranged in papillary groups, tight clusters, and well-formed acinar structures. Cells showed enlarged nuclei and moderate amount of cytoplasm, some of which displayed cytoplasmic vacuolation, displacing the nucleus eccentrically. They concluded that cytomorphic features of YST differed from that of other germ cell neoplasm's like embryonal carcinoma. Mishra *et al.*<sup>[8]</sup> found tight clusters and papillary fronds of cells associated with homogenous acellular eosinophilic globules in the FNA smears of ovarian yolks ac tumor. They concluded that a pre-operative diagnosis of this tumor by FNA is helpful in planning further diagnostic and therapeutic steps.



**Figure 2:** Increased amount of PAS-positive globules in the vicinity of these hyaline bands (PAS stain,  $\times 40$ )

Generally, two types of cells may be encountered in YST.<sup>[9,10]</sup> One cell (type A) has relatively well-defined borders and somewhat granular cytoplasm. Nuclei are round to slightly irregular and usually have well-developed nucleoli. In addition, many of these cells contain large "punched-out" clear vacuoles within the cytoplasm, corresponding to the deposits of intra-cytoplasmic glycogen in seminoma/dysgerminoma. The second cell (type B) usually forms syncytial clusters with indistinct cell borders. The cytoplasm is full of numerous vacuoles in all parts of the cytoplasm (hypervacuolated cells). The nature of these vacuoles is not known, but they are not related to glycogen. The relative numbers of type A and type B cells vary considerably from case to case. Generally, reticular and myxomatous patterns contain large numbers of hypervacuolated cells, whereas papillary, endodermal sinus, and solid patterns are usually dominated by type A cells. Further studies to correlate the various histologic and cytologic patterns are clearly needed. An important diagnostic feature in the fine needle aspiration biopsy (FNAB) smears from YST is the presence of eosinophilic inclusions within the cytoplasm. Globular or linear deposits of eosinophilic intercellular material may also be present in some cases, corresponding to the basement membrane-like material encountered in a histologic section. The background in aspiration smears from YST is usually mucoid. In addition, a few inflammatory cells, especially macrophages, may be present.

The presence of hyaline, PAS-positive material forming bands or connective tissue cores surrounded by tumor cells is not an infrequent finding in YST. There may be increased amount of PAS-positive globules in the vicinity of these hyaline bands, suggesting the possibility of their common origin. This feature was also appreciated in the PAS-stained smears of our case also.

To conclude, the cytomorphologic features of YST is characteristic and help in the early diagnosis and treatment even in their sacrococcygeal location.

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**Cite this article as:** Mardi K, Bakshi N. Cytodiagnosis of extragonadal pure yolk sac tumor of sacrococcygeal region. *Clin Cancer Investig J* 2014;3:105-7.

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