



## Case Report

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# Embryonic Yolk Sac Tumor of Mandible: A Rare Entity with Clinicopathological Features and Literature Review

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## Abstract

The present case report is about 1 and half year old boy presented with the complaint of aggressively growing swelling on right side of the face. This large tumour was extending from body of the mandible to the ramus of ipsilateral side of the face. Due to difficulty in mouth opening, intraoral examination could not be possible. Considering the aggressive behavior of the tumour, immediate surgery was planned and tumour was excised completely and excised mass had been processed for histopathological examination. Based on clinical, radiographic, histopathological and immunohistochemical findings, the lesion has been diagnosed as embryonic yolk sac tumour. The patient was referred to MINAR for post-operative radiotherapy.

## Introduction

Yolk sac tumor (YST) is a rare malignant tumor of germ cells that line the yolk sac of the embryo. These tumors usually grow in gonads while some extra-gonadal counterparts typically target the mediastinum, endometrium, sacrococcygeal areas, cervix and vagina [1]. This neoplasm accounts for 3.5% of all childhood cancers and its usual age of presentation is 15 to 19 [2]. The incidence of malignant yolk sac tumor is less than 1 per million per year while male to female ratio is about 1:2.5 [3].

Gonadal yolk sac tumors that are found in the ovaries or testes usually present as a painless, firm swelling that grows rapidly to the noticeable size. On the other hand extra gonadal yolk sac tumors present as a firm swelling with intermittent pain and fever [4]. Among extra gonadal sites, certain cases of yolk sac tumor have been reported in head and neck area [5].

The exact pathogenesis of YST is still not clear. However, most of the studies propose that these tumors arise from malignant

transformation of misplaced primordial germ cells. During the 4-6th week of embryogenesis primordial germ cells migrate along the dorsal mesentery to the genital ridge. Certain germ cells may not complete this journey and retain along the dorsal wall of embryo at areas near the midline [6]. Some extragonadal tumors may arise from totipotential cells that are scattered throughout the body and have the potential for additional growth when suitably stimulated [7].

There are numerous diagnostic modalities which can be used, such as US, CT, magnetic resonance imaging (MRI) and histopathological examination of tumor specimen. Another important diagnostic marker is alpha-fetoprotein (AFP), whose level was found to be elevated in > 90% of yolk sac tumors [4]. Cancer cells secrete human chorionic gonado-protein (HCG) and alpha-fetoprotein (AFP) and their levels are reported to be elevated in these patients [8]. These substances can be used as tumor biomarker for diagnostic and follow up cases.

The best treatment option for YST is surgery for eliminating the primary tumor without severe morbidity [9]. Surgical procedures usually involve the debulking surgery of tumor followed by adjuvant chemotherapy. The national comprehensive cancer

network recommend 3-4 cycles of chemotherapy of regimen of BEP (bleomycin, etoposide and cisplatin) [10]. BEP chemotherapeutic treatment is considered to be a gold standard first line protocol in all stages of germ cell tumors [11].

Both gonadal and extragonadal yolk sac tumors exhibit poor prognosis. Because of their rapid spread and metastasis, clinical presentation usually obscures the actual invasion. Minimally invasive surgery, fertility sparing surgery techniques and adjuvant chemotherapy has been proved to have better prognosis [9].

### Case Presentation

A three years old boy was presented to the Department of Oral and Maxillofacial Surgery, Bakhtawar Amin Dental Hospital Multan with aggressively growing mass with grotesque swelling on right side of the mandible with a history of one and half month only. The parents of the patients noticed a small swelling one and half month back. Initially, they ignored the swelling because it was painless and not causing any discomfort during eating. But the swelling attained a large size by growing aggressively in very short duration with gross facial disfigurement. On clinical examination, the swelling was firm and non tender. Patient's medical history was non-significant (Figure 1a).



Figure 1a

Intraoral examination could not be performed due to limited mouth opening and non-cooperative behavior of the child. The tumour mass was obstructing the oral cavity and a part of tumour

was protruding from the mouth opening. Blood clots were visible over the protruding front of the tumour (Figure 1b).



**Figure 1b**

**Figure 1:** An aggressively growing mass involving mid part of body of mandible, extending towards ramus.

On general physical examination, patients appeared weak, pale with signs of anemia by without any signs of jaundice, clubbing of nails and cyanosis. The pulse rate (85 BPM), respiratory rate (20/min) and blood pressure (130/80 mmHg) were quite normal in range. There was no abdominal distension or disturbed vision. Radiological and other laboratory investigations were quite normal in range.

On gross examination the excised specimen size was measured as 7.5x6.5x4. Actual tumour bulk was composed of a large necrotic mass and upon removal of this necrotic area, the remaining tumour bulk was measured as 7.5x3.5x3.5cm.

Tumor was 0.1cm from anterior margin, 0.3cm from posterior margin, 0.3cm from buccal margin and 0.2cm from lingual margin. On serial slicing, cut surface of tumor was partially homogenous and myxoid and partially necrotic and hemorrhagic. Grossly tumor is involving the bone. On histopathology, hematoxylin and eosin stained sections showed a neoplasm composed of loose network of anastomosing channels forming cystic cavities lined by tumor cells with clear glycogenated cytoplasm. Few Schiller Duval bodies were also seen. All the margins of excised mass other than anterior margin were free of tumour cells. On immunohistochemical staining, tumour tissue shows positivity for SALL4, cytokeratins and Glypican-3 and negativity for Oct-3 and Oct-4.

Based on clinical, radiographic, histopathological and immunohistochemical findings, the lesion has been diagnosed as

embryonic yolk sac tumour. The patient was referred to MINAR for post-operative radiotherapy.

### Discussion

Yolk sac tumors (YSTs) are a type of malignant germ cell tumor that usually grow in the gonads. They are difficult to recognize at other sites outside the gonads, and very few cases have been reported involving the head and neck region (Chen et al. 2017). However, between 10 and 15% of YST occur in the midline structures of the mediastinum, retroperitoneum and sacrococcygeal areas. Certain cases have been reported in other location such as in the pineal region and head and neck [3-5].

In the rare case discussed in the present study the diagnosis was established by histological and immunohistochemical examinations. The major pathologic characteristics of YSTs include a reticular pattern, a solid pattern, a hepatoid pattern with hyaline globules, a festoon pattern, enteric differentiation, Schiller-Duval bodies, the presence of a granulomatous tissue reaction and a polyvesicular vitelline pattern [6, 7]. Schiller-Duval bodies have been reported as a common feature to extragonadal YSTs, as observed in large quantities in a previous study [8]. Typical Schiller-Duval bodies comprise cuboidal or low columnar-shaped tumor cells surrounding the capillaries of a thin-walled sinus [9, 10]. Immunostaining plays very important role in diagnosing this tumour. Staining for alphafetoprotein by immunoperoxidase techniques is usually positive in this tumour [11] (Figures 2-9).



**Figure 2:** Aggressive behavior of tumour.



**Figure 3:** Aggressive lesion Compromising airway.



**Figure 4:** Per-op.

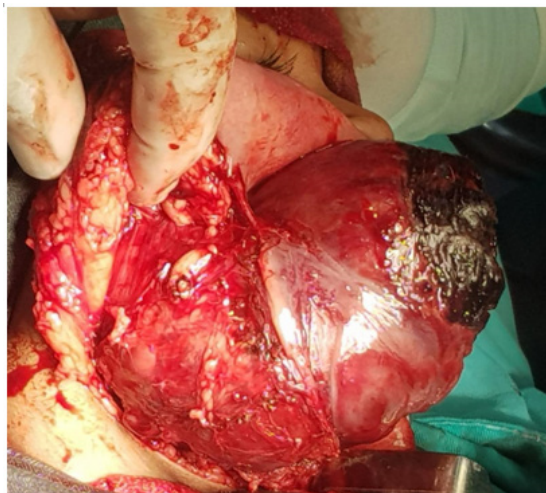


Figure 5: Per-op.



Figure 6: Post-op.



Figure 7: Post-op.

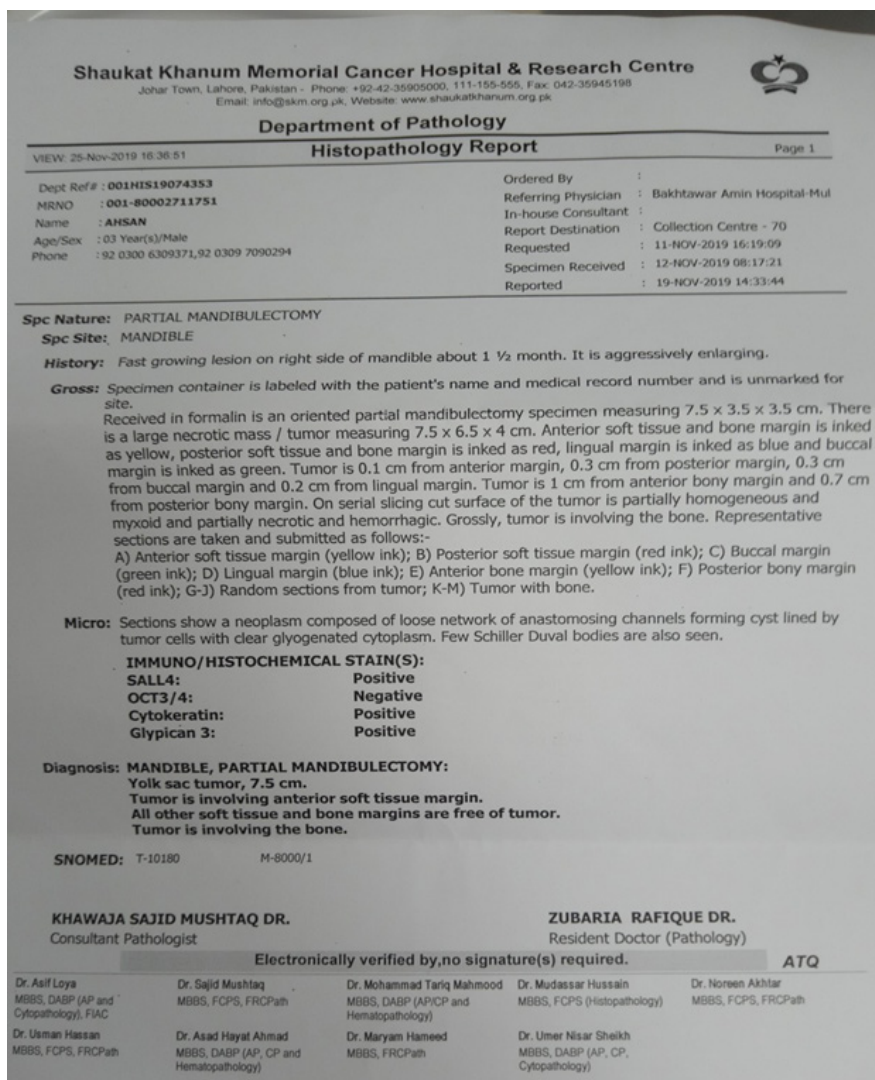


Figure 8: Biopsy report.



Figure 9: Post-op at 3 Months.

## Acknowledgement

None.

## Conflict of Interest

No conflict of interest.

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