Yolk Sac Tumor of the Parotid Gland in a Child and its Differentials

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Yolk sac tumors (YST) (or endodermal sinus tumors) are rare neoplasms of germ cell origin that have been reported in gonadal sites (testis and ovary). Extragonadal YST are uncommon and are extremely rare in the extracranial head-and-neck regions. Here, we present a rare case of a 1-year-old male with isolated YST of the right parotid gland initially diagnosed as malignant epithelial neoplasm with possibilities of mucoepidermoid carcinoma and epithelial–myoepithelial carcinoma.

Keywords: Endodermal sinus tumor, extragonadal germ cell tumor, parotid gland, yolk sac tumor

Case Report

A 1-year-old male presented with gradually increasing right neck swelling for 2 months.

On examination, the lump measured 3 cm × 2 cm. It was well-defined, firm, immobile, and nontender. Overlying skin was unremarkable. There was no locoregional lymphadenopathy. An excisional biopsy was performed by a pediatric surgeon and sent for histopathological examination at an outside institute which yielded a diagnosis of “malignant epithelial neoplasm” with possibilities of mucoepidermoid carcinoma and epithelial–myoepithelial carcinoma. The patient was then referred to our tertiary care facility for definitive management, where the biopsy findings were reviewed, and further workup of the patient was done.

Magnetic resonance imaging of the head and neck revealed a T1 intense and T2 hyperintense soft-tissue lesion in the right parotid space and right parapharyngeal space measuring 3.4 cm × 2.3 cm × 1.4 cm causing a mass effect over the tongue and oropharyngeal air column and displacing it toward the left side. The lesion was posteriorly abutting the carotid sheath structures, displacing them posteriorly and reaching up to the subcutaneous plane laterally, causing contour bulge of the skin (suggestive of neoplastic etiology) [Figure 1a].

Laboratory screen for alpha-fetoprotein (AFP) showed an initial level of more than 1,60,000 ng/ml.

Histopathological examination

Hematoxylin and eosin sections from the tumor showed malignant tumor arranged in sheets, anastomosing trabeculae and glandular formation showing a

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mixture of endodermal sinus pattern and reticular pattern [Figure 2a]. The endodermal sinus pattern showed labyrinthine anastomosing glands and papillae lined by cuboidal to columnar cells with moderate nuclear pleomorphism, vesicular chromatin, and moderate amount of amphophilic cytoplasm. Papillary structures lined by cuboidal to columnar cells with fibrovascular cores were seen projecting into cystic spaces covered by cuboidal tumor cells (Schiller–Duval bodies) [Figure 2b]. A fair number of eosinophilic hyaline globules were seen. The reticular pattern showed loose meshwork of microcystic spaces lined by cuboidal cells having atypical hyperchromatic nuclei and clear to amphophilic cytoplasm.

On immunohistochemical examination, tumor cells showed positivity for AFP [Figure 1b], CD117, and CK; vimentin, CD10, and CD30 were negative; and Ki67 proliferation index was positive in 70% of the tumor cells.

The definitive diagnosis of YST was signed out.

**Patient treatment**

The postoperative chemotherapy regimen in our patient comprised six cycles of bleomycin, etoposide, and cisplatin which was effective in achieving disease control, as evidenced by the return of AFP levels to baseline. The recovery period was uneventful, and he is currently 3 years post treatment with no residual disease and meeting his milestones as expected for his age.

**Discussion**

YST is also known as endodermal sinus tumor due to its similarity to the endodermal sinus of rat yolk sac [3]. YST comprises cells that have undergone malignant transformation along extraembryonic tissue lines, according to histology. The etiogenesis of GCT is explained by two principal theories. Malignant transformation occurs within primordial germ cells during their migration along the urogenital ridge, according to the “germ cell theory.” This explains the midline tendency, but it is difficult to account for a cephalad tumor that is located laterally. According to the “embryonic cell theory,” the totipotent embryonic cells undergo malignant degeneration into YST.[2]

YST is associated with higher levels of serum AFP, an α1-globulin with a half-life of 5 days, which is used to detect the presence, recurrence, or metastasis of the tumor.[4] The AFP levels return to baseline post treatment.

Some of the common reported histological patterns of yolk sac tumor (YST) include reticular or microcystic, perivascular or endodermal sinus pattern (showing characteristic Schiller–Duval bodies), macrocystic, solid, alveolar or glandular, polyvesicular vitelline, myxomatous, papillary, hepatoid, and intestinal (primitive endodermal) pattern. It reveals hyaline PAS-positive intra- and extracellular globules and AFP immunoreactivity of the tumor cells.[1]

Primary EGCT must be differentiated from the lesions occurring more commonly at this site which include...
infantile hemangioma, lymphangioma, dermoid cyst, rhabdomyosarcoma, pleomorphic adenoma, and lymphomas. YST is locally aggressive with a proclivity for recurrence. It can spread to regional lymph nodes, liver, lung, brain, and in rare cases, bone. Treatment includes surgical resection, followed by chemotherapy, radiotherapy, or both. The overall 2-year survival rate is reported as 70% (100% for testicular tumors, 60% for sacrococcygeal tumors). Chemotherapy improves the patient’s prognosis by 20%–50%. With the exception of testicular primaries, all cases of extragonadal YST should receive adjuvant chemotherapy.

In our case, the histomorphology along with immunoreactivity of the tumor cells for anti-AFP helped us exclude all the differentials and reach the definitive diagnosis of YST. The clinicoradiological findings of a localized disease ruled out the presence of metastases.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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