

CASE REPORT

Primary Orbital Yolk Sac Tumor

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ABSTRACT:**PURPOSE:**

To report unusual case of a girl with primary orbital yolk sac tumor.

METHODS:

A 2.3 –year-old girl presented with left eye proptosis and supraorbital swelling. CT revealed evidence of left sided proptosis with soft tissue mass about 2.7x2.2cm in the posterior part of optic nerve complex and causing destruction of the medial wall of Lt. orbit and extended into Lt. nasal cavity and also extended few mm intra-cranially with enhancement in post contrast study, signs of new growth? optic nerve glioma? Neurofibroma?

RESULTS:

The microscopical examination showed proliferation of cuboidal cells with slight pleomorphism in a reticulated fashion, frequent Schiller-Duval bodies, and intracytoplasmic and extracellular hyaline droplets, the neoplastic cells express cytoplasmic alpha fetoprotein (AFP). Features consistent with yolk sac tumor.

CONCLUSION:

Among common orbital masses in a childhood, yolk sac tumor must also be considered in the differential diagnosis of orbital mass with rapidly increasing size and adjacent bony destruction.

KEYWORDS: yolk sac tumor, AFP.

INTRODUCTION:

Yolk sac tumor is a rare malignant germ cell neoplasm. Primordial germ cell migrate from yolk sac to genital ridge. Most tumors are therefore gonadal, usually occurs in infants testes and ovaries. While aberrant migration may occur. The orbit is an unusual location of the primary development of this neoplasm.^(1,2,3) In the orbit they manifest by unilateral proptosis with or without restricted ocular motility, loss of vision, papilloedema, and congested retinal blood vessels.⁽³⁾ α fetoprotein had been used as a diagnostic and therapeutic follow up marker for gonadal yolk sac tumors.^(4,5) We report a case of primary pure yolk sac tumor of the orbit in a girl, discussing the histopathological and immunohistochemical finding.

CASE REPORT:

A 2.3 –year-old girl presented with left eye proptosis and supraorbital swelling, there was no significant previous problems, her developmental history was uneventful.

Left orbital CT revealed evidence of left sided proptosis with soft tissue mass about 2.7x2.2cm in the posterior part of optic nerve complex and causing destruction of the medial wall of Lt. orbit and extended into Lt. nasal cavity and also extended few mm intra-cranially with enhancement in post contrast study, signs of new growth? optic nerve glioma? Neurofibroma?

Two weeks later, the eye was significantly exposed, debulking orbital mass biopsy done.

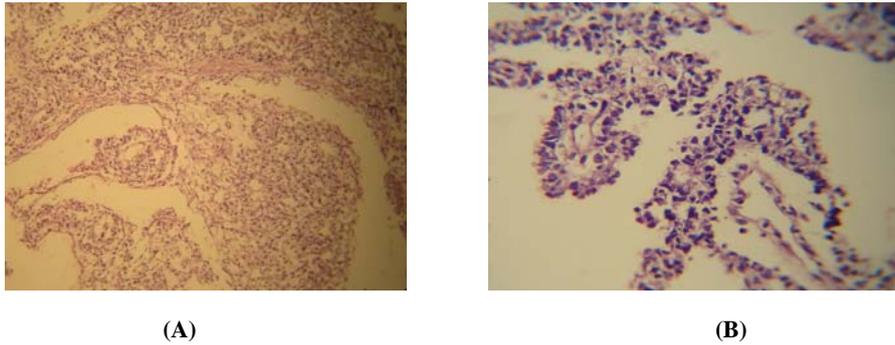
Histopathological examination showed irregular contoured glands and papillae covered by highly pleomorphic atypical cells surrounded by desmoplastic and inflammatory tissue reaction. the cells were stained immunohistochemically positive for CK and S-100 protein .weakly positive for NSE, while negative for desmine, actin, GFAP, and EMA, the overall picture in favour of metastatic poorly differentiated malignant tumour of the orbit.

Abdominal ultrasound and skeletal survey showed no significant finding, renal function tests, liver function tests, and bone marrow aspirate all were normal, S.LDH 306 u/l, Hb 11/5g/dl, WBCc 11.500/cmm, L 37%, N 56%, plt. 407.000/cmm.

Two weeks later, pathology slides and paraffin blocks referred to us at teaching Labs. Baghdad medical city complex for slide review consultation. The microscopical examination showed proliferation of cuboidal cells with slight pleomorphism in a reticulated fashion, frequent pseudo-papillary structures with central vessels (Schiller-Duval bodies), scattered mitotic figures, and intracytoplasmic and extracellular hyaline droplets, the neoplastic cells express cytoplasmic alpha fetoprotein (AFP). The histologic and immunohistochemical features consistent with yolk sac tumor. The patient started combined chemotherapy.

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**Fig. 1: (A) Loose reticular pattern and rounded papillary structures with central capillary are characteristic (original magnification x 200).
Fig. (B) :Also characteristic, Schiller-Duval bodies composed of cuboidal cell layer surrounding a fibrovascular core (original magnification x 400).**

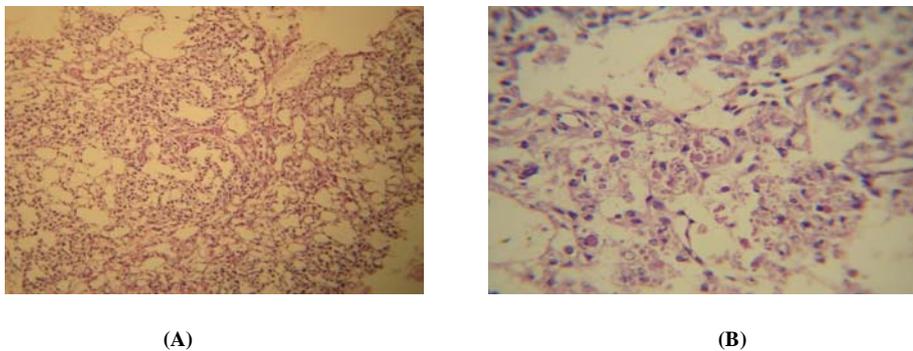


Fig. 2: (A) Demonstrating a microcystic area (original magnification x 200).. (B) Intracytoplasmic and extracellular PAS-positive hyaline droplets are nearly always present (original magnification x 400).

DISCUSSION:

Few patients with extragonadal location of yolk sac tumor described. Extragonadal germ cell tumor of the head and neck comprise only 5% of benign and malignant germ cell tumors, with yolk sac tumor is the most common histopathological subtype⁽⁶⁾ The presence of yolk sac tumor in extragonadal sites is difficult to explain. The theories include that, ectopic germ cells resulting from migration abnormalities and misplacement during embryogenesis.^(7,8)

Since the first published report of yolk sac tumor of the orbit by Katz et al⁽⁹⁾ in 1982, 13 orbital cases have been reported⁽⁶⁾. The patients presented between 3 months and 4 years of age, with eight of these patients had concomitant intracranial, nasopharyngeal, maxillary sinus, or pterygopalatine fossa extension.⁽⁶⁾

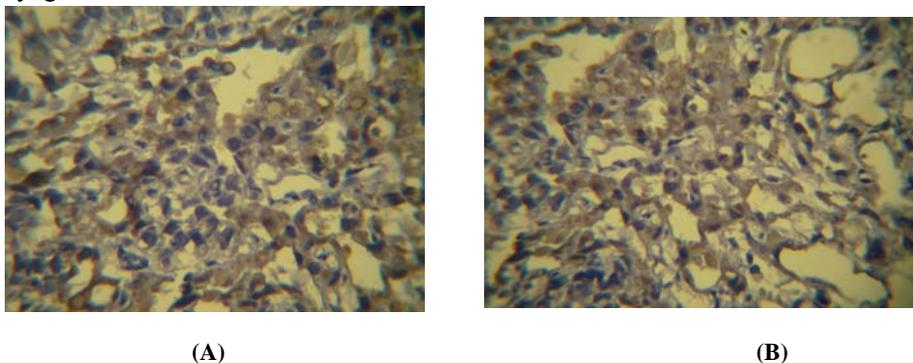


Fig. 3: (A & B) The tumor cells showed positive cytoplasmic immunostaining for AFP (original magnification x 400).

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Microscopically, yolk sac tumor is rather typical, featured embryonic structures, intracytoplasmic and extracellular PAS-positive hyaline globules and pathognomonic Schiller-Duval bodies,⁽⁷⁾ and AFP immunoreactivity⁽⁸⁾.

Yolk sac tumor may present as single isolated histological entity or as a mixed germ cell tumor.⁽¹⁰⁾ The mixed form seems to be the most frequent in YST in the nasal cavity and in adults.⁽¹¹⁾

Yolk sac tumor is extremely malignant, tend to recur locally⁽¹²⁾ and may present with early metastases in 50% of the cases.⁽¹³⁾ Kusumakumari et al⁽¹⁰⁾ suggested that extragonadal germ cell tumors have a worse prognosis than gonadal tumors. Although long-term follow up data are available in only a few cases, those tumors originating in the orbit may have a better prognosis compared with those originating at other extragonadal sites^(8,14) as the result of early recognition.^(2,7)

Radiotherapy was not effective for yolk sac tumors; adjunctive combined chemotherapy after aggressive surgical excision provided a relatively long disease-free survival.^(6,8)

Among common orbital masses in a childhood including rhabdomyosarcoma, optic glioma, metastatic neuroblastoma, lymphoma, teratoma, ruptured dermoid, and hemangimas,^(1,6,7,14) yolk sac tumor must also be considered in the differential diagnosis of orbital mass with rapidly increasing size and adjacent bony destruction.

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