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CASE REPORT

Malignant Yolk Sac Tumor in Neonate: A Management Dilemma

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ABSTRACT

Neonatal sacrococcygeal teratoma with yolk sac tumour is rare. We report a case of sacrococcygeal teratoma which was proven to be yolk sac tumor after excision. The patient developed complications related to chemotherapy. The baby was ultimately managed without chemotherapy on close monitoring.

Key words: Malignancy; Pediatric; Yolk sac tumor

INTRODUCTION

Sacrococcygeal teratoma [SCT] is the most common congenital tumour in neonates even after including still births [1]. Neonatal sacrococcygeal teratoma with yolk sac tumour is rare but incidence of malignant SCT is high when discovered after age of 2 months [2]. There is lot of controversy in medical literature about the management of neonatal SCT with malignant yolk sac tumour. We present here a case of neonatal SCT with malignant yolk sac tumour and discuss the difficulties faced during its management.

CASE REPORT

A one-day old male neonate presented to us with a swelling lower back. This baby was born at full term by normal vaginal delivery. There were no complaints other than a swelling lower back. Local examination revealed 6X8 cm cystic, lobulated sacrococcygeal swelling covered mostly by healthy skin except a small ulcerated area at the dome of the tumor. There was no evidence of neurologic deficit. Ultrasound of the sacrococcygeal region showed a predominantly cystic lesion with a small focus of a solid component. The mass was predominantly external and there was no intrapelvic extension [Altman type- I]. Alpha fetoprotein level [20880ng/ml] were in normal range for his age group [normal range for new born 4840 ±34718 ng/ml].

Complete surgical excision of the tumour with coccygectomy was done. Histopathological report showed malignant yolk sac tumour. Based on histopathology report and POG /CCG recommendations, patient was scheduled to receive four cycles of PEB regime and accordingly received first cycle after which he developed severe neutropenia and went into disseminated intravascular coagulation. Patient required care in intensive care unit for ten days and gradually recovered from neutropenia and disseminated intravascular coagulation. After this episode, we decided to discontinue the chemotherapy. Patient was kept on a close follow up without any further treatment. AFP was monitored monthly till it returned to normal adult range [< 10ng/ml]. Then AFP monitoring was done three monthly in second year and six monthly for the third year. Ultrasound abdomen and digital rectal examination and chest X- ray were done on each visit. AFP returned to normal at ten month of age. Patient did not develop any recurrence and is now seven years old without any complaints.

DISCUSSION

Sacrococcygeal teratomas are the most common teratomas in the neonate with female preponderance of 4-6:1 [3]. Most neonatal tumours are benign mature and immature teratomas that can be managed by surgery and postoperative observation.

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Complete resection of the coccyx is vital to minimize the likelihood of tumor recurrence [2].

The survey of the surgical section of the American Academy of Pediatrics noted the low risk of malignancy in neonates and young infants of less than 2 months of age [7% girls and 10% boys]; the risk of malignancy was miniscule if only the neonates with SCT were included. Yolk sac tumor or endodermal sinus tumor is the most common malignant element in SCT.

The management of neonatal SCT with malignant yolk sac component is controversial. As per NCI guidelines surgery and chemotherapy with four to six cycles of standard cisplatin, etoposide and bleomycin [PEB] is recommended [4]. AS per the UKCCSG GSII guidelines, bleomycin is to be omitted in children <1-year age [5]. Sarin et al. has reported neonatal malignant SCT in which they could successfully complete three cycles of chemotherapy [6].

Cytotoxicity of chemotherapy in neonate is always a concern. The strategy of close follow up without chemotherapy for completely resected tumour with malignant foci has also been evaluated in literature. Pediatric oncology group/ children cancer group intergroups study by Marina et al. reports only 4 recurrences in 23 patients of tumours with malignant foci treated with complete surgical excision and kept on follow up without chemotherapy [7].

Rescorla et al. reviewed six patients with Stage I SCT having yolk sac tumor components, who did not receive chemotherapy [8]. Of those six patients, two recurred and they were successfully treated with chemotherapy. Both achieved long-term survival of more than 9 years. Similarly, Schropp et al. and Gobel et al. reported good outcome in Stage I SCT patients with observation only [9,10].

Egler et al. in their review identified four Stage I and four Stage II SCT patients with malignant elements who were not treated with chemotherapy [11]. None of the Stage I SCT recurred but two of the four Stage II SCT had recurrence and were later treated with chemotherapy and surgery. They concluded that active surveillance after surgery in patients with Stage I SCT with malignant elements is reasonable.

In developing countries, the decision making become even more difficult where majority of the patients belonging to low socio- economic status and come from remote places. The loss to follow up is very common in such cases. Some authors prefer to give adjuvant chemotherapy to such patients. Considering all these facts, we also decided to give four cycles of adjuvant chemotherapy [PEB] to our patient who belonged to remote area and there was high likelihood of being lost to follow-up. Our patient developed febrile neutropenia and went in to disseminated intravascular coagulation after first cycle of PEB. The patient required prolong hospitalisation in intensive care unit and took long time to recover. After his recovery, we decided not to continue chemotherapy and put him on active surveillance. He also required telephonic reminders in between to ensure regular follow-up. Patient has not developed any recurrences after seven years of follow up.

One important parameter favouring active surveillance is Stage I SCT, where we can avoid adverse effects of chemotherapy especially in neonates. Stage II SCT has shown high risk of recurrence when not given adjuvant chemotherapy [11]. Whenever we are opting for watchful management, we should be committed for active follow up of patient by regular telephone reminders and emails.

Treatment of neonatal malignant yolk sac tumours needs individualization based on the socio economic status of the family, commitment towards follow up and resources available to manage the cytotoxic complications of the chemotherapy at the treating institute. If the decision for giving chemotherapy is made, then consideration to avoid bleomycin should be given as per UKCCSG recommendation. Considering the rarity of neonatal malignant yolk sac and lacunae in the available literature to manage such patients, a multi-centre study is needed to clear the controversy. Till then treatment should be individualised based on the available evidence.

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