

Maternal and Fetal Outcome in a Patient with Buccal Embryonal Rhabdomyosarcoma

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Abstract

Rhabdomyosarcoma is a rare malignancy that develops primarily in young individuals. We report a 19-year-old female with rhabdomyosarcoma who was classified with stage IIA disease, based on the clinical grouping system developed by the Intergroup Rhabdomyosarcoma Study Group in 1972. The patient was treated with surgery and chemotherapy. The histopathological and radiological assessment showed no residual tumor in adjacent tissues to contradict the clinical classification. The patient was scheduled for radiotherapy but left against medical advice and returned pregnant 6 months later with recurrence. She chose not to terminate the pregnancy, restricting the treatment options to chemotherapy. At 32 weeks of gestation, she underwent a caesarean section due to fetal distress. The patient deteriorated and passed away shortly after. The baby, however, is healthy. Although recent modalities of combined surgery, chemotherapy, and radiotherapy have improved the survival of such malignancies, they still have poor outcomes during a pregnancy.

Keywords

Rhabdomyosarcoma; Pregnancy; Chemotherapy; Outcome

Introduction

Rhabdomyosarcoma is the most common soft tissue sarcoma in children, though it is rare in adults; it represents 60% of soft tissue sarcoma in children^[1,2]. The majority of these tumors (90%) are diagnosed in individuals aged younger than 25 years^[1]. The most common sites of this tumor include head and neck (35%), followed by the genitourinary tract (23%) and extremities (17%). The most frequently diagnosed sites in the head and neck region excluding the orbit are nasopharynx, paranasal sinuses, middle ear and mastoid, and facial soft tissues. About 10-12% of all Rhabdomyosarcomas occurring in the head and neck

are located in the oral cavity mainly found on the tongue, palate and cheek^[2].

Adjuvant chemotherapy with surgery with or without radiotherapy improves the survival of patients with local or regional rhabdomyosarcoma; with modern combined modalities of therapy, over 70% of children with localized Rhabdomyosarcoma can be cured^[3,4]. Orbital or eyelid tumors are treated using surgical, radio therapeutic, and chemotherapeutic methods. Five-year survival rates vary between 44% and 80%^[5,6].

In this case report we discuss a 19 year old female who became pregnant during the remission

phase of her treatment. Approval for this report was obtained from the Ethical Committee at King Abdulaziz University Hospital, and permission was granted by the patient's family.

Case Report

A 19-year-old Pakistani female presented on September 23, 2006 to our hospital, complaining of a small mass in the right cheek that had gradually increased in size over 4 months. It had become painful and was associated with dysphagia and bloody offensive discharge.

Her physical examination was unremarkable, apart from a firm, tender 4x6 cm mass in the right buccal area of the face. The patients' laboratory values were all within the normal range. A magnetic resonance imaging (MRI) of the head and neck (Figs. 1A, 2A, and 3A) showed that the mass affected the entire masticator space (masseter, medial, and lateral pterygoid), pushing the right submandibular gland inferiorly with no invasion of the mandible or maxillary bone and no lymph node enlargement. A computed tomography scan of the chest, abdomen, and pelvis showed no distant metastasis.

A diagnostic incisional biopsy was performed on September 26, 2006 and the patient was diagnosed with embryonal rhabdomyosarcoma. The tumor was excised with a Weber Ferguson incision, Mandibular and coronoid process osteotomy, resection of the right zygomatic bone, and extraction of teeth #18, #48, and #47 was performed. An orbital mesh was adapted to the orbital floor.

According to the Clinical Grouping System, developed by the Intergroup Rhabdomyosarcoma Study Group in 1972, the patient was classified with stage IIA disease^[7]. Histopathological examination confirmed no residual microscopic disease contradicting the clinical staging. Two weeks later the patient began chemotherapy, comprising ifosfamide 5 g/m² and doxorubicin 50 mg/m² every 3 weeks. Six cycles were completed. Radiological imaging by MRI showed good regression of the tumor (Fig. 1B, 2B and 3B).

The patient was scheduled for radiotherapy discharged home for the weekend but failed to show for her appointment. She presented on October 28, 2007 to the emergency room complaining of a recurrence of the pain and a rapid growth in tumor size over the past 15 days. She was cachectic, had difficulty

closing her mouth, had lost vision in her right eye, and had developed right-sided anosmia.

She was admitted and began total parenteral nutrition. She was then referred for radiotherapy but was found to be 19 weeks pregnant. Thus, the radiotherapy was postponed, and the patient initiated chemotherapy (cyclophosphamide and doxorubicin). Termination of the pregnancy was rejected by the patient's family and was ill advised by the treating medical team.

The patient's condition improved after the first cycle. She received 4 cycles every 3 weeks. The patient and her fetus were monitored closely. At 32 weeks of gestation, the fetus was noted to be severely growth impaired. Her cardiotocogram showed fetal distress with persistent variable decelerations.

She underwent an emergency caesarean section under general anesthesia. Several difficulties were encountered during intubation, thus, fibro-optic intubation under sedation was performed with the patient awake. The patient gave birth to a 1.197 kg boy with an Apgar score of 6 at 1 minute and 8 at 10 minutes.

The patient was started on chemotherapy two weeks postoperatively. The patient insisted on going home, against medical advice.

She presented to the emergency room on June 6, 2008 in hypovolemic shock. She was pale, restless, and cachectic. The patient was offered admission and a blood transfusion (hemoglobin was 5.9 mg/dl), but her family refused any further medical intervention and took her home. She passed away soon after.

The baby was in good health and therefore discharged. He visited the hospital at age five months to have an umbilical hernia repaired. At follow-ups, he showed normal psychomotor development.

Discussion

Malignancy during pregnancy is a sad and distressing situation for the patient and the managing medical team. Although our patient's initial response to surgical and chemotherapeutic intervention was favorable, it was undermined by her early conception. The pregnancy created a conflict of interest with regard to chemotherapy between the mother and her fetus.

There is scant data on maternal and fetal outcomes post-chemotherapy during pregnancy. Normal

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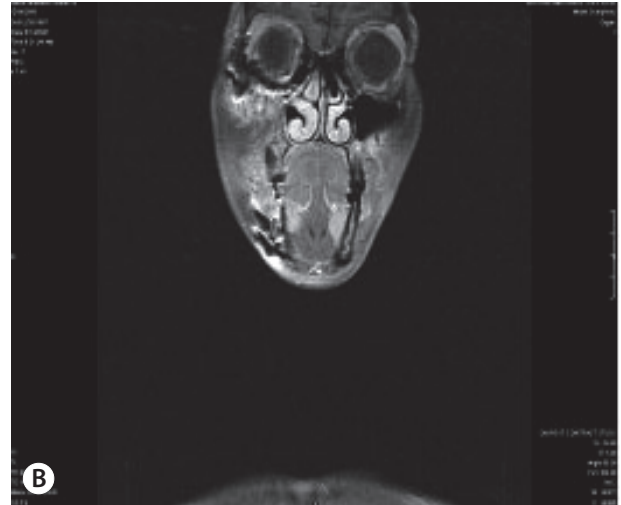
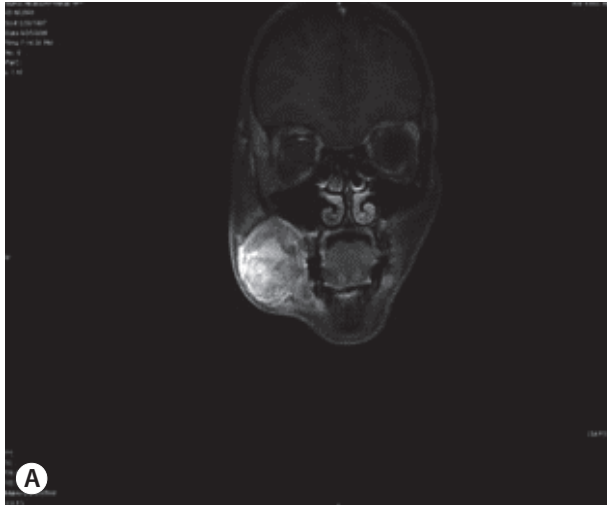


Figure 1. Magnetic resonance image of the head, (A) Sagittal section before surgery showing a mass in the masticator space 6 x 4 cm. (B) After surgery.

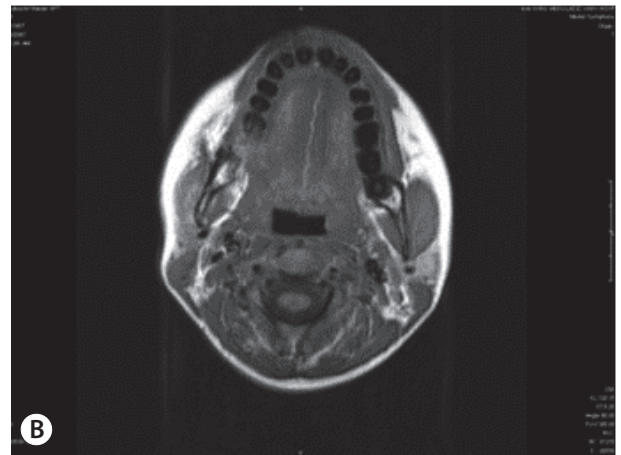
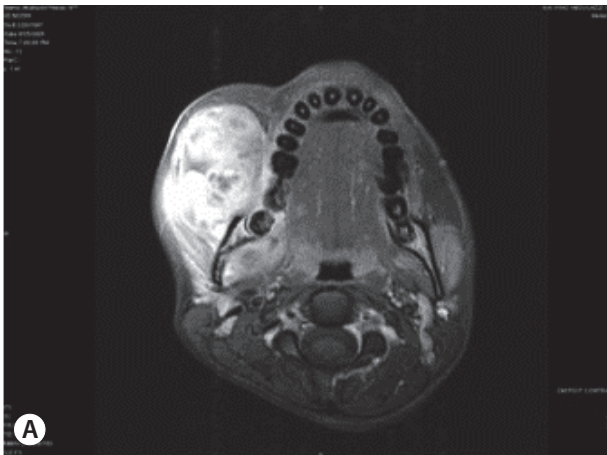


Figure 2. Magnetic resonance image of the head, (A) Coronal section before surgery, showing a mass in the masticator space 6 x 4 cm. (B) After surgery.

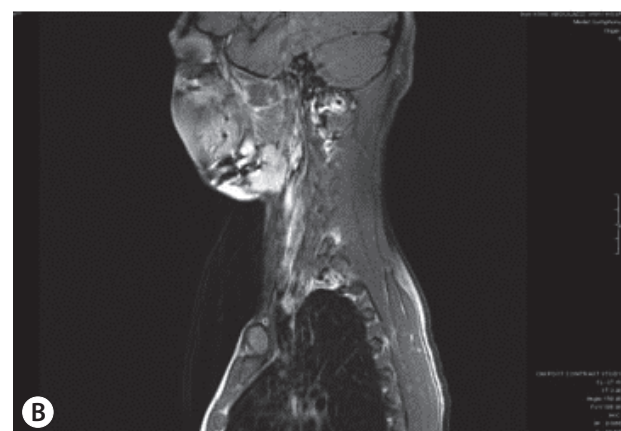
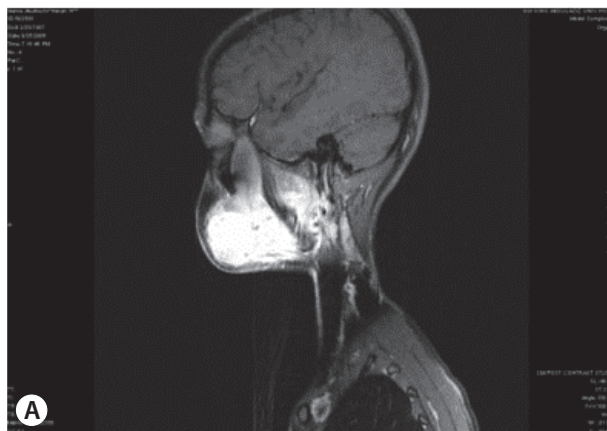


Figure 3. Magnetic resonance image of the head and neck (A) Sagittal section before surgery showing a mass in the masticator space 6 x 4 cm. (B) After surgery.

offspring have been born to mothers who received chemotherapy during pregnancy, and better outcomes are possible if exposure occurs in the second or third trimester^[8,9].

Due to the location of the tumor and the risk of intracranial extension, radiotherapy was planned for the patient immediately post-chemotherapy, but the patient married 3 weeks after the last course of chemotherapy and became pregnant immediately. She experienced rapid growth of the tumor and general deterioration; the planned radiotherapy was postponed due to the pregnancy, risking the possibility that the delay would decrease the likelihood of achieving local control in certain sites^[10].

Radiotherapy is most beneficial as an alternative to surgical management in cases where tumor removal would be disfiguring to the patient or cause loss of an essential organ such as the eye or bladder. Radiation therapy is usually given locally 6 to 12 weeks after chemotherapy, except when a tumor lies near the meninges or has extensions to the skull, brain, or spinal cord; in such patients, radiation therapy should be started immediately^[10].

Termination of the pregnancy was strongly recommended against for the patient, who presented with advanced recurrent malignancy at 19 weeks of gestation—particularly because it is not easily accepted by the Islamic population. As a nation, Saudi Arabia conforms to the more conservative Islamic schools of thought rather than more liberal Islamic interpretations. Islamic laws in such conservative countries legalize abortion at any gestational age if the pregnancy jeopardizes the life of the mother^[11]. However, this decision must be made by the hospital's medical and ethical committee and is usually weighed meticulously.

The patient delivered a severely growth retarded infant, which could have been attributed to the malnutrition associated with her medical condition or the result of the side effects of the chemotherapy.

Post-delivery, we initiated a more intensive chemotherapy regimen. Despite an initially good response, the disease had already progressed, and the patient passed away.

Conclusion

Malignancy during pregnancy is a sad and distressing situation for the patient and the managing medical team, as pregnancy creates a conflict of interest

between the health of the mother and the health of her fetus, with regard to medical interventions. Rhabdomyosarcoma is considered a very aggressive malignant tumor. Although a combined effort of surgical techniques, chemotherapy and radiotherapy can improve the prognosis, survival rates remain poor. Further worsening the outcome of the case presented here was the delay in commencement of radiotherapy as a result of the patient's pregnancy.

Conflict of Interest

The authors have no conflict of interest.

Disclosure

None of the authors received any type of commercial support either in forms of compensation or financial for this study. They have no financial interest in any of the products or devices, or drugs mentioned in this article.

Ethical Approval

Obtained.

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نتائج الأم والجنين عند مريض يعاني من الشدق الجنينية العضلية المخططة

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المستخلص. تعد سرطانات العضلات المخططة من الأورام الخبيثة التي تصيب بالدرجة الأولى الأشخاص اليافعين. في هذا البحث نلقي الضوء على شابة بعمر التسعة عشر عاماً أصيبت بهذا الورم وتم تشخيصها على أنها تعاني من سرطان الخد الجنيني للعضلات المخططة في المرحلة (الثانية أ) حسب تصنيف المجموعة السريرية المطورة من قبل مجموعة سرطانات العضلات المخططة عام ١٩٧٢م.

تم علاج المريضة جراحياً وبالأدوية الكيميائية، ولكن نتيجة التشريح المرضي للأنسجة أظهر وجود بقايا في الأنسجة المحيطة بالورم. تقرر إخضاع المريضة للعلاج الإشعاعي، ولكن المريضة قررت الخروج من المستشفى ورفضت النصيحة الطبية. بعد ستة أشهر حضرت المريضة الى المستشفى حاملاً ولديها نكسة للمرض، مما حصر فرص علاجها الى العلاج الكيميائي فقط. المريضة رفضت فكرة إنهاء الحمل. بعدها وعندما كانت المريضة في أسبوعها الثاني والثلاثين من الحمل تم اجراء عملية قيصرية لها بسبب تعب الجنين. كان المولود بصحة جيدة بينما توفيت المريضة بعد ذلك بمدة قصيرة. برغم كون التوجه مؤخراً لإستخدام العلاج الجراحي والكيميائي والإشعاعي قد حسن من نتائج العلاج لهذا النوع من السرطانات الخطرة، إلا بقي عاجزاً عن تحقيق نفس النتائج أثناء الحمل.