

Extragenadal germ cell tumour: Primary yolk sac tumour of the maxillary sinus

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ABSTRACT

Epistaxis in children is a common presentation, mainly due to local causes. The most common causes are due to inflammatory disease, infections and trauma. A small percentage of epistaxis in children can be caused by tumour whereby the differential diagnosis includes juvenile nasoangiofibroma and rhabdomyosarcoma. We report the case of a 2-year-old child who had presented with intermittent epistaxis that was later diagnosed to be secondary to a yolk sac tumour of the maxillary sinus.

Keywords: epistaxis, yolk sac tumour, germ cell tumour

INTRODUCTION

Yolk sac tumour (YST), an extragenadal germ cell tumour (GCT) is a subtype of neoplasm that arises from primordial germ cells mainly from the gonads. It has been reported in a number of sites which includes the sacrococcygeal area, retroperitoneum, mediastinum, neck and pineal region. Involvement of the head and neck region is considered rare, and accounts for only 5% of all benign and malignant GCTs. ¹ Although the causes of YST are still unknown, the classic theory suggests that GCTs in these areas arise from local transformation of misplaced primordial germ cells that occur during embryogenesis. ²

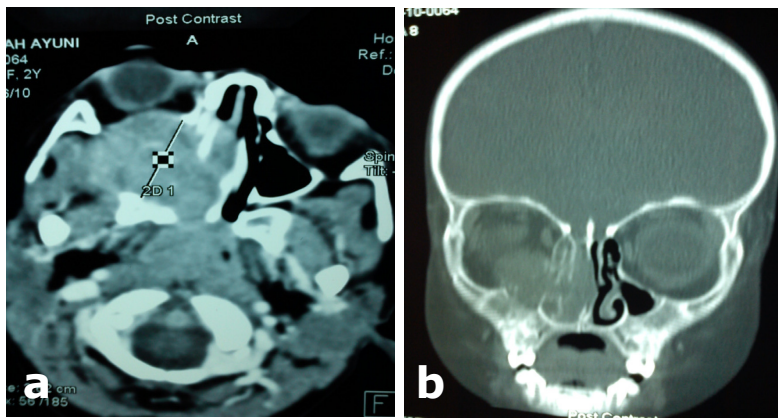
Majority of GCTs in children are gonadal in origin and benign, presenting either as a mature or immature teratoma. YST may occur alone or in combination with a teratoma. We report a case of isolated primary maxillary sinus YST in a two year old child.

CASE REPORT

A two-year-old Malay girl was referred to the Department of Otorhinolaryngology, Hospital Kuala Lumpur with a five days history of right sided epistaxis. The epistaxis was sporadic in nature, lasting for one to two minutes and would spontaneously stop. There was no history of trauma to the nose and the parent denied any history of previous nose bleeds or other nasal problems.

On general examination, we noted

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Figs. 1: a) Axial computed tomography view showing right maxillary sinus tumour, and b) A sagittal view showing the tumour infiltrating into the surrounding structures.

there was right eye proptosis with ophthalmologia. A Cold Spatula test revealed reduced patency of the nostrils on the right side. However, anterior rhinoscopy did not show any nasal mass in the nasal cavity.

A computed tomography of paranasal sinus (CT-PNS) revealed an enhanced lesion with its epicentre in the right maxillary region, extending to the ethmoid and right middle cranial fossa (Figures 1a and 1b).

The patient underwent examination under general anaesthesia which revealed a mass lateral to the right medial turbinate with the appearance of granulamatus tissue. Biopsies were taken with only minimal bleeding. Histopathology revealed a YST (Figure 2).

Serum alpha feto-protein (AFP) was remarkably increased (>5,000 U/L) and beta-chorionic gonadotropin (Bhcg) was within normal limit. She was diagnosed as right maxillary stage YST.

She was referred to the paediatric oncologist and started on chemotherapy

(carboplatin, etoposide and bleomycin). After completing six cycles of chemotherapy, the serum AFP level at 9 and 10 months declined to within the normal range: 3.75 U/L and 1.26 U/L respectively. A repeat CT-PNS 10 months post-chemotherapy showed significant shrinkage of the soft tissue mass in the right maxillary sinus.

DISCUSSION

GCT consists of a varied group of benign and malignant neoplasm occurring in the perinatal period. It accounts for 2% of human malignancies, but GCT is the most common malignant tumours in 15-35 years of age. GCT can

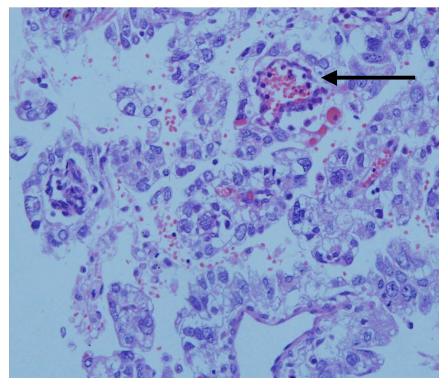


Fig. 2: Shiller-Duval body (arrow) on H&E stain. It consists of central vessel surrounded by tumour cells.

be divided into two major histologic types: seminoma and non-seminomatous GCT (NSGCT), with seminomas comprising about 40%–50% of GCTs.³

YST, also known as endodermal sinus tumour is a non-seminomatous GCT subtype. It is the leading malignant GCT of the perinatal period and throughout childhood.⁴ The International Germ Cell Consensus Classification (IGCCC) published in 1997 has become the gold standard used in clinical practice and research regarding GCT.⁵ Based on this, independent variables are used as a prognostic factors. These include histology and site of the primary tumour, the degree of pre-treatment tumour marker elevation (AFP, human chorionic gonadotrophin and lactate dehydrogenase) and the presence of non-pulmonary visceral metastases. A non-seminomatous germ cell tumour with non pulmonary visceral metastases and elevated level of tumour marker has a poor prognosis compared to other subtypes with normal tumour marker.

YST produces AFP and hence this is an important tumour marker for diagnosis and follow-up. Reduction of the serum levels correlates with response to treatment.⁶ AFP is a homologue of albumin, and is thought to act as a carrier protein in the foetus. During gestation, AFP is initially produced by the yolk sac and later by the foetal liver.⁷ After birth, circulating concentrations decrease with a half life of five days, falling to adult levels at 8-10 months of age.⁸ In adult, AFP is a sensitive marker for NSGCT.⁹

In general, the cytoarchitecture of a YST sac tumour shows flat to cuboidal cells

with vacuolated cytoplasm and variable atypia cells, with occasional single lines of tumour cells around blood vessels forming characteristic structures named Schiller-Duval bodies.¹⁰ In addition, some tumours demonstrate intracytoplasmic hyaline globules and/or aggregates of basement membrane-type material. Immunohistochemically, the tumour cells are positive for low molecular-weight cytokeratins and are usually positive for AFP with occasional positivity for Placental Alkaline phosphatase.

Germ cell tumours present differently depending on the age of patient, size, location, histologic types and the presence of metastases.² Its management varies with histologic aggressiveness and heterogeneous locations, and may include surgical exenteration in combination with various chemotherapy regimens and, occasionally, radiotherapy.

Many authors advocate surgical exenteration followed by adjuvant chemotherapy for YST in sites such as the ovary, vulva, vagina, liver, and ear.¹¹ Currently trans-nasal endoscopic surgery has been revolutionised and expands to include the treatment of selected cases of malignant tumour. Endoscopic sinus surgery decreases hospitalisation time, morbidity and mortality as compared to radical exenteration surgery. However, this advancement has been challenged by critics by who believe endoscopic surgery does not adhere to the principle of oncologic surgery.¹² In our case, we decided to refer the patient to the paediatric oncologist for chemotherapy.

YST is extremely malignant, and tends to recur locally. In view of the aggres-

aggressiveness and low survival rate of these tumours, extragonadal GCT have a worse prognosis than gonadal tumours.² Despite its poor prognosis, advances in chemotherapy drugs and diagnostic methods have improved survival rates.¹

Our patient responded towards chemotherapy with evidence of drastic decreased in AFP level post treatment. A repeat CT shows shrinkage of soft tissue mass occupies right maxillary sinus. We did not subject the patient to sinus surgery as she responded to treatment. She is being monitored at follow-up visits with serial CT PNS and tumour marker.

In conclusion, YST in the sinonasal region is a rare type of extragonadal GCT nevertheless, it should be considered in the differential diagnosis of any sinonasal tumour.

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