Case Report

# Malignant Sacrococcygeal Teratoma with Yolk Sac Differentiation: A Rare Case Report

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## Abstract

Sacrococcygeal teratomas (SCTs) are the most common germ cell tumours in neonates and infants, with malignant transformation often observed in postnatal cases. Yolk sac tumour (YST) is the most frequent malignant germ cell component seen in SCTs. We report a case of a 3-month-old infant presenting with a Type II SCT, showing histopathological features of yolk sac tumour differentiation.

*Keywords:* Sacrococcygeal teratoma; Yolk sac tumour; Neonate.

## Introduction

Malignant sacrococcygeal teratoma in children and new-borns can develop into yolk sac (endodermal sinus) [2]. However, during development, some of these cells may not complete their migration and remain along the dorsal midline of the embryo. These primordial germ cells then form an undifferentiated germ cell line, which can differentiate into embryonic somatic cells or extra-embryonic cells such as those of the yolk sac, chorion, and allantois. When these cells undergo malignant transformation, they can develop into tumours that mirror their embryonic origins [3]. Accurate histological diagnosis supported by immunohistochemistry (IHC) and radiological imaging is crucial for management and prognostication.

## Case Report

The parents of this young baby gave the history of appearance of painless swelling over the sacral region with gradual increase in size over a 3-4 months duration. The child was taken to the nearest doctor, where she got admitted in the Pediatric Surgery unit. Her Contrast-Enhanced CT (CECT) pelvis revealed a huge (solid) sacrococcygeal mass with foci of calcifications and intrapelvic extensions. CE MRI of the pelvis also confirmed it as a type II sacrococcygeal teratoma

(Altman classification). Her x-ray chest was clear and serum AFP was markedly (6300 IU) raised. The tumour was removed in piecemeal and sent for histopathology. No serious complications occurred during surgery and we followed up with the patient after two months; her recovery was excellent.

Gross examination of the tumor mass shows an irregular piece of tissue measuring 8.5cm x 6cm x 2cm with an attached skin flap measuring 7cm x 6cm. The cut surface is mostly solid, whitish, measuring 7cm x 5.5cm x 2cm with some areas of cystic degeneration.

Microscopically, along with the teratomatous component, atypical germ cells were seen arranged in microcystic, papillary, and glandular patterns. A cystic pattern was also noted at places. IHC for AFP, CK7, EMA, SALL-4, and Glypican-3 were done. AFP, SALL-4, Glypican-3 & EMA were positive, while CK7 came out to be negative. Based on histopathological as well as IHC findings, the lesion was diagnosed as malignant sacrococcygeal teratoma with yolk sac differentiation.

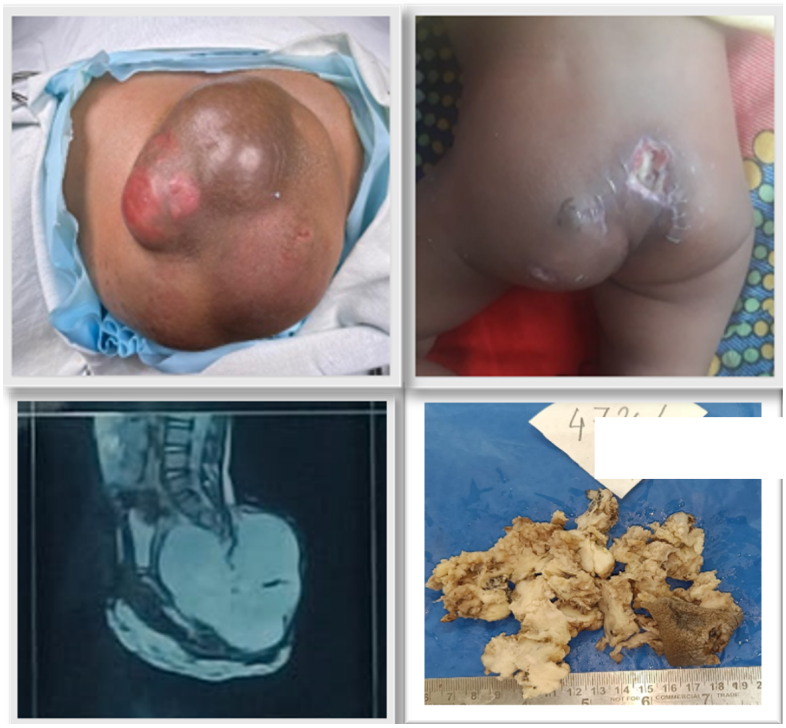

**Figure 1:** A: Clinical presentation of sacrococcygeal mass (Pre-op). B: Clinical presentation of sacrococcygeal mass (post-op). C: CEMRI appearance of MSCT. D: Gross appearance of MSCT.

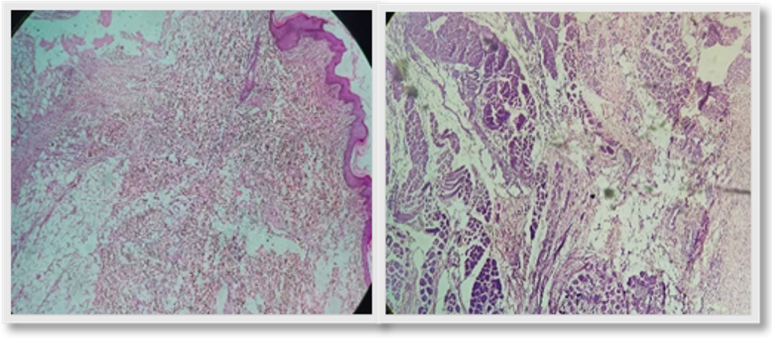

**Figure 2:** A: Showing teratomatous component (H&E, 4X). B: Showing mesodermal component (H&E, 10X).

## Discussion

Malignant extragonadal germ cell tumours are rare, comprising around 3% of all childhood cancers. The occurrence of malignant yolk sac carcinoma is very low, with an incidence of fewer than 1 case per million annually [4]. The male-to-female ratio is approximately 1:2.5 [5]. Sacrococcygeal yolk sac tumors may present as external masses or may be located internally, involving the presacral area and extending into the pelvis or abdominal cavity [6].

This case illustrates the diagnostic and prognostic importance of IHC markers in identifying malignant sacrococcygeal teratomas with yolk sac differentiation. The positive staining for AFP, SALL4, and GLYPICAN 3, combined with the high

serum AFP levels, strongly indicated the presence of a yolk sac tumor component, a highly malignant feature in germ cell tumors. Studies have consistently shown that AFP is a reliable marker for diagnosing yolk sac tumors, as it is highly specific for this tumor subtype. Research by Schneider et al. (2001) emphasized the diagnostic utility of AFP and its correlation with tumor burden [7, 8]. Our case is consistent with these findings, demonstrating both high serum AFP levels and diffuse cytoplasmic AFP staining in the tumor.

SALL4 is a well-established marker for germ cell tumors and is particularly useful in detecting yolk sac differentiation. Miettinen et al. (2014) reported that SALL4 is expressed in the nuclei of both gonadal and extragonadal germ cell tumors, including yolk sac tumors [9]. The diffuse nuclear positivity seen in our case supports the widespread use of SALL4 as a diagnostic tool.

GLYPICAN 3 is a key marker for yolk sac tumors, and its diffuse expression in our case mirrors the findings of Zynger et al. (2006), who reported that GLYPICAN 3, alongside AFP, helps confirm yolk sac differentiation in complex germ cell tumors [8]. The focal positivity of EMA in this case, while not a major feature of yolk sac tumors, is consistent with minor epithelial differentiation, as seen in some mixed germ cell tumors. Negative CK-7 staining was expected and aligns with the literature, where CK7 is typically absent in yolk sac tumors but present in other germ cell tumors like embryonal carcinoma.

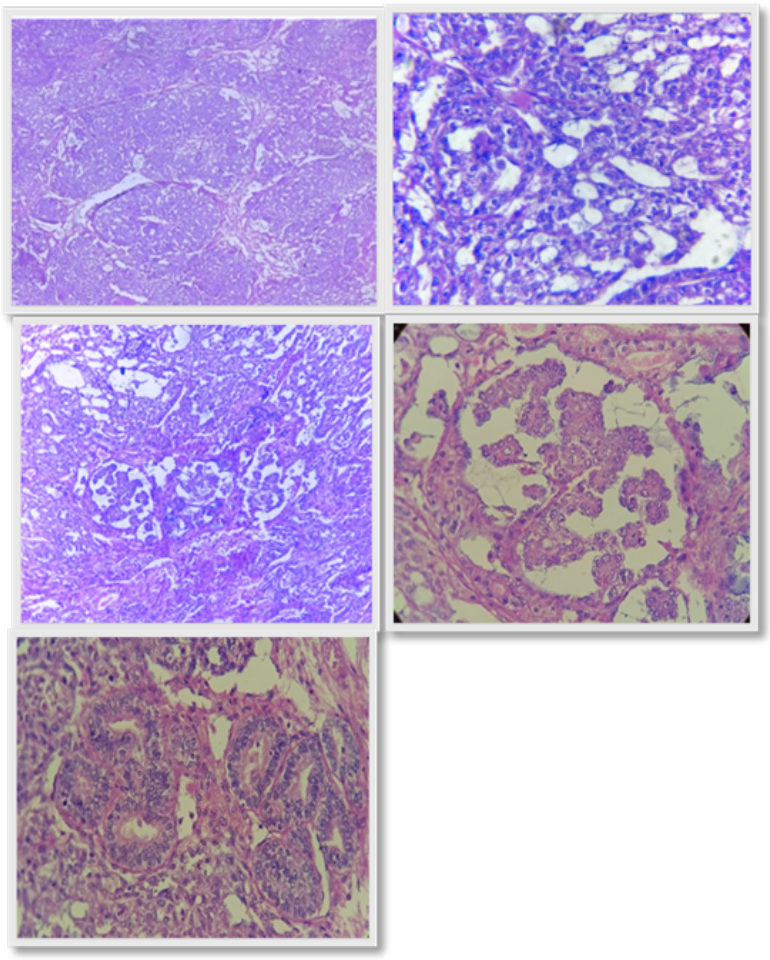

**Figure 3:** A: Showing cells are arranged in sheets (H&E, 4X). B: Showing microcystic spaces, yolk sac component (H&E, 10X). C: Showing papillary arrangement of tumour cells representing yolk sac component (H&E, 10X). D: Showing papillary arrangement (H&E, 40X). E: Showing glandular arrangement (H&E, 40X).

Prognostic Implications: The high AFP levels in this case (>5000 ng/mL) are indicative of a large tumor burden and more aggressive disease, as documented in several studies [6]. High AFP levels are associated with a worse prognosis, necessitating aggressive treatment, including surgical excision and adjuvant chemotherapy [6].

Treatment Considerations: Given the IHC findings and the high AFP levels, this patient underwent aggressive surgical resection followed by chemotherapy, typically a combination of cisplatin, etoposide, and bleomycin (PEB regimen), as per standard protocols for malignant germ cell tumors with yolk sac components. The success of this approach in improving survival has been well-documented in previous studies.

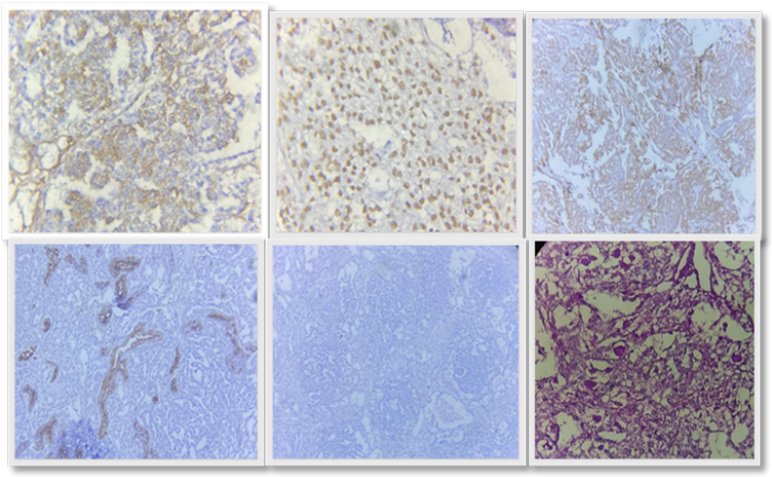

**Figure 4:** A: Showing granular cytoplasmic positivity for AFP. B: Showing nuclear positivity for SALL4. C: Showing cytoplasmic & membranous positivity for Glypican 3. D: Showing membranous positivity for EMA. E: Showing negative staining for CK-7. F: Showing PAS positive hyaline globules.

## Conclusion

This case of malignant sacrococcygeal teratoma with yolk sac differentiation highlights the aggressive nature of yolk sac tumors in pediatric patients. Complete surgical excision is critical to achieving a favorable outcome. Piecemeal excision, as in this case, increases the risk of recurrence, necessitating close postoperative monitoring of AFP levels and potential adjuvant therapy. Comparison with other studies confirms that timely, aggressive treatment and vigilant follow-up are essential in managing these cases to prevent recurrence and improve survival. Prognosis mainly depends on the age at presentation, completeness of the resection, and presence of malignant components.

**Abbreviations:** SCT: Sacrococcygeal teratoma; YST: Yolk Sac tumor; AFP: Alpha fetoprotein; FNAC: Fine needle aspiration cytology; EST: Endodermal sinus tumor; PAS: Periodic acid-Schiff stain; MSCT: Malignant sacrococcygeal teratoma.

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