Case Report

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Pathology Section

# Primary Nasopharngeal Yolk Sac Tumor: A Case Report

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## ABSTRACT

Yolk sac tumour also known as primitive endodermal tumour is the most common malignant germ cell tumour (GCT) in the paediatric age group. Most common sites of involvement are ovaries and testes, but rarely can occur in the extragonadal sites. In the head and neck region, yolk sac tumours have been reported in the nasopharynx, sinonasal tract, orbit, ear and parotid gland. Nasopharynx is an uncommon site for yolk sac tumour and very few cases of nasopharngeal pure yolk sac tumour have been reported so far. Yolk sac tumours are highly malignant and have a poor prognosis. This is a case of pure GCT in a three-year-old female child who presented with a rapidly growing nasopharyngeal mass. Histopathological examination followed by immunohistochemistry and serum AFP study clinched the diagnosis of yolk sac tumour. The tumour responded well to chemotherapy as evidenced by decrease in serum AFP levels.

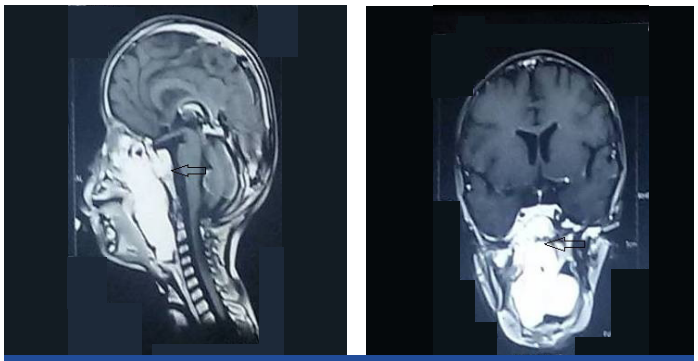
**Keywords:** Alpha- fetoprotein, Germ cell tumours, Schiller –Duval bodies

## CASE REPORT

A three-year-old female child presented with history of difficulty in breathing and snoring, which disturbed her sleep for two weeks. There was history of loss of appetite and fever of one month duration.

Clinical examination revealed a rapidly growing mass in the nasopharynx, pushing the soft palate. Airflow was absent in both nostrils.

All the routine investigations including complete blood count and peripheral smear were done and found to be normal. CT scan and MRI were done which showed large destructive lesion with increased vascularity in the nasopharynx extending upto the skull base. The provisional diagnosis was rhabdomyosarcoma and chordoma based on the clinical and radiological findings [Table/Fig-1a&b].



**[Table/Fig-1a]:** MRI-sagittal section. **[Table/Fig-1b]:** MRI-coronal section show a large destructive lesion in the nasopharynx extending upto the skull base.

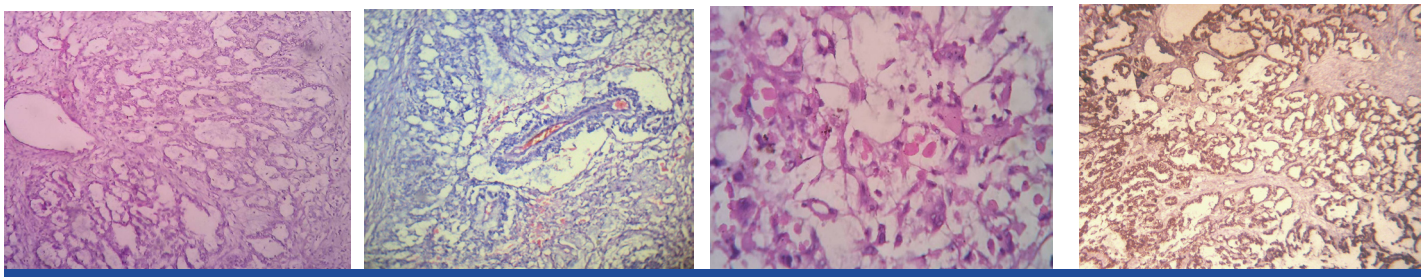
Excision biopsy was done and sent for histopathological examination. The specimen was received in multiple pieces together measuring 6x5 x 2.5 cm. Cut surface of the tumour was grey white and firm. Multiple samples were submitted for histopathological tissue processing. Multiple sections studied showed a cellular tumour composed of tumour cells arranged in solid sheets, glandular and microcystic patterns [Table/Fig-2]. Many Schiller-Duval bodies were noted [Table/Fig-3]. Intracytoplasmic and extracytoplasmic hyaline globules were also noted [Table/Fig-4]. No other germ cell elements were found.

The possibility of gonadal germ cell tumour was ruled out by imaging studies. Hence, a diagnosis of primary extragonadal pure yolk sac tumour was made.

Immunohistochemical staining with alpha-feto protein showed strong cytoplasmic positivity [Table/Fig-5]. Immunostaining with CD30 was negative, thereby excluding the co-existence of embryonal carcinoma. The results were consistent with the diagnosis of pure yolk sac tumour.

Serum hormonal assays were done for the patient. Serum alpha-fetoprotein was found to be 1098 ng/ml (normal value < 6 ng/ml). Serum lactate deydrogenase was also found to be elevated.

Patient was started on chemotherapy with BEP (Bleomycin, etoposide and cisplatin) regimen. Follow-up was done with serum hormonal assays. The tumour showed rapid response to chemotherapy, as indicated by decrease in the serum alpha-fetoprotein levels from 1098 ng/ml to 472ng/ml.



**[Table/Fig-2]:** Yolk sac tumor-Tumor cells arranged in microcystic and macrocystic pattern (10X). **[Table/Fig-3]:** Yolk sac tumor with Schiller-Duval body(10X) showing a central blood vessel surrounded by single layer of malignant epithelial cells. **[Table/Fig-4]:** Intracellular and extracellular hyaline globules in yolk sac tumor(40X).**[Table/Fig-5]:** Immunostaining with alpha-fetoprotein showing strong, diffuse cytoplasmic positivity (10X).

## DISCUSSION

Yolk sac tumours also known as primitive endodermal tumour or endodermal sinus tumour constitute 20% of malignant GCTs [1]. Yolk sac tumours are the most common malignant GCTs occurring in infants and children less than four years of age. These tumours represent 3-5% of paediatric malignancies [2,3].

As with other GCTs, yolk sac tumours are common in ovaries and testes. Extragonadal GCTs are relatively uncommon representing 1 to 5% of all GCTs [3]. Few cases have been reported in the mediastinum, liver, lung, brain, vagina, nasopharynx, sinonasal tract, orbit, ear and parotid gland [4-6].

In the head and neck region, yolk sac tumours have been reported with an incidence of 5% [7]. Yolk sac tumours are usually seen as a component with other GCTs like teratoma and embryonal cell carcinoma. Teratomas and yolk sac tumours are the common GCTs occurring in the sinonasal tract. Pure yolk sac tumour occurring in the nasopharaynx is rare and only very few cases have been reported so far in the literature. Mamoon et al., has reported a yolk sac tumour arising in a mature teratoma in the parapharngeal space [8].

Yolk sac tumours are grey to yellow, solid and cystic tumours, with areas of haemorrhage and necrosis. Yolk sac tumours are highly malignant tumours characterised by tumour cells arranged in various patterns like reticular or microcystic pattern, endodermal sinus pattern, papillary pattern, alveolar-glandular pattern, macrocystic pattern and in solid sheets [9]. The tumour cells are large with clear cytoplasm and vesicular nuclei and prominent nucleoli. Mitotic figures are numerous.

Schiller-Duval bodies are diagnostic of yolk sac tumours but their absence does not rule out yolk sac tumour if other histological features are present [9]. These tumours show intracellular and extracellular PAS positive hyaline material.

Serum AFP levels are elevated in patients with yolk sac tumours. Assessment of serum AFP levels is useful for diagnosing yolk sac tumours and also in monitoring response to therapy and prognosis [10]. Serum AFP levels decrease after treatment in 2 to 3 weeks. Immunohistochemistry is useful in confirming the diagnosis of yolk sac tumour and also in excluding other GCTs. Tumour cells shows strong immunoreactivity with AFP antigen, LMWCK, Glypican-3 and SALL4 [11,12].

In the present case, elevated serum AFP levels and positive cytoplasmic immunoreactivity of tumour cells with alpha-feto protein antigen confirmed the diagnosis.

Nasopharyngeal yolk sac tumours should be differentiated from chordoma based on the histological features and immunohistochemistry, as chordomas often skull based, can also rarely present as nasopharngeal mass. Kataria et al., has reported a case of skull based chordoma presenting as nasopharyngeal mass [13]. It is important to diagnose pure yolk sac tumour because these tumours are aggressive and require surgical excision and adjuvant cisplatin based chemotherapy [14].

## CONCLUSION

A diagnosis of yolk sac tumour should be thought of in the differential diagnosis of nasopharyngeal mass, especially in children, as early diagnosis has a significant influence on the clinical outcome and survival of the patients.

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